

F. Hefti



# Pediatric Orthopedics in Practice

R. Brunner · F. Freuler  
C. Hasler · G. Jundt

Forewords by  
J.R. Kasser  
G.D. MacEwen

 Springer

Fritz Hefti

**Pediatric Orthopedics in Practice**

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# Pediatric Orthopedics in Practice

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Translated into English from the German by Robert Hinchliffe, Lörrach

With 679 Figures (and 1164 individual Illustrations),  
79 Cartoons and 121 Tables

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ISBN-13 978-3-540-69963-7 Springer-Verlag Berlin Heidelberg New York

Bibliographic information Deutsche Bibliothek

The Deutsche Bibliothek lists this publication in Deutsche Nationalbibliographie;  
detailed bibliographic data is available in the internet at <<http://dnb.ddb.de>>.

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**Springer Medizin Verlag**

springer.com

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Planning: Antje Lenzen  
Project management: Claudia Kiefer

Translated into English from the German by Robert Hinchliffe, Lörrach

Freehand drawings: F. Freuler  
Schematic drawings: F. Hefti

Cover design: deblik Berlin  
Typesetting: TypoStudio Tobias Schaedla, Heidelberg, Germany



*For Christiane and for my three sons  
André, Philippe and Stephan,  
who have repeatedly taught me  
how children and adolescents feel  
and what their needs are.*

The English translation was made possible thanks to the financial support of the following individuals, companies and institutions:

Department of Surgery, Basel University Hospital, Switzerland  
Dr. Andreas Oeri, Basel, Switzerland  
Professor André Gächter MD, St. Gallen, Switzerland  
Dr. Alex Staubli, Luzern, Switzerland  
Dr. R. Peter Meyer, Baden, Switzerland  
Dr. Urs Kappeler, Baden, Switzerland  
Synthes AG, Oberdorf, Switzerland  
Stryker-Osteonics, Grand-Lancy (Geneva), Switzerland  
Centerpulse Orthopaedics, Münsingen, Switzerland  
Smith + Nephew, Solothurn, Switzerland  
Biochimica, Lugano, Switzerland

# Forewords

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The specialty of pediatric orthopaedics involves a vast array of primary and secondary musculoskeletal conditions influenced by growth and development. Writing a comprehensive textbook is a somewhat herculean task which Fritz Hefti and his group have done with wit and style, making this book both instructive and readable. The sometimes humorous, cartoon-like illustrations are reminiscent of those drawn by Mercer Rang. These drawings emphasize important facts, as does the bold colored print throughout the textbook.

As near as I can tell, all important aspects of pediatric orthopaedic conditions are covered in this textbook. Each new disease or section begins with a discussion of etiology, complemented by a full description of diagnostic, historical and physical findings. Diagnostic studies are recommended with an eye towards the efficient use of resources in »working up« a patient. Recommended treatment is outlined with frequent discussion of personal, Basel, Swiss, or European experience. Reference to the Basel Tumor Database and other datasets is helpful to the reader.

Whether you are searching for information on tumors, regional pediatric orthopaedic conditions or genetic diseases, you will find it in this textbook. In addition to being a reference source for answering a question about a specific disease, one can read the text in a journey through pediatric orthopaedics as it is written in a stimulating and entertaining style rather than simply a listing of facts.

I've known Fritz Hefti for 25 years and have admired his thoughtful approach to pediatric orthopaedics. Through this textbook, others will have an opportunity to be exposed to the wit and wisdom of an outstanding Swiss pediatric orthopaedic surgeon.

**James R. Kasser, M.D.,**

Chief of Orthopaedics at the Children's Hospital Boston  
John E. Hall Professor of Orthopaedic Surgery at Harvard  
Medical School

I have just finished reading this enjoyable book. I started out only to scan it but found it to be most enjoyable and educational and I learned a great deal.

Fritz Hefti has been the first who was appointed as a full Professor of Pediatric Orthopedics in Switzerland and has had a very large clinical experience at the University of Basel. He is, therefore, very qualified to include his personal indications for chosen treatment programs. His book covers all areas of pediatric orthopedics including fractures.

It is a detailed work that is educational as well as humorous. The line drawings make the points in an unforgettable way.

The references are up to date. Since the work was done in Europe it includes many important references from journals not included in the English works.

Fritz Hefti worked with me at the beginning of his career. He was an excellent clinical and research fellow and since then I appreciate him very much. He is very proficient in English and the book is easily read.

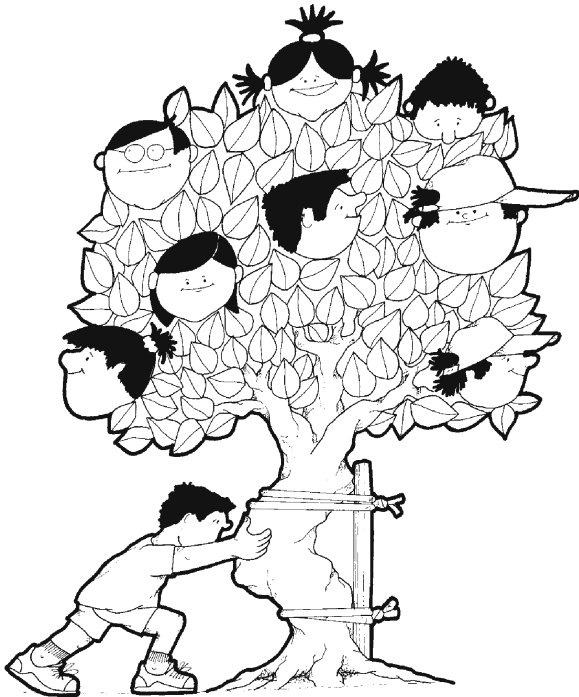
I know that you will enjoy it as much as I have.

**G. Dean MacEwen, M.D.,**

Newark, Delaware

# Preface

» Children are »patients«, not »customers«, they require »care«, not »management« ‹‹ (G.A. Annas)



The term »childhood illnesses« conjures up images of a feverish condition with red spots or a baby's teething troubles – reminiscent of the scenario with a brand new car when the engine mysteriously starts to shudder on an uphill incline – but rarely evokes crooked backs or bandy legs. Orthopaedics has long since outgrown its children's shoes, particularly since its first steps stretch right back to Hippocrates (...on clubfeet one might say). Orthopaedics has since veered in the direction of orthogeriatrics, as orthopaedists worldwide are now predominantly occupied in alleviating the infirmities of the elderly (and since bone is the »firmest« structure in the human body, »infirmities« might well be viewed primarily as an orthopaedic problem...). Nevertheless, we still need the »straight trainers«, as »orthopaedists« might be described according to a literal translation from the Greek. Trains are pushed or pulled. But trainers should not »pull« or »push« (see cover illustration) too much, since this is of limited benefit with today's children, unless the child actually wants to be pulled or pushed. Pediatric orthopaedics ultimately involves motivating children »to want to be straight« (which explains why it is the child himself

that is pushing the crooked tree trunk in the cover illustration). This requires close cooperation with parents, pediatricians, other therapists, orthopaedic technicians and nursing personnel. The idea for this book originated from pediatricians who were frequently encountering patients with musculoskeletal problems and who, during a course in pediatric orthopaedics, expressed a wish for a book that would take into account the standpoint of the pediatrician, as well as those of the children and parents. It has since grown into a comprehensive volume. Not all readers will have so much to do with »crooked« children that they will want to read everything. But perhaps they might wish to refer to this book upon encountering a specific problem. There may also be those who are not directly involved in treatment, but who would probably like to know the various available options and the corresponding factors considered in their selection. For practical purposes, this book also aims to stress the regional (rather than a systematic) subdivision of disorders. After all, a child does not come to the doctor's office saying »I'm suffering from a growth disorder« or »I have a congenital condition«. Rather he or she will say »my back hurts« or »I have a stabbing pain in my knee«. The reader will therefore find most conditions presented under the relevant body region, whereas complex diseases are addressed in a »supraregional« manner only at the end of the book. Where possible we have cited current literature sources to back up all our statements. For ease of legibility, authors' names are only mentioned in the text if they designate a classification or treatment method.

The variability in clinical pictures in pediatric orthopaedics is considerable, and no single individual can be an expert in every field. We in Basel are in a doubly fortunate position: not only is the pediatric orthopaedic department located in a children's hospital (with all pediatric specialists in-house), it is also an independent department with attending physicians in charge of their own specialist departments. My former boss and teacher, E. Morscher, realized that pediatric orthopaedics offered the greatest opportunities in terms of autonomy and, prior to his retirement, he led what was then a subdivision of adult orthopaedics into independence. In our unit the attending physicians R. Brunner and C. Hasler are primarily concerned with neuro-orthopaedics and pediatric traumatology respectively. The chapters contributed by my two highly esteemed colleagues represent extremely valuable additions to this book. My own specialist areas are spinal surgery and orthopaedic tumors. In addition to the collaboration with pediatric oncologists, my cordial relationship with the bone pathologist G. Jundt has proved particularly fruitful. He heads the Basel-based

Bone Tumor Reference Center and has contributed his considerable expertise to the corresponding chapters of the book. I have also been especially fortunate to benefit from the amicable collaboration with the privately practicing orthopaedist F. Freuler. On the one hand, he has clearly depicted the examination methods with his outstanding drawings (and in such a way that anyone can see that children are involved rather than sexless and ageless examination dolls). On the other, he has translated many ideas into visual gags with his numerous amusing cartoons. This adds a playful touch to the book which, after all, deals with children, who always want to be taken seriously, but ideally in a humorous way. Certain situations can be described much more quickly and precisely with the help of drawings than with text alone. Who would grasp the meaning of the terms »achievement by proxy« or »early childhood development program« so quickly without the drawings on pages 8 and 9? Nor is there any reason, why reading a scientific book should not also be fun. Since our brain can store information only via the emotional center (the amygdala) we should make every effort to ensure that the transmission of knowledge is associated with positive emotions, so that what is read is also stored.

I should like to thank the staff of Springer Verlag for readily accepting these illustrations, which are unusual in a textbook, and for their active support for the project. The first edition of this book appeared in German in 1997. A completely revised 2nd edition of the German version was published in 2006. This has now been translated into English by Robert Hinchliffe. He has produced an extremely competent translation, in both subject- and language-related respects, which required almost no further editing. I should like to thank him for his excellent work. The content of certain chapters has been updated since the publication of the German edition several months ago. The translation was made possible thanks to generous financial support, and the necessary funding would not have been obtained without the initiative of my friend, the orthopaedist Dr. Rainer Peter Meyer in Baden, Switzerland. He deserves my special appreciation. I should also like to thank the individuals, companies and institutions listed below for their financial contribution to the translation costs. My thanks are also due to my staff at Basel University Children's Hospital, who made many useful suggestions. Numerous ideas also emerged from discussions with the pediatricians in our hospital. Finally, I should like to thank my dear wife Christiane, who has always shown understanding for this time-consuming »leisure« activity, who also helped read through the texts and repeatedly made useful suggestions.

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## General

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## 1.1 What do the »straight-trainers« do with crooked children? – or: What is pediatric orthopaedics?

» Those who have no clear recollection of their own childhood are poor educators. «  
(M. von Ebner-Eschenbach)

So you have decided to read a book on pediatric orthopaedics – or are at least considering this as an option. You probably do not fully realize the risks involved in making such a decision. Perhaps you have already browsed through this book and noted the many schematic illustrations of impressive operations or x-rays of dreadful conditions and successful treatments. But such illustrations only relate to a small part of your work. As we all know, the term *orthopaedics* derives from the Greek words *orthos* (ὀρθός) = straight and *pais* (παῖς) = child, or *paideuein* (παιδεῖν) = to educate, or train, children. A pediatric orthopaedist would thus be considered a »straight-trainer«. But when we actually try to quantify the work of the pediatric orthopaedist the following picture emerges:

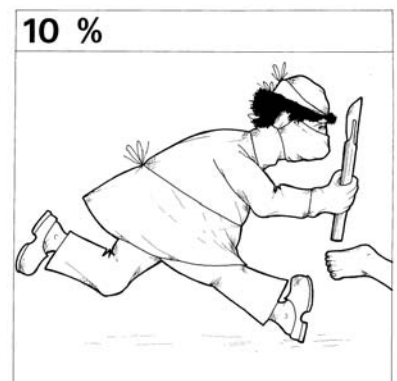
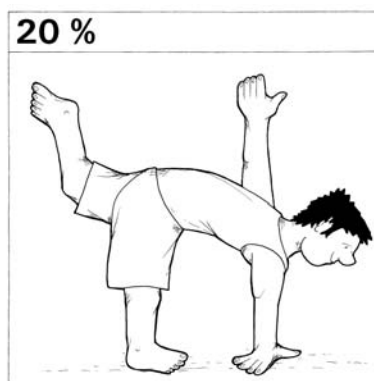
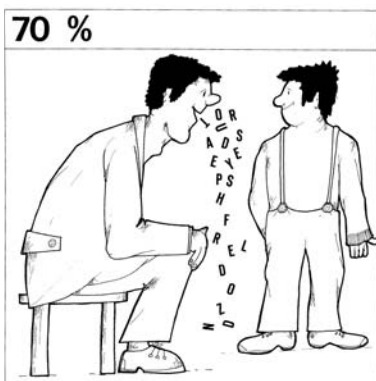
1. Orthopaedic counseling: Explaining to the parents that the child is »straight« enough, that the condition is harmless and will not have any consequences in later life: 70%
2. Conservative treatment: »Straight-training« by non-invasive means (physical therapy, plaster casts, orthoses): 20%
3. Surgical treatment: »Straight-training« by surgical means, the actual hands-on practice of the »orthopaedic surgeon«: 10%

Those of you who are orthopaedists or pediatric surgeons will think this breakdown unusual. You are accustomed to performing your handiwork with the scalpel. But this only benefits a small proportion of your patients in pediatric orthopaedics.

The second distinctive feature of pediatric orthopaedic work is that you have not just one person to deal with – the patient – but at least one or more additional individuals – the parents. This means that you not only have to understand the psychology of children or adolescents, which differs significantly from that of adults, but also that of parents, who moreover behave differently when they are worried about their children than when confronting their own illnesses.

While the reader, who is already a qualified pediatrician, is perfectly aware of these facts, there are a number of additional salient features that can seem strange even to a pediatrician: The pediatric orthopaedist is not primarily an orthopaedist specialized in »children«, but one specialized in »children and adolescents«. Some two-thirds of patients seen by a pediatric orthopaedist are adolescents. Disorders of the musculoskeletal system are the commonest reason for consulting a doctor in this otherwise extremely healthy age group. The second salient feature is the fact that bone possesses its own growth system and that actual body growth is primarily bone growth. Biomechanics provides the basis for our knowledge about the forces and their effects in respect of the musculoskeletal system. However, since relatively simple biomechanical relationships are greatly influenced by growth, the sequence of events becomes much more complex in children than is the case for full-grown individuals. In fact, it is probably naïve to imagine that only bone has a growth system and that all the other tissues grow by a kind of passive expansion.

While this view is probably incorrect, we nevertheless remain ignorant of the growth systems of other tissues and organs. There is evidence to suggest that a growth system exists in the muscles at the transition between aponeurosis and muscle tissue. But our knowledge of this system is still very deficient and these gaps in our understanding will form the subject of extensive research in the future.



### Why do parents bring their children for an orthopaedic consultation?

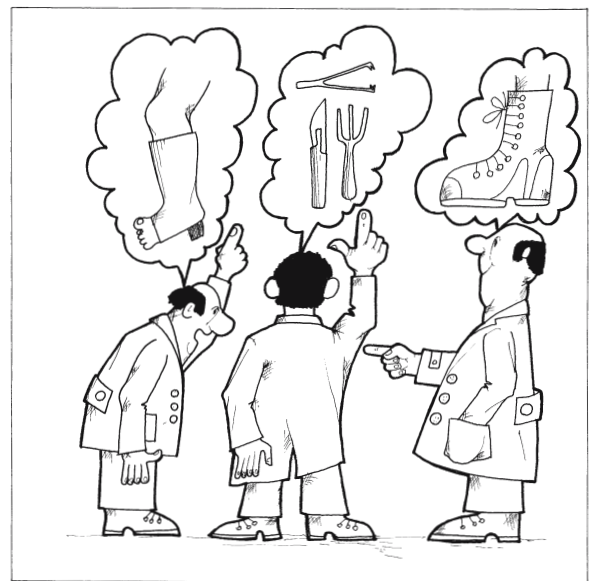
The visit to the orthopaedist or pediatrician for an orthopaedic »problem« may be prompted by the following reasons:

- The parents are worried about neglecting to do something, i.e. not starting a treatment at the right time, and thus be reproached by the child in later life as an adult. The parents fear, for example, that the intoeing gait may persist for life, that flat feet may make their child ineligible for military service in later life or that the knee pain experienced after a football training session could be an early sign of an imminent sporting disability. In many cases, the visit to the doctor is ultimately prompted by people who are not even present during the consultation: neighbors who are appalled by the »knitting needle« gait of the child, or grandparents who have compared the feet of the child with duck's feet, or even shoe retailers who justify the selling of expensive specialist shoes by citing the misshapen appearance of the child's foot. Another important reason for the parents' concern may be the experience from their own childhood, i.e. from the 1950's to the 1970's at a time when orthopaedists tended to over-treat their patients. An intoeing gait, for example, would be treated by »breaking and rotating the femur«, children with knock-knees or bow legs were forced to wear leg splints for years, and growing up without shoe insoles was only permitted to a few eccentrics.
- The parents are worried about the possibility of a tumor: If pain and/or a palpable bulge are present, there is the fear (generally unspoken) that a tumor might be involved.
- The parents are seeking support for their own rearing methods: Their children always sit crookedly on chairs or ruin their shoes in double-quick time. The parents hope that a forceful word from the orthopaedist or pediatrician will bring the children (and the shoes) to their senses.
- Referral by a colleague to investigate and/or treat a condition.
- Follow-up after a treatment or for monitoring a child's natural development.
- The parents are seeking a second (third, fourth...) opinion, having already been advised by another colleague, and are now unsure as to whether they should follow the advice or not.

One frequently asserted – but in reality non-existent – motivation for consulting the orthopaedist is the parents' »desire for treatment«. Orthopaedists, in particular, repeatedly justify the provision of treatment for a peripatellar pain syndrome, for example, by arguing that if



Some parents seek the doctor's support for their own rearing methods...



Some parents consult the doctor to obtain a second, third or even higher opinion...

they were to refuse treatment, the parents would simply go to another doctor who would be more amenable to their wishes. While parents certainly do seek the opinion of another doctor when the first has not provided treatment, the reason is not the lack of treatment, but the fact that they felt that the first doctor did not take them seriously enough. This is due to the inappropriate conduct of the first doctor. Of course, he can very probably make a diagnosis on the basis of the medical history. But

he must still examine the patient with meticulous care: Firstly, in order to avoid missing some other possible diagnosis, and secondly, to give patients and parents the feeling that they are being taken seriously. The next occasion for pushing parents into the arms of another doctor is when, after the examination, the doctor flatly states: »There's nothing wrong with your child!« Of course, there's something wrong. The child hurts and has been experiencing pain for a long time and it's getting worse all the time. The correct response in such situations is to explain to the patient and the parents that the pain is due to a very unpleasant problem connected with growth that cannot be influenced by treatment, but one that will not leave any permanent damage after the child has stopped growing. Patients will fully understand that the growing body is defending itself against overexertion and that a temporary reduction in sporting activity may be needed. The parents may still ask: »And can nothing be done to treat the condition?« I generally respond: »*Naturally something can be done, in fact a lot can be done. The question is whether it is appropriate and sensible!*« ...and, as we all know, »nothing« is the sensible response in such cases. The parents may still insist on treatment, however, because, as ambitious parents, they are unwilling to accept a reduction in sporting activity for their daughter who is, after all, about to join the regional junior team.

### **The pediatric orthopaedic consultation**

#### **Behavior of patients**

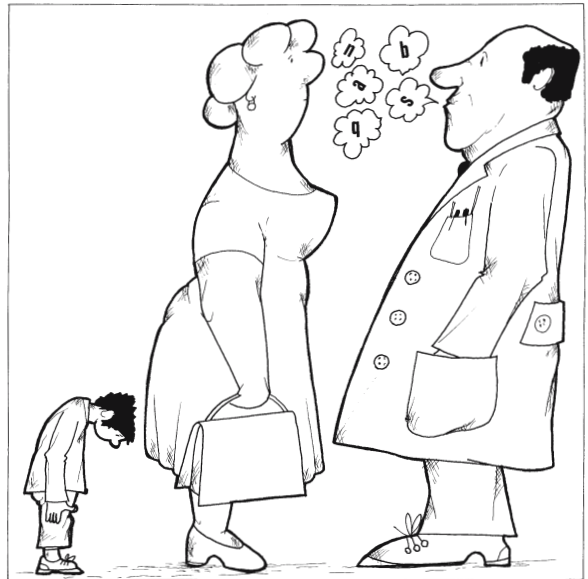
##### **Infants**

Infants generally don't care whether you're a doctor, an uncle or an aunt. What is important is that you should smile. For this means that the world is just fine – at least unless the infant is feeling hungry or thirsty. The immediate reaction of some infants is to reject unknown individuals, they just don't take to strangers, but even with these babies the odds will be in your favor if you flash them a smile.

##### **Children**

» **Children have no concept of time, hence their protracted and detailed observations.** «  
(Jakob Bosshart)

Children are extremely diverse creatures and differ fundamentally in the way they communicate with the environment of adults. They are not simply »adults on a small scale«. Children display their primary feelings spontaneously, openly and honestly. If you give an adult an injection and then ask him whether it hurt, he will probably say: »No, not at all«, and look at you in the expectation of receiving a medal for bravery. But it wouldn't occur to a child to react in this way at all, it simply yells out in pain. But in their honesty, children can also sense very



Children must always be included in the discussion...

accurately whether you are also being honest with them. If, before giving the injection, you tell the child »*this won't hurt at all*«, and then it definitely does hurt, the child will never forgive you for this deception. Why didn't you say to the child: »*This will hurt just for a moment, but it will soon be over!*«? You should always remind yourself of this need for honesty.

Children are quick to notice when you are talking about them with their parents but don't want them to hear what you are saying. Nor will they forgive you for this attitude. The parents sometimes feel that the child would not be able to cope with certain types of bad news. But if the child has a malignant tumor, who will subsequently have to cope with all the unpleasant treatment, if not the child itself? So why should it be excluded from the discussion and thus cause the child to lose the trust in the doctor right from the start? Even if they don't understand or take in everything at the initial consultation, it is extremely important from the psychological standpoint that you should include even small children in the discussion so that they never feel that you are trying to hide something from them.

Incidentally, adults find it far more difficult to cope with such news than the children themselves, because they have a much better idea of what the children will have to face. Fortunately, since most pediatric orthopaedists rarely have to administer injections, children don't categorize them as »bad doctors«. But pediatric orthopaedists do occasionally have to cause children pain, for example when removing transcutaneously inserted Kirschner wires from bones or applying a plaster cast to fresh fractures.

## 1.1 · What do the »straight-trainers« do with crooked children? – or: What is pediatric orthopaedics?

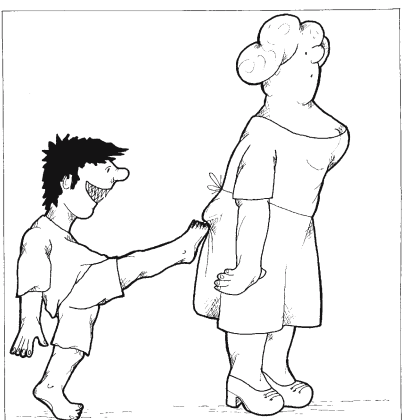
» *The reason why we adults find it so difficult to deal with the honesty of the child is that we have learned so efficiently how to lie. We all lie repeatedly; it is an essential aspect of social acceptability [1]. But children are unable to do this (yet); they have a very finely-tuned sense that tells them whether someone is telling them the truth or not, even though they may not usually be able to express directly their feelings about the truthfulness of what is being said. It is not possible to withhold the truth from children in the long term without negative consequences.* «

Children show widely differing behavioral patterns during the consultation:

— Well-behaved children will do everything that you ask of them: They will walk in the suggested direction, jump at your command, stand straight like soldiers, bend down when asked and not show any opposition to even the most adventurous contortions of the legs. In fact, most children act in this way and no great skill is required to examine them, but even well-behaved children will also appreciate a joke, a smile or a little game before the examination.

— Anxious children are afraid of the doctor. In most cases, they have previously received an injection that hurt (e.g. a vaccination) from a man in a white coat. For this reason many pediatricians these days dispense with the identifying feature of the »medicine man«, i.e. the white coat. In my experience, however, children are still able to identify the doctor in the sweater disguise as a person that can cause potential hurt. Most children therefore show a certain degree of anxiety. Especially anxious children hide their face in the mother's lap and, when asked to walk while holding mummy's hand, will suddenly disappear between the mother's legs, almost causing her to trip over, whereupon the mother will say to the child: »*You must be good now, after all you did promise me!*« At which point the child starts to cry.

This is where your skills are needed. The surest method of making any further examination impossible is to look at your watch and think about your busy schedule. Even though you may not say it out loud, the child can sense the sentence forming in your head: »*Must you behave so stupidly just at this particular time!*« Children have an incredible sixth sense for such thoughts and respond to the slightest sign of irritation



Well-behaved, anxious, defiant and hyperactive children are encountered in the doctor's office

with even more defensive behavior. You must therefore keep calm and try to distract the child with a toy (ideally one that makes a noise). Perhaps you could even play a suitable game with the child. Or you could let the mother examine the child (this only works if the mother is not impatient).

What you should never do during the examination is to lay the child down. In this position the child will feel helpless and even more anxious. What should prove successful, however, even with a crying child, is to examine it while sitting on the mother's lap. You may also manage to sit the child on the examination table next to the mother. Most examinations are possible with the child in the sitting position. With much patience, friendliness and a playful approach, it is almost always possible to perform the most important tests, calm the child and also stop the flow of tears.

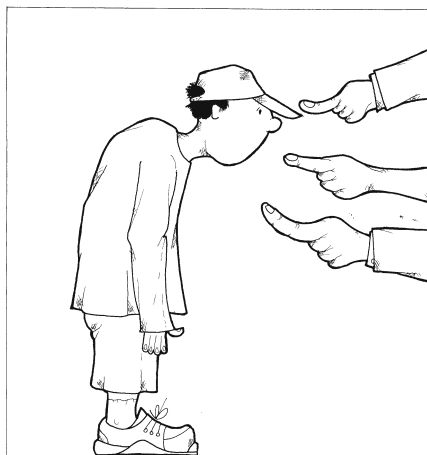
- Defiant children are similar to anxious children, they simply express their anxiety in a different way. Between the ages of 2–4 years, defiance toward the parents is, to a certain extent, physiological, since this is when an initial detachment takes place. Defiant children stamp their feet on the ground when made to undress, kick out at the mother when she pulls off their trousers, run away when asked to demonstrate their intoeing gait, dial the toy telephone when asked to stand up straight, or thrash around when the doctor tries to examine their arms. Here, too, patience, a friendly attitude and playful conversation can help produce the desired result.
- Hyperactive children will operate all the noise-producing devices at the same time while you are discussing the medical history with the mother. They will shake the armrest of your chair and possibly even climb up your back. While you are palpating the iliac crest to assess leg lengths they will get the giggles and start laughing uncontrollably because it tickles so much. In these situations also, the greatest possible calm is required. Sometimes such children can be made to listen to reason with a little game. For example, you could ask the child to learn by heart, during the examination, certain features of a picture hanging on the wall. Naturally, you must ask the child afterwards about the picture.
- Mentally handicapped children: Communication is possible even with the most severely handicapped children. The mother always knows how the child feels and what it is sensing even if the child is unable to speak. The fact that a child cannot give adequate responses should not stop you from talking to the child. Even a mentally handicapped child will notice the attention, register the friendliness in your voice and will react, possibly strongly, to physical contact, which you should not shy away from.

## Adolescents

» *Young people yearn for the future.* «  
(Jean-Paul Sartre)

Adolescents deserve to be taken just as seriously as adults. Although adolescents themselves hardly ever want to attend a consultation and tend to be pressured into it by their parents, they should nevertheless be allowed to state the problem from their own viewpoint. If the parents reply to a question posed to the young patient, the doctor should insist that the latter answers the questions. In many cases, the adolescents don't believe that there is a problem and are then »corrected« by the parents. But while parents are often the only ones to feel that something is not right, the young patients themselves will sometimes play down their problem for fear of a possible treatment.

Adolescents passing through puberty are in a phase of physiological detachment and have a tendency to revolt against adults in varying degrees, and naturally against their parents in particular. There is nothing abnormal about this. Quite the opposite, in fact, since this is a necessary phase of development. Posture plays a very strong symbolic role at this time. The muscles are not sufficiently developed physiologically to cope with the growth spurt that occurs during puberty, since the increase in muscle cross-section lags behind the growth in height and the corresponding increase in muscle length. Consequently, a certain amount of postural weakness is inevitable during this phase of development. Yet it is precisely this poor posture that often causes perpetual conflicts with authority. The constant nagging by parents exhorting the child to sit up straight provokes the adolescent to ostentatiously adopt an even more crooked posture. Mothers hold the unshakeable belief that



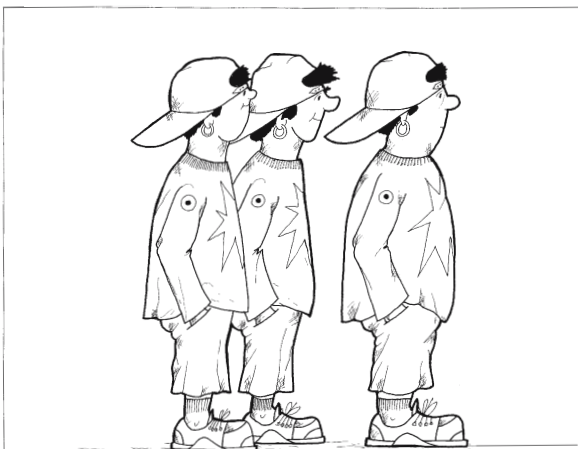
Constant nagging provokes an even more crooked posture...



poor posture can lead to scoliosis (which is absolutely not the case).

The same cannot be claimed for Scheuermann disease. Psychological factors play a significant role in this growth disorder, and the influence of an extremely dominating parent is very frequently apparent. The parents naturally expect to be supported in their constant admonitions about correct posture. However, since such admonitions are counterproductive, it is preferable to encourage the young patients, who often tend to be very passive, to take up some pleasurable sporting activity. A particular feature of adolescents is also their great need not to appear different from their peers: They have to wear the same brand of shoes, the same cut of jeans and the same type of sweater as their friends. Strict standards also apply to hairstyles within a student's class, and the earring is likewise a badge of identification. This predominant tendency of wanting to be the same as others gradually disappears after puberty, to be replaced by a greater need for individuality.

Unfortunately, this penchant for uniformity presents particular problems to those of us working in pediatric orthopaedics. Adolescents, in particular, find it very difficult to accept treatments that change their external appearance, e.g. a brace treatment. They generally prove to be the sole individual wearing a brace in their class, or possibly in the whole school. By contrast, other measures that change the outward appearance in equally unflattering terms, but which are employed much more commonly, are readily accepted: dental braces, for example, are prescribed so frequently nowadays that dentists even complain that young people without any dental problems are coming to their offices and asking to be fitted with braces just because all the other students in their class have them.



Adolescents have a great need to resemble their peers...

## Behavior of parents

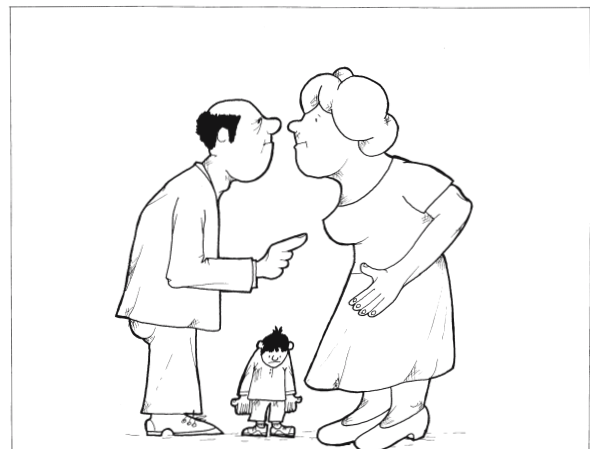
» The training of children is a profession where we must know how to lose time in order to gain it. «  
(Jean-Jacques Rousseau)

From the doctor's standpoint there are easy and difficult parents.

*Easy parents* want the best for their children, are hugely relieved when it emerges that nothing serious is present but, if their child does have a serious illness, are prepared to travel considerable distances in order to obtain the appropriate treatment, accept fairly long waiting times without complaining, are understanding in the event of difficulties during treatment, reassure the child in the face of procedures that will necessarily prove painful and leave the child in the care of the nursing staff confident that the child will be treated well. Most parents act in this way and it is always a joy to work with them.

Certain mothers and/or fathers, however, can be classed in the category of *difficult parents*:

— Parents with mutually conflicting ideas: It is not always easy to establish whether serious conflicts exist between parents during a medical consultation. Even parents who are divorced will sometimes jointly attend a consultation arranged to review a medical problem affecting their child and initially act as if they are in agreement. Only when something fails to proceed according to plan do conflicts come to the surface, with corresponding accusations being made against the medical and nursing staff. Such conflicts are always very distressing for the child and can also frequently influence the subsequent course of the illness. While orthopaedic conditions tend to be very typical somatic disorders, predominantly with well understood somatic etiologies, nevertheless the influ-



Parents sometimes resolve their conflicts through their children...

ence of the patient's mental state on the development and course of these illnesses should not be ignored. Mental stress can have very adverse consequences particularly if complications are present.

- **Conflicts with the child:** Parents occasionally have serious conflicts with their child, particularly during puberty. In many cases the cause of the problems can be traced back to the parents themselves. Perhaps the child does not fulfill the parents' expectations, whether in terms of intellectual performance or external appearance. The intoeing gait or the curved back does not correspond to the set standard and must therefore be corrected by all means. Although physical shortcomings are usually better accepted than intellectual failure, physical attributes are not infrequently interpreted as a sign of intellectual weakness (e.g. the intoeing gait).

Intellectual weakness can thus prove difficult to accept because the parents think that they are to blame. The scenario is particularly bad for children with deformities when their parents believe that this is a »punishment from God«, and that everyone can see how badly they have sinned. The deformity must therefore be corrected primarily because this provocative parading of their own sins must come to an end. Sometimes this attitude will result in the surgical correction of deformities that are of no particular importance either from the functional or esthetic standpoint (e.g. syndactyly separation on the foot).

But even parents who behave quite appropriately when it comes to the indications for surgery will often have the idea of »original sin« at the back of their mind. For this reason I avoid taking an excessively detailed history in cases of deformities occurring as a result of toxic damage during pregnancy. After all, the type of harmful substance is of almost no relevance to the nature of the damage (this is only determined by the particular moment during the pregnancy), and excessively detailed probing can unnecessarily make an already bad conscience even worse.

- **Parents requiring achievement by proxy from their child:** Sometimes parents who have failed to achieve their own dreams of great sporting, musical or other success pressurize their children into undertaking an unhealthy training regimen that doesn't really meet their needs. This occurs more frequently with girls than with boys since girls are less likely to demonstrate any great ambition.

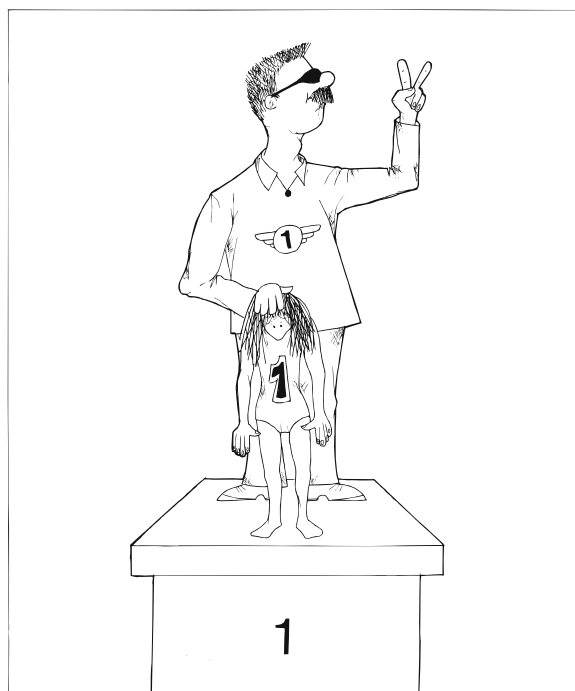
Such children, or adolescents, arrive at the doctor's office with symptoms that fail to respond to treatment. No measure proves successful. The parents become increasingly annoyed by the inability of the doctor to cure their offspring as the next competition, the one that will bring (inter)national acclaim, approaches. If you then ask the child whether the need for a medal is

really so great, the patient will reply in the affirmative, not daring to speak out against the pressure, hence the need for the disease symptom.

The treating doctor often finds it difficult to understand the real reasons for the protracted course of the illness. If you have perhaps been cajoled into arranging an operation the conflict is exacerbated, because you will then be partly to blame for the fact that the cabinet at home remains empty, instead of being filled with silver and gold trophies.

One subtype of this parent category will send their (small) children to early childhood development programs. The child must be able to play the violin by the age of 3, perform artistic tumbles on the trampoline by the age of 4 and have internalized Pythagoras' theorem by the age of 5. Brain research has discovered the huge learning capacity of children at this age and some parents now believe that it is never too early to start the learning process. While it is doubtless true that the learning ability (including for complex movement sequences) is much greater in childhood than in later life, we should not forget that the appropriate learning model for children is based on playing and not training.

- **Overstressed parents:** In many cases these are single mothers who are in employment. Children notice the



Many children (particularly girls) are pressurized by their parents to achieve sporting results that the children don't actually want themselves (achievement by proxy). Such children often respond to the pressure with chronic disease symptoms whose true causes will need to be explored...

## 1.1 · What do the »straight-trainers« do with crooked children? – or: What is pediatric orthopaedics?

constant tension and frequently react irritably and defiantly. Money is often short and every minute is planned. Any additional burden – for example a brace treatment or necessary surgery – causes the system to decompensate. This is not infrequently expressed in aggressiveness towards medical and nursing staff, and can be particularly bad if the child is handicapped. If a hospital stay is planned, social support should be arranged at an early stage.

- **Demanding parents:** These are closely related to the aforementioned subtype. Such parents are convinced that their child is the only one with a problem and that it is their duty to suspend all other activities and concentrate solely on their child. If surgery is planned, the operation must take place immediately even if no medical urgency is involved. Of course, anxiety is frequently the trigger for this attitude. Even though they may have received a detailed explanation, such parents will still telephone up to 10 times a day in order to emphasize the priority of their concern.

People are largely unaware of what the term »patient« actually means. They are often amazed to discover

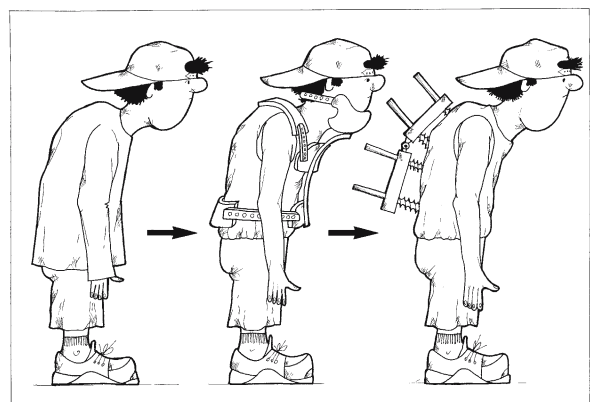
that it has something to do with »patience«. These days an illness is no longer »endured«. Rather, people expect the medical system to deliver health in double-quick time. Other parents will expect a scheduled operation to be performed on a very specific date, because school, recorder lessons, tennis camp, hockey training, best friend's birthday party or the parents' scheduled wellness weekend rule out any other date. While one should certainly accommodate the parents' wishes insofar as possible, the priorities must be based on medical considerations. Special requests or even the health insurance category should remain of secondary importance.

- **Pessimistic parents:** Certain parents are convinced from the outset that a treatment will not prove successful. This places you in a difficult situation, since you will have to be prepared for the possibility that things will actually go wrong. You would be well advised to give a detailed explanation to such parents, be very restrictive in establishing the indication for surgical treatments, and describe possible complications in great detail. This doesn't mean that you yourself should be pessimistic. A surgeon should never be a pessimist, since this would be incompatible with the practice of his profession. Nevertheless, the negative attitude of the parents will complicate matters and the blame for even the slightest complication will be laid at your door.

There are also certain parents who see problems where none exist. It is all too easy to be cornered by such parents and you should guard against this possibility. For example, you explain to the mother of an adolescent with a slight postural problem that it is harmless and will resolve itself after a little sporting activity. The mother insists: »But what if it gets worse?«. You mutter something about a brace treatment that would then



Many parents think that it is never too early to encourage (and push!) the child – but: *The grass won't grow any faster if you pull it...* (African proverb)



Pessimistic parents are convinced that everything is always worse than it is and will ultimately go wrong



produce the desired result in most cases. But again the mother asks: »What happens if the brace treatment doesn't work?«. You mention a possible operation. – ... »What happens if the operation goes wrong?« – »Well, possible complications include infections, rod failure, paralysis...«. With a cry of indignation, the mother now accuses you of initially having said that everything was harmless, but are now talking of paralysis. While remaining completely open in your explanation, you should avoid this tricky situation and not let the parent be led astray into such disproportionate conclusions.

- Parents with justified misgivings after poor results: Discussions with such parents can be very stressful. Particularly after surgical treatments, you will always feel partly responsible for the poor result, regardless of whether the indication was not completely watertight, whether the technical procedure was incorrect or whether an unavoidable complication (e.g. infection) occurred despite all the precautions. While it is only human nature to want to avoid such discussions, you should under no circumstances shirk from them.

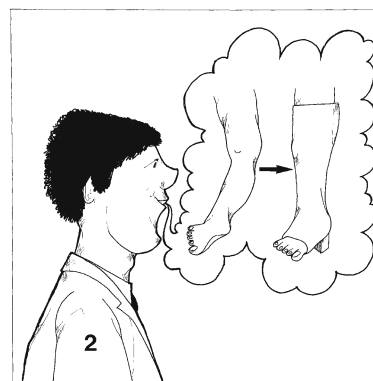
**! Of all your patients, those who have suffered complications deserve your fullest attention.**

- If patients and parents notice that you are giving their problem your complete attention, are not trying to avoid the issue and are doing everything humanly possible to minimize the negative consequences, they are much more likely to accept the setbacks, than if they have the impression that you would rather steer clear of the problem. From my experience of writing expert reports I know that it is rarely the extent or the consequences of the complication that prompt the legal liability claims, but rather the fact that communication with the treating doctor deteriorated after the occurrence of the complication.
- The parents come to you for a second opinion: Parents are increasingly less likely these days to accept the indication for surgical treatment just like that, and

therefore like to obtain a second opinion. Frequently, the health insurers will also demand this second opinion to ensure that operations are not being performed for frivolous reasons. If there are perfectly good and clear reasons to operate, your task is simple – you can confirm the opinion of your colleague. The parents will then go back to their first doctor to arrange the scheduled operation.

Your task is more difficult, however, if you have a differing opinion. Try to obtain as much information as possible relating to previous investigations. Bear in mind that the information available to the first doctor may not match your own knowledge of the facts. The parents may have presented the situation to you differently than to your colleague. Perhaps they told him that they could no longer accept the child's condition and that something just had to be done. This colleague might then have suggested an operation. The parents now tell you that your colleague has proposed surgical treatment for their child: »Is there no other way of resolving the problem?« While you should naturally not be deterred from giving your own personal opinion, you must neither protect nor disparage your colleague.

It is only natural that doctors should often have widely differing opinions, because they have had widely differing personal experiences. One or two poor experiences with a certain method or a certain indication can substantially influence the thinking of a doctor, despite the lack of any statistical basis. As the saying goes: »If two people share the same opinion, one of them is not a doctor.« This is in the nature of the profession and does not mean that any one doctor is more intelligent than another. Parents are often astonished, therefore, to discover how many different opinions emerge, particularly if they visit four or five doctors. You should not allow the previously consulted doctors to play off one against the other, nor should you feel proud if the parents talk negatively about other doctors while praising you yourself. They



If two people share the same opinion, one of them is not a doctor...

will no doubt talk about you just as negatively to the next doctor.

Express your own opinion about the treatment in accordance with your personal conviction, whether or not this differs from previous opinions. If my own opinion differs only slightly, I try to minimize the differences, explaining to the parents that I share exactly the same opinion as my predecessor and that they should follow his suggestions. This avoids any unnecessary uncertainty. Any diversity of opinion will confuse the parents. While the reasons for the various opinions may be perfectly understandable, they contribute little to the successful outcome of an operation. But if you differ fundamentally from your colleague, you should say so.

Whether to inform your colleague is a more difficult question. If the parents have no objection, it is usually a good idea to let him know of the outcome, even if you hold a differing view. But if the parents do object I will respect their wishes.

- The parents would like to obtain a second (third, fourth...) opinion: This is a legitimate wish. You should support the parents by promptly forwarding the complete documentation relating to the case, if possible, to the colleague in question. The situation is more difficult if the parents are unwilling to mention the name of the doctor they intend to contact. Your only option in such cases is to hand over all the documentation to the parents.
- The parents would like to inspect, or take with them, the medical records: The patient and the parents are entitled to view the records and make copies of them. Of course, you should not simply hand over the original files. Bear in mind that everything you

record in writing may be viewed by the parents and should be worded accordingly. Derogatory remarks are completely inappropriate. If you frequently find yourself being irritated by patients or parents and think disparagingly of them, then pediatric orthopaedics is probably not the right branch of medicine for you.

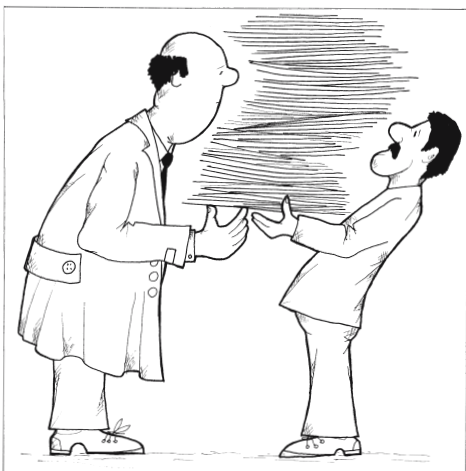
### Behavior of the doctor

**» You won't understand children unless you yourself have a childish heart, you won't know how to treat them unless you love them, and you won't love them unless you yourself are lovable. «**  
(Ludwig Börne)

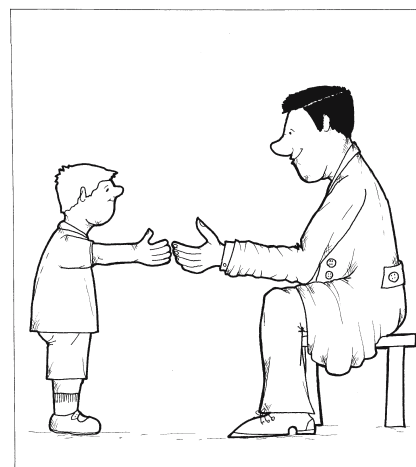
### Medical history

Whether you as a doctor will get along with a child will be decided after only a few minutes. A child wants to be taken seriously, just like every adult. Since the visit to the doctor is arranged because of a problem experienced by the child, it is important that you talk to the child, not primarily to the parents. For the pediatrician this goes without saying. But orthopaedists who deal mainly with adults tend to forget this fact all too readily. So it is almost a mortal sin to ask the parents first of all how things are, or to fail to welcome the child. As a rule, I always welcome the child first, after all the child is the leading character. Any siblings that attend will also want to be welcomed.

The first question concerning the reason for the visit to the doctor and any symptoms should likewise be addressed to the child. You must always pose questions to children in much more concrete terms than to adults: If



The patient and the parents are entitled to view the records and make copies of them



Children appreciate it when the doctor doesn't talk down to them from on high...

you ask the child: »*Why have you come to see me?*«; or: »*Have you any problems?*«, you will only receive a shrug of the shoulders in reply and the child will look at the mother inquiringly. You could ask the child whether it hurts anywhere or ask it to point to where it hurts. Nor will you receive a useful answer to the next question: »*Since when has it been hurting?*«, unless a very short period is involved (since yesterday, a week ago). But it is perfectly possible to obtain such information from the child. Make specific suggestions about periods of time that will be significant to the child, e.g.: »*Was it hurting at Christmas?*«, or: »*Did you notice the pain during the summer vacation?*«

If pain is present it is always important to establish whether or not the pain is related to loading or movement. You can likewise discover this from the child itself if you ask very specific questions. But since most children don't visit your office because of pain they will be unable to say exactly why they have come. No child says: »*I've come to see you because of an intoeing gait!*« While the reason for the visit will usually be apparent from the referral letter, it is still important to speak primarily to the children. I might ask such children, for example, whether they came by train or by car or whether they have visited the mall or the zoo. In this way the child comes to learn to trust you and feel as if it is being taken seriously.

In many cases the mother will, of course, answer the questions that you have posed to the child. However, I always insist that the child should reply by rephrasing the question differently and asking the child again. You can draw certain conclusions about the psychological situation of the child within the family from the behavior of the parents in this situation. Other fathers or mothers immediately correct the child's reply. The child might say: »*It doesn't hurt anywhere!*«, whereupon the mother says: »*Of course you hurt dear, remember the pain you felt in your knee when you were playing blind man's bluff!*« In such cases, also, the child should be given a chance to make further clarifying remarks. If the child says that nothing hurts, then the level of suffering is obviously not so serious.

Frequently the parents will dramatize the pain, while the children will keep quiet about it. Having talked to the child you will naturally want to obtain specific details from the mother or father. The parents should be allowed to present their version of the problem, but always in the presence of the child. I refuse, as a matter of principle, any request of the parents to send the child out of the room, as this would break the bond of trust. The child would feel as if it were being deceived and not taken seriously, and would sense that people were talking about it behind its back. While it is not important for the child to understand everything that is said, if the child asks for an explanation this must be provided.

## Examination

Not all children can be examined with equal ease. If you have managed to gain the child's trust during your questioning, possibly by playing with it and, above all, if you radiate calm and do not let yourself be rushed by pressure of time, you will be able to examine almost any child. Any edginess on your part will remorselessly convey to the child something that would have much less direct impact on adults.

As a rule, I perform a *full physical* examination on every child that I have not seen for more than six months. For this purpose the child will have to undress down to its underpants. Adolescent girls may keep on their brassiere or an undershirt. It is important to respect the privacy of children and adolescents. If adolescents come to the office without their parents it is advantageous to have a third person present (nurse, secretary) during the examination. This will avoid raising any suspicion of sexual abuse. A female person can help reassure the child in such situations.

A *pediatric orthopaedic examination* includes the measurement of height. As this is our most important growth parameter it should never be forgotten. Pediatric orthopaedic problems are usually long-term problems, and you will often see children over a period of years, if not decades. Since the illness changes constantly as the child grows, height as a growth parameter is extremely significant. Arm movements can be tested very summarily. I always check the pelvic tilt and examine the back in the forward-bending test (I note the fingertip-floor distance and the presence of rib or lumbar prominences).

During the forward-bending test the back can even be examined if the girl is wearing an undershirt. I also always examine the hip, knee and ankle mobility, the arch of the foot and the foot axis, regardless of the reason for the visit. A comprehensive examination of the part of the body that prompted the consultation is then indicated.

There are two reasons for this thorough examination: Firstly, it would be inexcusable for an orthopaedist to overlook a scoliosis in an adolescent girl presenting with a peripatellar pain syndrome. Secondly, the examination has an important psychological effect. As an experienced doctor you already realize, having taken the history, that a peripatellar pain syndrome is involved. If you now ask the girl to pull up her pant leg, briefly palpate the patella and then declare that nothing's wrong and it doesn't need to be treated, the patient will feel as if she is not being taken seriously and will not accept this non-treatment. As most doctors are aware of this scenario many will prescribe treatment in order to fob the patient off as quickly as possible. This ploy initially works because she has received treatment, so it must be a serious problem, and the doctor has taken her seriously even though he spent very little time on her case.

## 1.1 · What do the »straight-trainers« do with crooked children? – or: What is pediatric orthopaedics?

If the problem persists despite the treatment, the same doctor will prescribe a different treatment at the next consultation, and so it continues until the patient perhaps decides to change her doctor. The next doctor, who likewise appears to be in a mad rush, learns from the patient that three different conservative treatments have failed to banish the pain, and therefore proposes surgical treatment. The patient gives her consent because, after all, the conservative treatment proved ineffective. As a result, an unnecessary operation is performed that likewise proves ineffective, since it is unable to resolve the underlying problem, namely the muscle imbalance resulting from the increased pressure beneath the patella during a pubertal growth spurt. This is followed by more operations, until the circulation to the patella is so bad that lifelong pain is the result. Unfortunately, such cases are not particularly rare, and all this simply because the first doctor failed to take the situation seriously and spend sufficient time on the patient's problem.

Patients who are not driven by a strong, unnatural, sporting ambition (or who are not goaded on by their parents to achieve record athletic performances, see above) are perfectly prepared to accept that the peripatellar pain syndrome is a temporary problem during growth that does not require treatment. Nevertheless, they still want to be taken seriously, for it does hurt after all. If you are going to tell the patient of your intention not to provide any treatment you, as the doctor, will need to much more time to explain this than if you were to offer treatment. The complete physical examination has an important psychological effect and helps you avoid unnecessary costs and the possible consequences of surgery.

### Diagnostic procedure

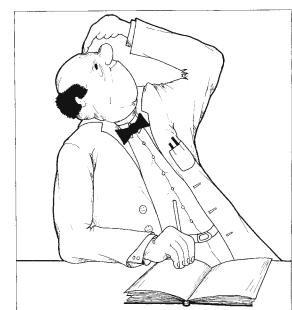
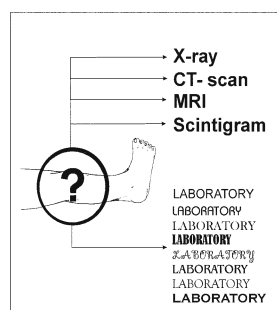
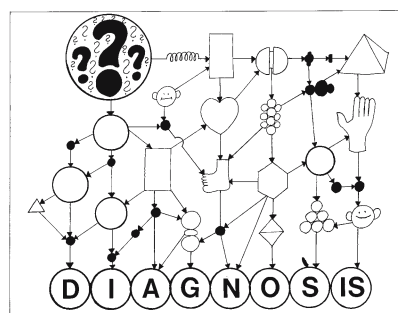
In establishing the diagnosis, most doctors proceed according to one of the following approaches:

1. Systematically according to an algorithm: Algorithms are decision trees, in some cases with complex branchings, which plot the stepwise procedure to be followed in each case according to the outcome of certain investigations. While this is certainly an efficient approach, almost no-one is able to remember such algorithms. It is fairly laborious, and there are always those patients who do not follow the specified paths of the algorithm and show findings that do not fit anywhere, obliging the doctor to pursue other avenues. Algorithms are only rarely useful in practice.
2. Investigate everything: At the onset of a symptom, the complete battery of tests is performed on the assumption that a pathological result will emerge from somewhere and thus reveal the diagnosis. This method is, alas, often employed for medical problems: All available laboratory tests are performed, the laboratory sends back the results and the pathological values are

already ticked or highlighted in red. Unfortunately, this strategy now often proves cheaper than performing targeted individual investigations.

This approach is also possible in orthopaedics. The patient presents with knee pain and the doctor prescribes a bone scan, a CT scan and an MRI. The radiologist will then report on the site of the problem. In my view this is the most indiscriminating way of practicing medicine. It is also hugely expensive and therefore unacceptable in the face of increasing cost pressures on the healthcare system. Only rarely will you establish the correct diagnosis by this method.

Imaging procedures are only meant to support the clinical examination findings. Only for a limited number of conditions is the radiologist, who is unaware of the patient's clinical situation, able to correctly evaluate and rate the changes on the images. Since he is, moreover, under pressure to provide a diagnosis, he will also tend to describe possible findings as pathological, instead of assessing them as not relevant. Hardly any patient undergoes an MRI scan of the knee without a meniscal lesion being discovered. The discrepancy between radiological findings and clinical relevance is at its most extreme for degenerative changes of the spine. In this situation the x-ray on its own is almost meaningless. While the situation is not so extreme in pediatric orthopaedics, most findings can still only be assessed in connection with the clinical examination.



The diagnosis can be determined systematically according to an algorithm, according to the »investigate everything« principle or intuitively...

3. Intuitively: A hypothetical diagnosis is made on the basis of the signs and symptoms. This initial diagnosis is then further explored in order to get to the root of the problem. Most doctors proceed according to this method, which is definitely the most sensible approach in practice. In pediatric orthopaedics, however, it is important to assess the degree of urgency before exploring the problem.

In many cases, the course of the disease will decide on the diagnosis. Legg-Calvé-Perthes disease will only be visible on the x-ray after several weeks, whereas changes will be apparent on a bone or MRI scan even in the initial stages. Thus, if you are not sure whether transient synovitis of the hip or Legg-Calvé-Perthes disease is present in a 4-year old boy who has been suffering from hip pain for 1 week, you might make the diagnosis after an MRI scan, although the actual diagnosis would not have any consequences for the treatment. At any rate, I would not treat a case of Legg-Calvé-Perthes disease in the early stages unless restricted hip motion were also present. If this finding persisted for more than 2 weeks I would refer the patient to a physical therapist, regardless of whether the patient was suffering from Legg-Calvé-Perthes disease or some other condition. Since the Perthes disease would readily be diagnosed on the basis of conventional x-rays six weeks later, we could have spared the child from having to undergo an expensive MRI scan.

**! We should proceed according to the following principle: We should never order a diagnostic measure if it is clear from the outset that the result will not have any therapeutic consequences. The more uncertain the doctor is, the more unnecessary diagnostic procedures he will order and the less clear will be the resulting diagnoses.**

Before a treatment can be initiated, the *diagnosis* must be *explained* to the parents and the child. The child must *always* be present during this part of the consultation. In my view, it is unacceptable to send the child out if a bad diagnosis is involved, e.g. a malignant tumor. Since the child is the one that will have to undergo the whole treatment, it would be inconceivable to conceal the diagnosis. Children regard it as a breach of trust to talk about their own problems behind their back. Parents will sometimes find it difficult to accept this situation, but will perfectly understand once the necessary explanation is forthcoming.

Always avoid using unfamiliar words when talking about the diagnosis. If the listener does not understand what the speaker means, this never indicates that the listener is too stupid to understand, but rather that the speaker has been unable to express the main elements in simple terms that the listener can understand. In

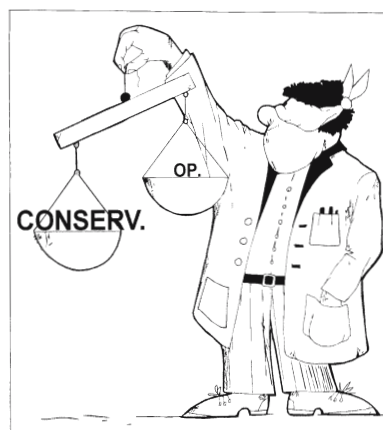
your explanation you should also avoid the use of certain negatively loaded or anxiety-triggering words, e.g.: »deformity« (better: malformation); or: »tumor« (better: swelling); or also the words: »crippled«, »deformed baby« or »feeble-minded«!

### Treatment

Many parents ask whether nothing can be done to resolve the child's problem. While, in our experience, 70% of pediatric orthopaedic problems do not require any treatment, one can, of course, always do something. The question is whether that something is appropriate. A treatment will always be judged against the spontaneous progression of the illness and should only be prescribed if it will produce a better result than this spontaneous outcome. The therapeutic objectives should always be clear and also discussed with the parents. Parents often have very unrealistic expectations and believe that their deformed child can be made completely normal again, that a leg that is shorter by 20 cm will be a perfectly normal leg once it is lengthened, that their paralyzed child with the myelomeningocele will be able to walk again with agility, or that the child with cerebral palsy can be made completely normal. This situation must be addressed and steps taken to counter such unrealistic hopes. Any hint of such starry-eyed notions must be corrected.

Most parents find *conservative treatments* to be much more acceptable than *surgical procedures*, even though conservative measures can sometimes be more drastic than surgical treatment. For example, I personally consider that treating a child with Legg-Calvé-Perthes disease with an abduction orthosis for 2 years can sometimes be more drastic and stressful than an trochanteric varus osteotomy involving a 10-day hospital stay. The effect of the surgical procedure is absolutely identical.

Occasionally it may be necessary to prescribe a conservative treatment even in the knowledge that subsequent surgery is inevitable, simply to make the operation more acceptable. This particularly applies with scoliosis





patients. If a girl with a scoliosis of more than 40° and who has not yet reached the menarche attends the office for the first time, then surgery is indicated in principle. But considerable sensitivity and tact is needed to detect whether the parents would accept this measure. Parents are often so shocked by this suggestion that they refuse the whole treatment. In such cases it is sometimes more appropriate to start with a *brace* treatment and tell the parents that surgery will be necessary if the condition progresses despite this treatment. The parents will then feel that everything has been tried in order to avoid an operation.

When surgery is indicated, the parents must be given a detailed *explanation* of the operation. They should understand the principle of the operation, they should know what outcome to expect compared to the spontaneous course of the condition, and they should be informed of any alternative therapeutic options. They should be aware of the most important complications, i.e. both those that occur with particular frequency and those that are particularly serious. They should also be given information about the circumstances of the hospital stay and follow-up treatment. This explanation must be provided at the time the indication is established and should always be given verbally. This can be backed up by an

information sheet that ideally describes the scheduled operation in specific detail.

For elective procedures the comprehensive explanation should always be provided in the office once the indication is established and not delayed until the day before the operation. In the immediate preoperative period the parents will feel under pressure and lack the courage to refuse an operation because they were unaware of a particular risk. We nevertheless restate the risks on the day before operation and record them in writing (together with the parents) on a sheet of paper, which is then signed by the parents (for legal reasons).

When information is being given about the treatment, certain anxiety-generating terms should be avoided, e.g.: »*break the bone*« (better: divide the bone) or »*cut the tendon*« (better: lengthen the tendon). The child should be included in the discussion of the treatment. After all, since the child is the one that will have to »suffer« the treatment, it should know what it will have to face.

### **What orthopaedic problems are encountered in children and adolescents?**

The risk of children or adolescents having, or acquiring, a problem with the musculoskeletal system is approx. 90% (■ Table 1.1). Accordingly, the risk of encountering an

■ Table 1.1. Orthopaedic problems in children and adolescents. (Figures from [2] and personal experience)

	Incidence (%)	Conservative Treatment <sup>a</sup> (%)	Surgical treatment (%)
Fracture	50	45	5
Flat feet (physiological)	20	0.5	0
Anteverted hip	15	0	0.1
Benign tumor <sup>b</sup>	15	0	0.1
Spondylolysis	5	1	0.1
Metatarsus adductus	3	2	0
Scoliosis >10° (girls)	3	0.5	0.15
Hip dysplasia	2	2	0.1
Cerebral palsy	0.2	0.2	0.1
Clubfoot	0.15	0.15	0.15
Trisomy 21	0.14	0.01	0.01
Legg-Calvé Perthes disease (boys)	0.13	0.13	0.07
Slipped capital femoral epiphysis	0.05	0.05	0.05
Malignant tumor	0.002	0.002	0.002
Neurofibromatosis	0.002	0.001	0.001
Osteogenesis imperfecta	0.001	0.001	0.001
Achondroplasia (and all other skeletal dysplasias)	<0.001	<0.001	<0.001

<sup>a</sup> Including physiotherapy.

<sup>b</sup> Non-ossifying fibroma of bone, in particular, is very common in childhood.

orthopaedist is very high. Around 50% require conservative treatment (most commonly a plaster cast for a fracture). Less than 10% of children will need surgery, half of whom will likewise require a fracture repair.

**Why, despite all the risks, is it still such a pleasure to work in the field of pediatric orthopaedics?**

- Sonja is happy because she had been so anxious about the possibility of requiring surgery for her knee pain, but now she only has to reduce her running program slightly. While she still experiences the occasional knee pain, she does not feel greatly bothered by it.
- Six years ago Kevin was diagnosed with an osteosarcoma of the femur. Today he can walk without a limp. Although he cannot take part in sports, he is satisfied with his current situation. He has completed an apprenticeship in electrical engineering and now works for a company specialized in metrology.
- Françoise was born with a deformed left leg. By the onset of puberty this was 15 cm shorter than the right leg. Today, aged 16, and after protracted treatment, both legs are the same length. Although she needs a splint and limps noticeably when tired, she is satisfied with her situation. I have known Françoise and her family since she was born. I also attended her school play and know some of her school friends.
- Sakine entered the world with a dislocated hip. When she came to us she was already 2 years old. She had to undergo 4 operations and remain in hospital for many weeks. Today she is 17. When a change in the weather occurs, she notices her hip. She frequently returns to

the hospital and is pleased to see the nurses that she has come to know very well.

While Sonja, Kevin, Françoise and Sakine may not yet be completely healthy and free of symptoms, we have been able to help them in some way and they are grateful in return. Moreover, we have known Kevin, Françoise and Sakine for many years, and they also tell us their private joys and worries. The very fact that we repeatedly see the same children and adolescents with serious musculoskeletal problems over many years and that we also become well acquainted with one another over time is a particularly pleasurable aspect of pediatric orthopaedics. Pediatricians and general practitioners are also familiar with this positive aspect of the job, while surgeons in other disciplines rarely have the opportunity to observe their patients over such a long period and develop such a close relationship.

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**1.2 Orthoses, prostheses, theories and inventive individuals – a historical review**

» Those who cannot remember the past are condemned to repeat it...! «  
(George Santayana)

» The past must be a springboard not a sofa «  
(Harold Macmillan).



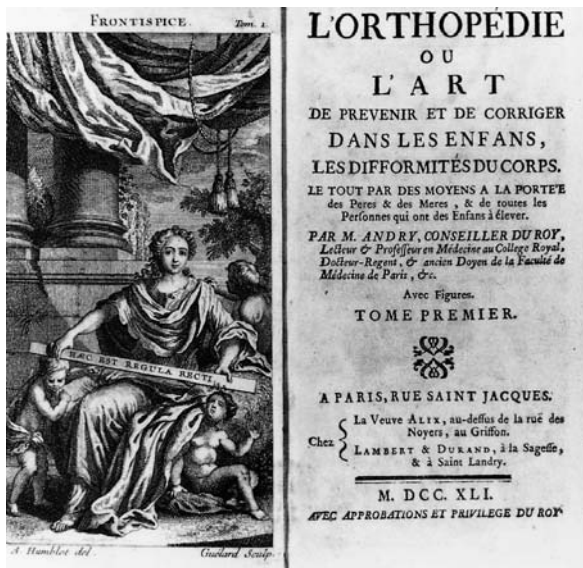
Sonja, Kevin, Françoise and Sakine

Disorders of the musculoskeletal system are thought to have been a concomitant of the human condition ever since we arrived on the scene some 5 million years ago. Some orthopaedic conditions are linked with a very specific feature of human evolution, i.e. the straightening of the spinal column and walking on two legs. This particularly applies to spondylolysis and idiopathic scoliosis, neither of which occurs in animals. But degenerative spinal conditions also originate largely from the fact that the spine stands erect. The history of orthopaedic treatment is much shorter and dates back to antiquity.

The development of orthopaedics has always primarily been that of pediatric orthopaedics. The term »orthopaedics« is known to have been coined by Nicolas Andry in his book »L'Orthopédie ou l'art de Prévenir et de Corriger dans les Enfants, les Différences du Corps. Le Tout par des Moyens à la Portée des Pères et des Mères, et de toutes les Personnes qui ont des Enfants à élever« (1741) [9] (■ Fig. 1.1 and 1.2).

The Greek root »pais« refers to the »child« and the idea incorporated by this term was that of »straight-

training«. Only in the last 30 years has the emphasis in orthopaedic therapy clearly shifted from pediatric to adult orthopaedics, and the treatment of degenerative disorders (particularly arthroses) is now more significant, in terms of number of patients, than the treatment of childhood diseases and musculoskeletal injuries. Developments in orthopaedic conditions over recent decades are described in ► chapter 1.3.



■ Fig. 1.1. Nicolas Andry: Title page of the book *L'Orthopédie ou l'art de Prévenir et de Corriger dans les Enfants, les Difformités du Corps. Le Tout par des Moyens à la Portée des Pères et des Mères, et de toutes les Personnes qui ont des Enfants à élever*, 1741. The woman holds the »Regula recti«, or straight rule, for measuring the straightness of children



■ Fig. 1.2. Nicolas Andry: The famous illustration from the book *L'Orthopédie ou l'art de Prévenir et de Corriger dans les Enfants, les Difformités du Corps*, 1741. The crooked tree splinted by a rope to a straight post has become the globally recognized symbol of orthopaedics

From very early times, people have wanted to replace missing or defective limbs with *orthoses* and *prostheses*. This has required some *inventive individuals*. Naturally, such people were also needed in the development of operations. The discipline of biomechanics has emerged as a theoretical basis, and many proposed *theories* have prompted the development of treatments, though not all schools of thought have subsequently proved to be correct.

### **Orthopaedic diseases through the ages**

Orthopaedic diseases can be traced back to the beginnings of human history, because the actual supporting structure for the locomotor apparatus, i.e. the skeleton, can remain preserved for millions of years. Two pathologies in particular have repeatedly been observed in archeological finds dating back to the Paleolithic Age: changes attributable to tuberculosis of the bone and post-traumatic conditions. Thus, spinal columns with collapsed vertebral bodies and gibbus formation in particular have been found [9]. There have also been a number of observations from that period of post-traumatic changes following femoral, pelvic or vertebral fractures [8]. Interestingly, spinal finds with degenerative changes have been unearthed from the Neanderthal period [8].

Humans evidently paid the price for their upright gait at an early stage. The clinical conditions observed become more diverse in the Neolithic Age, particularly in ancient Egypt. In addition to numerous tuberculous and post-traumatic changes, one skeleton was discovered with signs of ankylosing spondylitis. Numerous cases of clubfoot and equinus deformity of the foot, as well as hereditary diseases, have been observed in mummies. The visual art of ancient Egypt reveals numerous images of dwarfism, and even Ptah, the Egyptian god of the dead, and the god Bes are often depicted as (achondroplastic?) dwarfs [9]. Certain illustrations show evidence of the presence of poliomyelitis [1, 3, 7–9].

*Hippocrates* (born in 370 BC) was the first to provide a written record of diseases. He would have been well acquainted with congenital conditions such as clubfoot, hip dislocation and scoliosis. Traumatology and tuberculosis played an important role even during that era. Bone curvature in children was described in the early post-Christian period (around 110 AD) by *Soranus of Ephesus* [9] – evidently referring to rickets. It was only in 1650 that this disease was eventually described in detail in Glisson's treatise. Rickets doubtless played a substantial role in past centuries, primarily in northern countries.

Nevertheless, while Glisson believed that the curvature of the spine was also attributable to this condition, this was likely to be the case in only a few instances. Most cases of scoliosis, even at that time, were probably »idiopathic« or neurogenic in origin. Then, as now, the cause of these conditions remains unexplained. The only difference is that we can now describe it in more sophisti-



1 cated terms. At all times, however, paralytic scoliosis due to poliomyelitis has probably been more prevalent than idiopathic scoliosis. It was not until the introduction of vaccination at the start of the 1950's that this disease was finally eradicated, first in the industrial nations and nowadays also largely in the developing countries.

Clubfoot has remained a common condition across the centuries. Only in recent years has there been a decline in its incidence. A similar situation also applies, incidentally, to idiopathic scoliosis. The frequency of inherited systemic disorders is very closely dependent on the degree of relationship of the parents and is therefore also indirectly influenced by religious, cultural and social conceptions. Incest was quite usual in ancient Egypt. The idea that incest might be sinful only emerged in the Old Testament. The consequences of marriage between relatives were no doubt observed and clear conclusions drawn. Thus we read in Deuteronomy 27, verse 22:

» ***Cursed be he that lieth with his sister, the daughter of his father, or the daughter of his mother.*** «

The taboo of inbreeding has persisted in the Jewish and Christian religion to the present day. This taboo is less strict in the Islamic social order and is also less likely to be observed in certain primitive peoples. As a result, hereditary diseases are more common in these societies, although such illnesses – particularly among primitive peoples – have not become a social problem. Even today in certain tribes, children with obvious birth defects are abandoned and left out to be killed by wild animals. This also applies to infants with Little disease or other types of cerebral palsy.

In Europe we probably see more of these kinds of children nowadays compared to earlier centuries, when children who were evidently failing to thrive were left to their fate. The proportion of infants with mild cerebral palsy attributable to difficult births has declined thanks to improvements in obstetrics and neonatology. In births with a high risk of complications, the decision to proceed to cesarean section is now taken at an early stage.

However, the proportion of severe cerebral palsies has not declined, but rather increased. This generally involves children with cerebral malformations who would not even have been capable of surviving at all in the past, but who now receive treatment. Bone tumors have likewise always been with us, although these were neither correctly diagnosed nor treated in previous centuries. Patients with such conditions tended to be left to their fate. We have no evidence to suggest that the incidence of these tumors has changed over time.

### **Conservative treatment**

The history of the conservative treatment of orthopaedic conditions starts with *Hippocrates*. Although fractures were doubtless splinted and bandaged well before this Father of Medicine appeared on the scene, we lack the writ-

ten or graphic portrayals of such treatments. Only the use of crutches has been depicted repeatedly in records from ancient Egypt [2, 3]. But the era of corrective measures starts with Hippocrates. He described corrective manipulations similar to those that are still in use today. He also recommended the application of a bandage to exert a corrective action and prescribed shoes that were capable of correcting the position of the foot.

Hippocrates was also doubtless familiar with congenital hip dislocation, even though he was unable to offer a corresponding treatment. For curvatures of the spine he recommended the following treatment: The patient is tied to an upright ladder either by the feet or around the chest. This ladder is then repeatedly raised using ropes and allowed to fall under its own weight. Evidently this involved the application of the extension principle [9], which was subsequently described in the book *Chirurgia è Graeco in Latinum conuersa* by Guido Guidi (Vidus Viduus, approx. 1500–1569) in 1544 (■ Fig. 1.3).

Plaster treatment was introduced by Arabian doctors around the 10th century AD. While fractures were treated with this material right from the start, this application of plaster only reached Europe at the end of the 18th century [9].

The options for conservative treatment were neither significantly extended nor refined during the Middle Ages. Although the archetype as it were of the *brace* was created with the arrival of medieval iron armor, this did not have any corrective effect, nor was it used as a therapeutic device. Corrective splints for treating contractures of the knee or elbow joints were described and depicted by *Hans von Gersdorff's Feldtbuch der Wundartzney* (Strasbourg 1517) (■ Fig. 1.4). These are very reminiscent of a knight's armor. Actual braces appeared in the 16th century. *Ambroise Paré* (1510–1590) treated cases of scoliosis with braces made from thin plates of perforated iron in order to minimize weight [9]. The extension principle was refined by *Francis Glisson* (1597–1677) with his swinging or suspension device. Even today, the Glisson sling is still to be found in orthopaedic hospitals. Traction beds also subsequently came into widespread use. Braces, made primarily of metal, wood and fabric, were constantly refined.

Then, in the 20th century, came the arrival of plastic, a lightweight, dimensionally-stable material. An important milestone was reached in the 1940's with the development of the Milwaukee brace, which operates according to the principles of both extension and correction. Subsequent brace developments were limited to the application of the correction principle, for example in the very popular Boston brace. Traction beds were also frequently used for the treatment of spinal deformities.

The correction principle employed for *clubfoot treatment* also hardly changed at all for centuries after *Hippocrates*, even beyond the Middle Ages. Pioneering work

in this field was achieved by *Ambroise Paré* with the development of a clubfoot splint. This and other splints of the time were able to maintain a particular position to a certain extent, but produced almost no corrective effect. This was only achieved by *André Venel* with his »sabot de Venel«. This boot, which was the archetype of all current clubfoot splints, produced an actual corrective effect. The correction of clubfoot with plaster casts was only subsequently introduced in the 19th century.

*Congenital hip dislocation* is a condition whose dissemination is closely associated with civilization. It is largely unknown among primitive peoples, but has been known in Europe, particularly Central Europe, since ancient times. The condition is even mentioned by *Hip-*

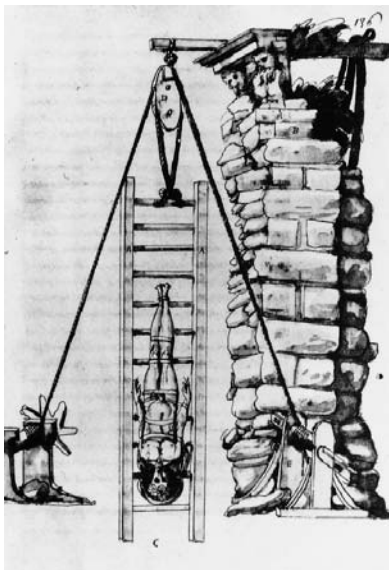
*pocrates*. The congenital aspect of the problem was only established in the 17th century (*Theodor Kerckring* 1640–1693, *Theodor Zwinger* 1658–1724). At that time, so-called »bone-breakers« would try to correct the deformity, apparently with little success [9]. The first successful attempts at closed reduction were achieved by *C.G. Pravaz* in around 1842 [9]. The work of *Adolf Lorenz* (1854–1946) also represented a milestone in the treatment of congenital hip dislocation. His bloodless method of reduction with retention of the patient in a frog-leg plaster cast developed at the end of the 19th century was, for many decades, the standard method for the early treatment of congenital hip dislocation. It was not until 1968 that this plaster treatment was finally replaced by the less pronounced abducted position in a pelvis-leg cast described by *Fettweis* and associated with a reduced risk of femoral head necrosis. Other therapeutic landmarks included the development of splints (*Hilgenreiner*, *Brown*) and bandages (*Pavlik*, *Hoffmann-Daimler*).

The *treatment of fractures* by splinting dates back to ancient times. Numerous illustrations from the earliest historical records testify to the existence of such treatments [3, 7, 8]. The extension principle also dates back to that period. In the 19th century, the fixation technique was significantly improved with the introduction of plaster. The actual plaster of Paris cast was invented by the Dutchman *Antonius Mathysen* in 1851. A particularly discriminating approach to fracture management, with standardization of treatment according to the type of fracture, was developed by *Lorenz Böhler* in Vienna at the start of the 20th century.

The history of *prosthetics* likewise dates back to ancient times. *Pliny the Elder* relates how the Roman soldier *Marcus Sergius* lost his right hand in the Second Punic War (218–201 BC) and ordered an »iron hand« to be fashioned so that he was able to return to active duty in later military campaigns. In the Middle Ages, the use of prostheses as replacements for arms and legs was widespread, in the latter case generally in the form of peg legs [8] (Fig. 1.5).

One famous prosthesis wearer was *Götz von Berlichingen*, who had lost his right hand in the Landshut wars of succession (1504–1505). Prince *Frederick of Homburg* (1633–1708) wore a silver artificial leg. The options for prosthetic production were substantially increased by *Otto Bock* (1888–1953), who designed a system for the mass production of individual functional components. Prosthetic joints allowing much smoother movement, particularly of the lower extremity, were also developed around this time.

The above-mentioned *André Venel* also achieved pioneering work in another field by establishing the world's first *orthopaedic institute* in Orbe (Canton of Vaud, Switzerland) in 1780. This institute provided conservative treatment exclusively for children with orthopaedic conditions. In Germany, *Johann Georg von Heine*, was the first to open an orthopaedic hospital, in 1812 in Würzburg. In France, *Jacques Mathieu Delpèch* founded an orthopaedic institute



■ Fig. 1.3. Vidus Vidius: Hippocrates' *rachiotherapy* (scoliosis treatment), from: *Chirurgia è Graeco in Latinu conuersa*, 1544. The patient is tied to an upright ladder. This ladder is then repeatedly raised using ropes and allowed to fall under its own weight.



■ Fig. 1.4. Hans von Gersdorff: Corrective knee extension, from: *Feldtbuch der Wundarztney*, 1517

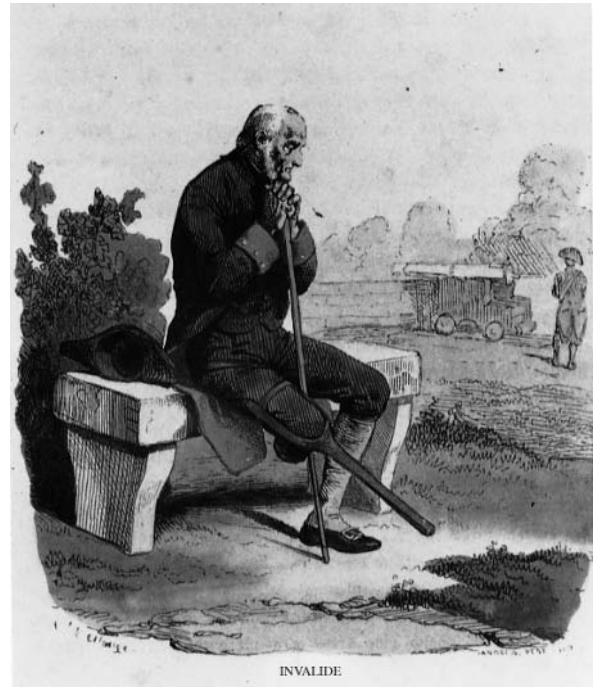
in 1825 in Montpellier, while *Jules-René Guerin* and *Charles-Gabriel Pravaz* began their work in an orthopaedic hospital in Paris in 1826. *Delpech* (1777–1832) is also considered to be the actual founder of the science of orthopaedics.

In England an orthopaedic institution was founded in 1837 by *William Little*. The first American orthopaedic institute was inaugurated in Boston in 1839 by *John Paul Brown* [10]. Other important institutes were founded by *Wilhelm Schulthess* in Zurich, Switzerland (*Wilhelm Schulthess Klinik* and *Balgrist Hospital*) [6] and the *Rizzoli Institute* in Bologna, named for the orthopaedist *Francesco Rizzoli* and opened in 1896 [6].

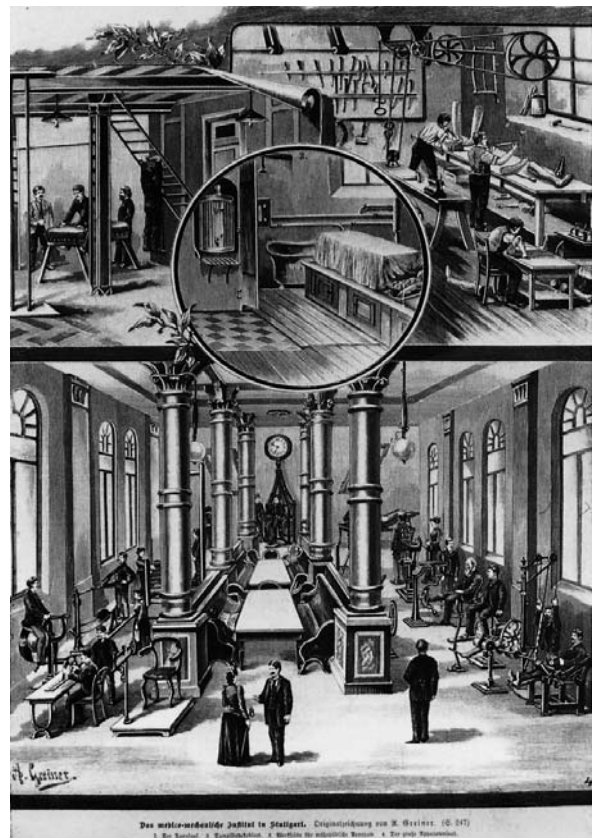
*Physical therapy* is another form of treatment that was already known to the ancient world. *Hippocrates* was aware of this mechanical therapy, while *Aesculapius* and *Galen* recommended massages. Gymnastic exercises were also a familiar feature. Hydrotherapy and balneotherapy arrived from the Orient and were known to the ancient Greeks and Romans. In Central Europe, bath houses and bathing masters are even mentioned in legislative texts (*Volksrechten*) dating back to the 6th–8th centuries. The bathing masters, who also worked as barbers, subsequently adopted the role of surgeons [4]. The steam bath, evidence for which dates back to the 13th century, has reappeared in the 20th century in the familiar guise of the sauna. Electrotherapy was introduced in the 18th century with the discovery of electricity. Galvanic treatment in water in a »Stanger bath« was very widespread. Actual therapeutic exercises were developed by *Fufelan* (1763–1836).

The German doctor *Daniel Gottlob Moritz Schreiber* refined these to produce a system of »medical gymnastics«. He also invented the allotment garden, which is known as a *Schreibergarten* in German-speaking countries. *Friedrich Ludwig Jahn* was the founder of an actual gymnastics movement with a patriotic outlook (*Die deutsche Turnkunst*, 1816). *Jahn* is considered to be the German »father of gymnastics«. *Pehr Henrik Ling* subsequently founded the »Swedish physical therapy« program, a dynamic method that competed with the mechanical techniques of the time. *Jonas Gustav Zander*, on the other hand, developed various apparatuses for use in therapeutic exercises. Numerous institutes employing *Zander's* machines were founded towards the end of the 19th century [6] (■ Fig. 1.6). This form of treatment was also known as »mechano-therapy«.

Physical therapy in the current meaning of the term was developed towards the end of the 19th century with the support of the clinicians *Theodor Billroth* and *Albert Hoffa*. The pioneers also included *Rudolf Klapp*, who developed a creeping treatment. Numerous physical therapy schools were formed in German-speaking countries. New therapeutic options for neuromuscular disorders were introduced in the 1950's by *H. Kabat* and *B. Bobath*. Hippotherapy for disabled children was also developed around this time.



■ Fig. 1.5. »L'invalidé«: Anonymous depiction of a below-knee amputee with peg leg, Paris, 18th century



■ Fig. 1.6. Medico-mechanical Zander institute in Stuttgart. Advertising copy published at the end of the 19th century. A certain similarity with today's fitness centers is unmistakable...



### Surgical treatment

Since classical antiquity, surgical treatment in orthopaedics was limited for many centuries to a single procedure, namely *amputation*. This mutilating operation was necessitated by the numerous war injuries. Even in the Middle Ages people realized that wound fever would lead to death if the injured limb was not amputated in time. This development reached its zenith with *Dominique Larrey* (1766–1842) who, as Napoleon's chief surgeon, was able to perform amputations in less than a minute [5]. Given the lack of effective anesthetic techniques, speed was an important requirement in performing the procedure. Apart from amputations, the only other orthopaedic procedures commonly undertaken at that time were *tenotomies*, which were performed particularly for congenital muscular torticollis (using what were known as »neck-cutters«) and for clubfoot or equinus deformity.

The two important preconditions for the development of surgical orthopaedics were only satisfied in the mid-19th century: *Anesthesia* and *asepsis*. The pioneers in anesthesia were the Boston dentist *William Thomas Morton* who, in 1846, was the first to administer an ether anesthetic, and the doctor *James Young Simpson* in Edinburgh who, in 1847, used chloroform in obstetrics. An important forerunner in employing asepsis was *Ignaz Philipp Semmelweis* (1818–1865) in Vienna, Austria, while *Lord Joseph Lister* (1827–1912) introduced antisepsis. As a result of these developments, complex operations soon became a possibility.

While isolated attempts at *osteotomy* date back to the era before anesthesia, this operation only gained acceptance in the second half of the 19th century. Pioneers in this field were *Bernhard Langenbeck* and *Theodor Billroth*, the latter introducing the use of the chisel [9]. By the end of the 19th century, *bloody reduction* was commonly employed for hip dysplasia, and *arthrodeses* were also possible. Methods for *lengthening muscles and tendons* for the treatment of the consequences of poliomyelitis were developed at the start of the 20th century. The trailblazers were *Oskar Vulpius* and *Richard Scherb*.

*Arthroplasty* first emerged at the start of the 20th century in the form of joint transplants. While experiments with artificial joints were conducted as early as the 1940's, the breakthrough was only achieved at the start of the 1960's with the development of the hip prosthesis by *John Charnley*. In pediatric orthopaedics, arthroplasties are only relevant in the context of malignant bone tumors and possibly rheumatoid surgery.

*Spinal surgery* dates back to the 1920's. Spinal fusion was introduced by *R.A. Hibbs* and fixed postoperatively in a plaster cast. At the end of the 1950's *Paul Harrington* in Houston developed instrumentation for the posterior realignment and fixation of the spinal column. At the start of the 1960's, *A.F. Dwyer* proposed an anterior approach to achieve the same objective. Spinal surgery experienced

a boom in the 1980's with the advent of numerous refinements in this field.

Another milestone in the development of modern orthopaedics was the introduction of *arthroscopy*. The beginnings of this technique date back to *Eugen Bircher* (in Aarau, Switzerland) in the 1920's. A school for arthroscopy was formed in Japan by *Keniji Takagi* in the 1930's. The current technique was primarily developed in the 1950's, likewise in Japan, by *Masaki Watanabe*, and led to a boom in knee surgery during the 1970's and 1980's. Knee ligament reconstruction, in particular, flourished during this period. Although rarely performed on children, this procedure certainly is of relevance for adolescents.

*Surgical leg lengthening* dates back to *Alessandro Codivilla* in the Rizzoli Institute in Bologna, who performed an osteotomy and extended the leg by traction with weights. During the 1950's, *Gavril Ilizarov* in Russia developed the ring fixator for which he is named. Surgeons in Europe and America remained unaware of this development for a long time and instead used the apparatus introduced by *Heinz Wagner* in the 1960's. It was not until the 1980's that the Wagner lengthening method was abandoned in favor of the Ilizarov technique. Many other unilateral devices and ring fixators have since been developed.

Pioneers in *surgical fracture treatment* at the start of the 20th century were *A. Lambotte* in France and *R. Danis* in Belgium. At the start of the 1960's the Association for the Study of Internal Fixation (AO/ASIF) in Switzerland provided considerable impetus in this field. The stable techniques of internal fixation developed by the AO/ASIF also play an important role in pediatric orthopaedics (primarily in connection with osteotomies), although even today the plaster cast and certain »unstable« internal fixation methods are much more prevalent in the fracture treatment of children than stable osteosynthesis procedures, which are only indicated in exceptional cases.

The surgical treatment of malignant *bone tumors* predominantly involved amputation up until the end of the 1970's. It was only with the development of modern chemotherapy methods, suitable tumor prostheses and the use of solid homologous grafts that modern limb-preserving tumor surgery was able to progress in the 1980's and 90's.

### Principles – Theories – Biomechanics

Orthopaedic treatments cannot change a diseased organ into a healthy organ, they can merely steer the body's own healing powers in a positive direction. The basis for our ideas on therapeutic indications is *biomechanics*. The first fundamental insights were published by *Julius Wolff* (1836–1902) in his treatise entitled *Gesetz der Transformation der Knochen* [The law of the transformation of bone] (1892). Wolff discovered that bone adapts to stress, i.e. it is deposited and resorbed in response to increased and decreased loads respectively. The term »functional adaptation« originates from *Wilhelm Roux* (1850–1924). In

his publication *Der Schenkelhalsbruch, ein mechanisches Problem* [Femoral neck fracture, a mechanical problem] (1935), *Friedrich Pauwels* explained current biomechanical thinking using mathematical models of forces and lever arms. Such model-specific ideas are generally based on static considerations.

*Gait analysis* introduced a dynamic approach to processes in the locomotor system. The first scientific examination of the human gait was published in the monograph by *W. Braune* and *O. Fischer* entitled *Der Gang des Menschen* [The human gait] (1896–1903), in which the kinematics of a walking soldier was measured in minute detail. Since the 1960's gait laboratories have been established in various centers. Modern computer-aided electronic methods are now used to calculate forces and torques in the joints during the dynamic process of walking and produce conclusions for subsequent treatments. This is a very valuable technique especially for patients with neuromuscular disorders.

For *diagnostics* in orthopaedics, as in most other disciplines of medicine, the invention of the x-ray by *Wilhelm Conrad Röntgen* in 1895 was of crucial importance. This technique was supplemented at the start of the 1970's by the computer tomogram (an invention attributed to *Godfrey Hounsfield*) and, at the start of the 1980's, by magnetic resonance imaging (MRI). The basic principles of the MRI technique date back to the early 1950's. In 1952 *Felix Bloch* and *Edward Purcell* were awarded the Nobel Prize for their discovery of magnetic resonance spectroscopy. The medical application of ultrasound began in 1947 with *Douglas Howry*. In orthopaedics this technique only became significant at the start of the 1980's when it was used for evaluating soft tissue processes and investigating infant hips.

*I should like to thank my friend Professor Beat Rüttimann MD of the Institute and Museum for the History of Medicine in Zurich for critically reviewing this chapter and for providing the illustrations.*

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## 1.3 Changing patterns of pediatric orthopaedic diseases – Developments, trends

» *We cannot see the future, but we do see the past. That's strange, for we don't have eyes in the back of our heads.* «  
(*Eugene Ionesco*)

### Achievements of recent decades

Orthopaedics has grown considerably in importance as a discipline in recent decades. This is largely attributable to the application of endoprosthetics in the treatment of arthroses. But internal fixation techniques for fractures, arthroscopy and ligament reconstruction for the knee and shoulder, and new instrumentation systems in spinal surgery have also contributed to the revival of this specialty. All of these achievements predominantly benefit adults, who are increasingly dogged by sports injuries and degenerative disorders. But significant developments with benefits for children and adolescents have also emerged:

- In the treatment of highly malignant *bone tumors*, survival rates and the simultaneous preservation of the affected extremity have increased over the past 20 years from 10–20% to 60–90%.
- New instrumentations have greatly improved the correction options for *scolioses* and *kyphoses*.
- New techniques have enabled better and more efficient *limb lengthening procedures* to be developed, thanks to innovative advances in Russia (G.A. Ilizarov). Ring fixators can now be used to correct contracted joint deformities. Even very complex abnormalities can now be corrected in three dimensions with the Taylor Spatial Frame.
- Thanks to rapid advances in the development of electronic devices, gait laboratories facilitate the complex analysis of an impaired gait, allowing conclusions to be drawn for subsequent treatment.
- In severe *neurogenic disorders* hip reconstruction can prevent secondary dislocation and stabilize the hip in a central position, thereby eliminating pain and improving patient care. Even cases of severe neurogenic and musculogenic scoliosis can now be straightened and stabilized efficiently, allowing the patient to retain the ability to sit up.
- Microsurgery has opened up new avenues in the treatment of *congenital malformations*. Thus, for example, the pollicization of a finger in hand deformities restores the pinch grip, producing a substantial functional benefit.
- In *severe spinal deformities* the use of the titanium rib can not only straighten the crooked vertebral column without stiffness, it also efficiently solves the problem of the much reduced chest and lung volume.

- The *ultrasound examination* of infant hips has significantly improved the early detection of hip dysplasia and dislocation, resulting in a substantial reduction in the costs of treating this condition and its late sequelae.
- Knowledge of the *problem of impingement in the hip* has contributed much to our understanding of the development of arthrosis and opened up the possibility of preventive treatment even in adolescence.

But it is not just the introduction of new techniques that has led to advances in pediatric orthopaedics. Thanks to recent findings *many surgical treatments that used to be considered essential are hardly used at all these days* (for example, the procedure of trochanteric derotation osteotomy for an anteverted hip, the resection of harmless benign tumors, e.g. non-ossifying fibroma of bone, or of popliteal cysts, operations for treating a peripatellar pain syndrome, etc.). But many conservative treatments have also proved to be unnecessary (for example, the insertion of insoles for the treatment of flat feet, splints for metatarsus adductus or knock-knees, etc.).

### Unsolved problems

Various classical pediatric orthopaedic problems can now be considered as largely solved. Thanks to the early detection of hip dysplasia by sonography, the risk of late sequelae is almost non-existent. Clubfeet can subsequently be made to work properly in the majority of cases, even allowing the patient to participate in sport with no functional restrictions.

However, there are a number of problems that remain unsolved or conditions for which current treatments are still unsatisfactory:

- Adolescents undergoing correction of scoliosis or some other spinal deformity still have to cope with the complication of stiffness of the affected section.
- In certain conditions, for example Legg-Calvé-Perthes disease, little can be done to influence the fateful course.
- The limb-preserving treatment of highly malignant bone tumors is not usually a real long-term solution for adolescents. Even with optimal management with a tumor prosthesis or allograft, major problems can be expected after 10–20 years.

### Development of morbidity

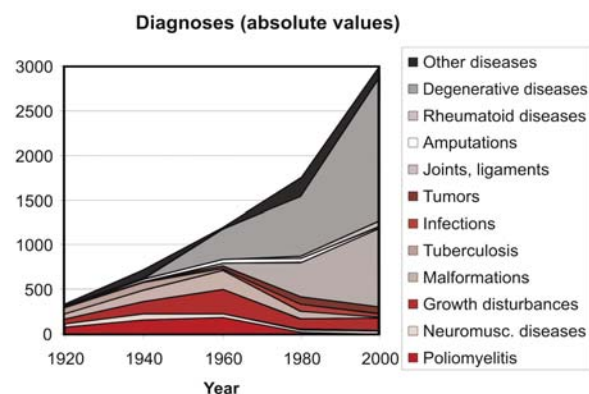
An analysis of population trend indicators suggests that the frequency of pediatric orthopaedic conditions is on the decline. In Central Europe, the birthrate in most countries is 1.4–1.6 (children per woman). To keep the total population at the same level (without immigration), the birthrate would need to be approx. 2.1. This figure is not currently reached anywhere in Europe. In Southern Europe the birthrate is even lower, at 1.15 (Italy), while

it is somewhat higher, at 1.8 (Ireland, Finland), in certain Northern European countries. This trend is exacerbated by the so-called »secondary pill dip«, i.e. the fact that, as a result of the drop in the birthrate in the 1960's, there is now a shortage of potential mothers for bringing children into the world. But not only will the number of children decline. The incidence of certain diseases is definitely falling, even though this trend is not yet clearly apparent in epidemiological studies.

To enable more substantial statements to be made about the occurrence of pediatric orthopaedic illnesses over time, I consulted the annual reports of the two oldest orthopaedic institutions in Switzerland, the Orthopaedic University Hospital of Balgrist and the Orthopaedic Hospital in Lausanne, dating back to 1920 in intervals of 20 years. To these can be added the records of the Orthopaedic University Hospital of Basel dating back to 1960. Between 1920 and 1960, all diagnoses for patients treated in hospital increased (apart from TB), primarily because of the general improvement in the options for hospital treatment.

A more differentiated picture appears from 1960. A substantial increase in degenerative diseases (particularly the arthroses) and sports injuries can be contrasted with reductions in most categories relating to pediatric orthopaedics. ■ Fig. 1.7 shows the frequency of these diagnoses since 1960. Marked reductions are observed not just for polio and TB (which has played a negligible role since 1960), but also for hip dysplasia, slipped capital femoral epiphysis and clubfoot, while Legg-Calvé-Perthes disease has remained fairly constant, inpatient scoliosis is on the increase again following a decline between 1960 and 1980, and the overall number of treatments of infantile cerebral palsy has increased.

The disappearance of hip dysplasia requiring inpatient treatment is (in central Europe) generally attributed to the



■ Fig. 1.7. Patients admitted to the Hôpital orthopédique in Lausanne, the Balgrist hospital in Zurich and the Orthopaedic University Hospital of Basel in the 20th century, listed according to *disease groups*. The groups that are relevant to pediatric orthopaedics are shown in shades of red

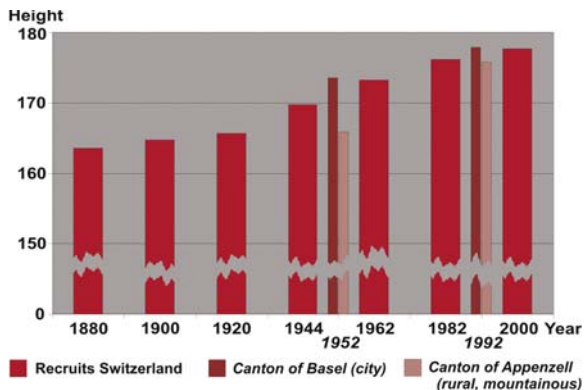
ultrasound examination. But this explanation fails to tell the whole story, since the marked reduction in treatments occurred between 1960 and 1980, i.e. *before* the development of ultrasound diagnosis. Another striking finding is the reduction in slipped capital femoral epiphysis despite an increase in the risk factors; there are now more overweight adolescents and those who overstress their hips with sporting activities than before.

Furthermore, the incidence of Legg-Calvé-Perthes disease is probably declining, although hospital (surgical) treatments are now indicated more frequently for this condition. The same applies to scoliosis. Inpatient treatments declined substantially between 1960 and 1980, but we have seen an increase in the number of operations in recent years probably because compliance with the brace treatment has deteriorated. We find a genuine increase for infantile cerebral palsy, particularly for the severe forms. The (rather milder) cases due to a traumatic birth are now rare, but the serious cases are definitely more common. Advances in neonatology have often preserved life in cases where the infant would previously have died of its cerebral injuries.

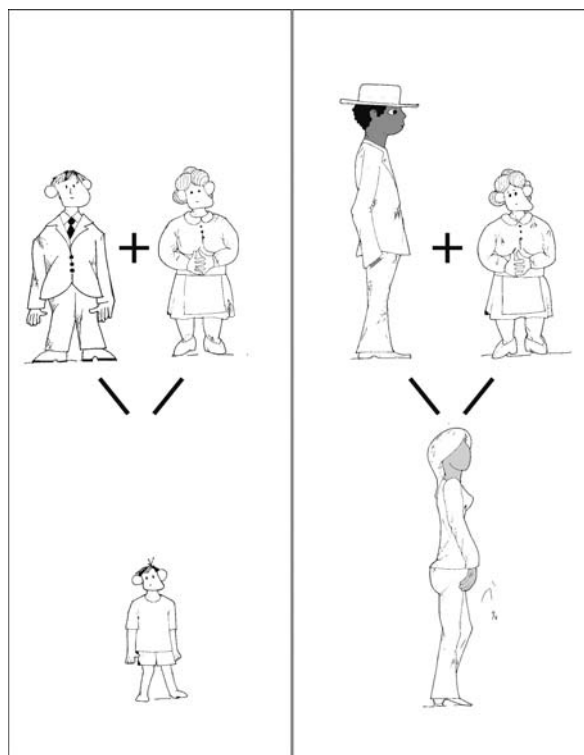
In order to isolate the causes of the reduction in most pediatric orthopaedic diseases we have examined another growth phenomenon, namely »acceleration«, i.e. the fact that children, on average, grow taller than their parents. I have obtained figures from the Swiss Army relating to the average height of conscripts recruited since 1880. Between 1880 and 2000 the average height of the Swiss recruit has increased by 15 cm (6 in.), from 163 cm (5 ft. 4 in.) to 178 cm (5 ft. 10 in.). (■ Fig. 1.8).

Swiss cantonal statistics are also available for the years 1952 and 1992. If we compare the typically rural-mountainous canton of Appenzell with the urban canton of Basel-City, the Appenzellers in 1952 were 7 cm (2.75 in.) shorter than their Basel counterparts (166 versus 173 cm or 5 ft. 5 in. versus 5 ft. 8 in.). In 1992 the Appenzellers were still shorter, but in this case only by 2 cm (176 versus 178 cm = 5 ft. 9 in. versus 5 ft. 10 in.). Diet is the usual explanation for the phenomenon of acceleration. But surely no-one could claim that the Appenzellers consumed substantially greater quantities of proteins than the Basel residents between 1952 and 1992. The only valid explanation is genetic intermixing. The Appenzeller population have known this for a long time as this is mentioned in the familiar local joke: »*The short people result from inbreeding and the tall people from tourism...*«. It is certainly true that most cases of hereditary skeletal dysplasia are associated with stunted growth, the sole exception being Marfan syndrome.

I think it is likely that the decline in pediatric orthopaedic diseases since 1960 also has something to do with mobility and the associated increase in genetic intermixing. After all, hip dysplasia had been common primarily in the Alpine countries of Austria, Switzerland,



■ Fig. 1.8. Average height of recruits in Switzerland from 1880–2000. The figures for specific cantons are shown for the years 1952 and 1992. In 1952 the average height in the rural and mountainous canton of Appenzell was 7 cm (2.75 in.) shorter than in the urban canton of Basel-City. By 1992 the difference had fallen to just 2 cm (1 in.)



Increased genetic intermixing has not only resulted in an increase in the average height, but is also responsible for a decline in the frequency of congenital malformations...

Czechoslovakia and Southern Germany, whereas in Italy only the mountainous region of Lombardy was affected. Inbreeding in these regions was evidently greater than in those countries with coastal borders. The incidence of hip dysplasia in English-speaking countries back in the 1960's was about as low as the current figure for the Alpine countries.



### Future

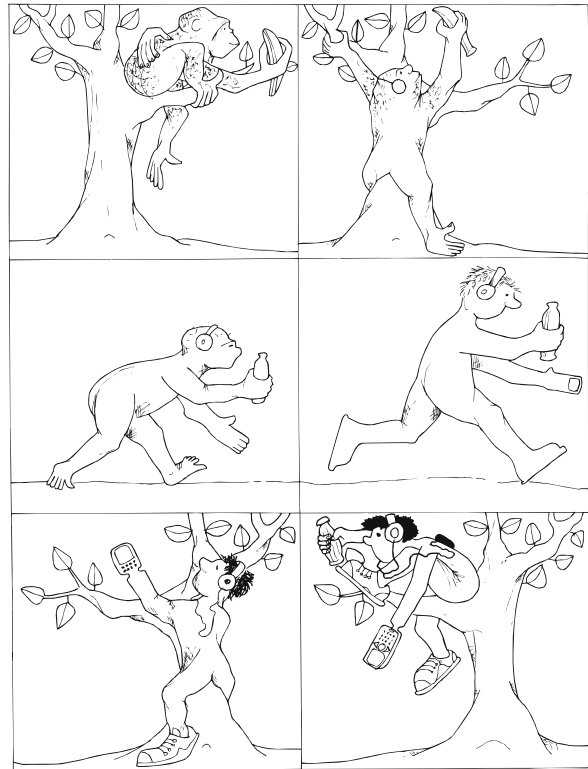
» *Those who didn't fulfill a childhood dream are not the ones who are poor, but rather those who didn't dream at all during their youth.* «  
(Adolf Nowaczynski)

For the reasons outlined above, we shall be seeing fewer pediatric orthopaedic problems in our offices in future. On the other hand, there will be an increasing need for a small number of individual pediatric orthopaedic treatment centers where the latest treatments are provided and where children and adolescents can receive appropriate, age-specific care. Such centers *must* be located in a children's hospital, where all the specialists in neighboring disciplines are available (pediatric anesthetist, pediatric neurologist, oncologist, geneticist, pediatric surgeon, etc.). Such centers will also need pediatric orthopaedists with subspecialties who are qualified particularly in disciplines such as neuro-orthopaedics, pediatric traumatology, tumor, spinal or hand surgery and microsurgery. While several such centers already exist in English-speaking countries, and we have also implemented this concept in Basel, it is still not very widespread in Central Europe. The future will bring further improvements in pediatric orthopaedics. Prenatal ultrasound diagnosis will provide a further reduction in malformations. Even now we are technically capable of detecting an abnormality as small as syndactyly, for example, in the 15th week of pregnancy. But there are still too few investigators with adequate knowledge of the whole spectrum of possible malformations, which means that many children are still born with deformities that remained undetected.

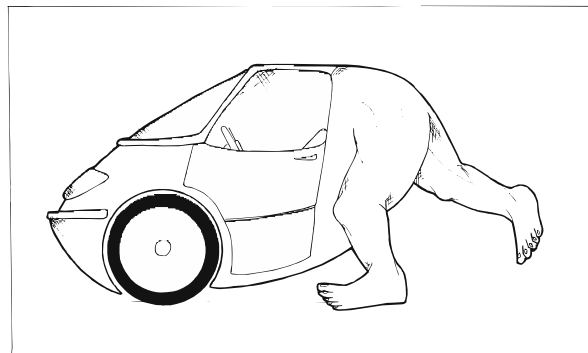
We can also dream of other conceivable advances. Perhaps one day we shall be able to straighten scolioses without stiffness, resect sarcomas more precisely thanks to tumor markers (possibly in a computer-navigated, or even completely computer-controlled, procedure), offer stable, long-term bridging options and, thanks to gene technology substitute missing enzymes in hereditary disorders...

» *We should not only dream during youth, but also for youth...* «

But pediatric orthopaedics will also have to face other challenges. The growing *pressure on costs* in all countries is increasingly prompting political authorities and health insurers to ask what a treatment actually provides and what price should be paid for that treatment. It is no longer sufficient to demonstrate that a lesion can be successfully repaired by a treatment. It is rather a question of demonstrating that a treatment can not only correct the impairment, but can also positively influence subsequent individual disabilities or handicaps in society. The criteria for conducting evaluations at this level are listed in the ICDH (=>International Classification of Impairments, Disabilities and Handicaps).



Evolution will continue to perfect humans and their functions...



Genetic intermixing probably holds a few surprises in store for the future...

The goal of treatment must be to produce a benefit in terms of abilities, or at least the maintenance of functions that would be lost without treatment. If it cannot be demonstrated that a treatment will achieve these objectives, then the health insurers will probably be unwilling to pay for such treatments in future, or at least only willing to pay a part of the cost. Demonstrating the maintenance of abilities will prove extremely difficult in pediatric orthopaedics.

As a rule, our work is not based on a time scale of weeks, months or years, since a positive effect may only



1 emerge after decades. No-one now implements treatments that were common 20 or 30 years ago according to the approach that prevailed at that time. Although we should undoubtedly focus our attempts primarily on improving or maintaining abilities, cosmetic aspects should not be completely disregarded (for instance in cases of thoracic scoliosis). But how can one statistically prove the maintenance of abilities in pediatric orthopaedics, given the small patient numbers involved and the considerable variation in therapeutic methods?

Even though large numbers of patients undergo hip ultrasound examination, there is still no agreement at all as to whether this diagnostic method reduces costs or not. Specialists in preventive medicine (in Switzerland) consider that the statistical data is still inadequate, even though most pediatricians and orthopaedists are convinced that the incidence of dysplasia-induced dislocation and the costs of treatment have declined substantially since, and because of, the introduction of this method (although there is no doubt that the natural occurrence of the condition has also declined as a result of the greater genetic intermixing of the population). It will be even much more difficult to demonstrate statistically the efficacy of rarely performed surgical treatments.

## Basic principles

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## 2.1 Diagnosis

### 2.1.1 General examination technique

#### 2.1.1.1 Medical history

The specific aspects of dealing with children and adolescents were explained in detail in ► chapter 1.1. This chapter will focus solely on the *systematic aspects of the interview*.

#### Current problem

##### Has a trauma occurred?

If so:

- When did the trauma happen?
- During what type of activity (sport, play, daily routine)?
- Direct or indirect trauma?
- With what movement was the trauma associated?

#### Pain history

- Where is the pain located?
- When did it occur?
- Is the pain *load-related*, *movement-related*, or does it also occur *at rest* or even at night?
- *Load-related* pain is usually caused by a problem in the joints, but can also be induced by muscular or intraosseous problems.
- *Movement-related* pain: What specific movements elicit the pain? Movement-related pain without load-related pain indicates the presence of muscle problems.
- *Nocturnal pain*: Does the pain only occur when the patient changes position or does the patient awake at night because of the pain?

#### ! One-sided pain that is not clearly load-related always raises suspicions of a tumor or inflammation.

- Asking about the *type of pain* (stabbing, dull, burning, etc.) is not usually very productive with children and adolescents.
- On the other hand, the *duration of the pain* can often be established with great accuracy with precise questioning.

#### Other events

- Habitual or voluntary dislocation of joints,
- Cracking or rubbing sounds,
- Clicking in the joints.

#### Personal history

**Course of pregnancy and labor history:** Special events during the pregnancy, head or breech presentation at delivery, cesarean section, difficulties during labor are significant factors in many conditions that are of relevance

to orthopaedics. A persistent intoeing gait or excessively frequent traumas during sport may be attributable to a very slight cerebral motor dysfunction. Labor complications can not only be the cause of problems but also the result of cerebral injury to the fetus.

**Early childhood development, onset of walking:** *The onset of walking is a simple and effective parameter for evaluating motor development.* Almost all mothers can remember this point, even many years later. The onset of walking normally occurs between 10 and 18 months. Impaired motor development should be suspected in children with a delayed onset of walking.

**Previous illnesses and accidents:** List all previous illnesses, accidents and operations in chronological order.

**Pubertal development:** *In girls, the menarche is an extremely accurate and useful parameter for evaluating the development status. After the menarche the pubertal growth spurt continues for a further 2 years or so.* Girls, and their mothers, can almost always date the onset of the menarche to the exact month. No similarly effective parameter exists for boys. Breaking of the voice occurs gradually over a prolonged period. Adolescents and their parents are usually unable to state exactly when it occurred. They just happen to notice one day that it has taken place.

**Sport:** Adolescents should always be asked about sports activities in and out of school. Before prescribing an expensive (and futile) course of physical therapy for a slack posture, you should explore the options for practicing a sport that the young patient might also enjoy.

#### Family history

**Hereditary disorders in the family:** Asking about hereditary illnesses in the family is not usually very productive. On the one hand, the parents rarely know what illnesses are inherited, on the other, they tend to keep quiet about outwardly visible hereditary conditions as these can elicit feelings of shame. Only the question about *hip conditions* (hip dysplasia, osteoarthritis of the hip) is usually answered correctly. *Tumors* in the family are also reliably itemized in response to the appropriate question.

**Family relationships:** Questions about social relationships should not be posed systematically as this can prove very hurtful. It is often possible, however, to incorporate relevant targeted questions in the interview. While the number of *siblings* can usually be elicited without difficulty, establishing the precise number of *parents* who are physically present in everyday life can prove more problematic. The fact that a father is attending the consultation by no

### 2.1.1 · General examination technique

means indicates that he is also present at home. Another »father«, of whose existence you are completely unaware, may be responsible for the patient at home, and his differing opinion (from that of the other parents) may greatly influence the therapeutic decision. As the treating doctor you are then astonished when the parents, who had been in complete agreement with your proposals during the consultation, subsequently decide on the opposite course of action. Other factors in the *social environment* (mother's or father's job, unemployed father, financial situation, relationships at school, drug scene, etc.) can also influence the course of an illness, frequently to a considerable extent. Questions about such topics must be posed with considerable sensitivity and tact.

#### 2.1.1.2 Instruments, measuring instruments

Just as the internist is identified by the stethoscope hanging from the neck, so the orthopaedist is characterized by the *protractor* poking out of the pocket.

Other *important* utensils for the pediatric orthopaedic consultation are

- the ruler on the wall for height measurement,
- the tape measure (non-extendable tailor's tape measure for circumference measurements),
- Boards for indirect leg length measurement, in various thicknesses (5 mm, 1 cm, 1.5 cm, 2 cm, 3 cm, 4 cm, 5 cm),
- reflex hammer,
- stool (for examination of the back),
- a flat examination table.

The following are also *useful*

- weighing scales,
- podoscope,
- safety pin,
- scoliometer
- flashlight,
- box or children's chair on which toddlers can stand so that their back is at eye-level during the examination,
- camera for documenting outwardly visible deformities.

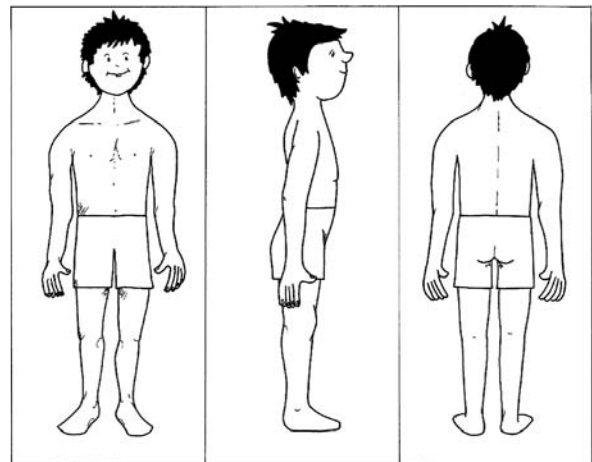
#### 2.1.1.3 Measuring the range of motion by the neutral-0 method

» If you need to measure joint motion, think of the neutral-zero notion. «

In joint measurement according to the neutral-0 method, all the movements of a joint are measured from a uniformly defined neutral or zero position. The measured angle gives the range of deflection from the zero position [1]. The zero position relates to the *anatomical zero position* or *baseline position for joint measurements*. This position has been defined as standing erect with the arms hanging by the side, thumbs pointing forward, legs extended and feet together and parallel. The patient's gaze is directed forwards (■ Fig. 2.1).

The range of motion is *recorded* according to the *zero-crossing method*. The mobility of each joint in each direction is noted in 3 sections: The extreme positions are noted on the left and right, and the zero-crossing point in the middle. If this cannot be achieved because of a contracture, the angular position of the contracture is specified as the middle figure, and a 0 is entered on the side of the extreme position. Examples are provided in

■ Table 2.1.



■ Fig. 2.1. Standard anatomical position for the measurement of joint motion by the neutral-zero method. All joints are shown in the zero position. (After [1])

■ Table 2.1. Possible options for recording joint mobility according to the zero-crossing method

Joint	Direction of movement	Angle [°]
Normal hip mobility in the sagittal plane	Flexion/extension	130–0–10
Flexion contracture of 30°	Flexion/extension	130–30–0
Normal rotational movements of the hip	External/internal rotation	70–0–60
Normal knee mobility in the sagittal plane	Flexion/extension	160–0–0
Hyperextensibility of the knees	Flexion/extension	160–0–10

The neutral-zero method allows joint mobility to be recorded precisely and unambiguously at any time. This method also offers an extremely succinct way of recording the range of motion since there is no need for a detailed description of a pathological condition. For example, rather than writing: »The hip flexion is 130°, but full extension cannot be achieved because of a flexion contracture of 30°«, we write: »Hip flexion/extension: 130–30–0«. The joint ranges of motion in this book are all stated according to the neutral-zero method.

#### 2.1.1.4 Orthopaedic examination technique

##### Inspection

**Contours:** We record *swellings, redness, protuberances* or any *atrophy of the muscles*.

**Deformities:** We note any *abnormal curvature* of the extremities or the spine, *axial deviations, high or low positioning* of joints.

**Skin:** We observe *color, pigmentations* (e.g. café-au-lait spots), *callusing*.

**Anomalies:** We record any outwardly visible anomalies.

##### Palpation

We can palpate the following parameters

**Pain points:** In every region of the body there are various pain points at which local tenderness can typically be elicited in the presence of certain lesions. These will be described under the specific body region. *Children will appreciate it if you avoid provoking pain any longer than necessary.*

**Skin:** *Skin temperature, sweating and mobility* of the skin (particularly important for tumors).

**Swelling, effusion:** *Swellings* are accumulations of fluid outside the joints, while intra-articular fluid is termed an *effusion*. A skilful examiner can clinically distinguish between the two.

**Protuberances:** These can be *hard, firmly elastic* or *soft* depending on whether solid tumors, cysts, soft tumors or other soft tissue protuberances are involved.

**Crepitations:** *Crepitations* are common in the knee (particularly behind the patella), but are not necessarily an indication of arthrosis. Crepitations rarely occur in other adolescent joints and are usually induced by wear phenomena.

**Ligament stability:** The condition of the *ligaments* can be examined specifically in each joint (see relevant joint). Note that children generally tend to show much greater ligament laxity than adults. Both sides should always be compared during this examination.

##### Pediatric orthopaedic status

Height and possibly weight should be measured at every examination, since it constitutes the simplest parameter for growth and one that influences the course of almost all orthopaedic conditions while the child is growing.

Girls who have reached the age of puberty must be asked about the onset of the *menarche*.

Insofar as possible a *systematic examination procedure* should be adopted: e.g. examine the patient while walking, standing, lying, organized according to topographic considerations: spine, hips, knee, feet, upper extremities

- *height, weight,*
- *walking with/without shoes, heel and toe walking,*
- *static position* (pelvic tilt, leg and feet axes, Trendelenburg sign),
- *spine* (shoulder position, pelvic tilt, waist triangles, sagittal profile, frontal profile, forward-bending, sitting up, lateral mobility),
- *upper extremities* (cursory examination of mobility, apron grip [i.e. adduction, retroversion and internal rotation, as if tying an apron at the back], neck grip, turning up the thumbs),
- *hips* (mobility),
- *knees* (mobility, stability, meniscus signs)
- *feet* (rearfoot, forefoot, arches, mobility),
- *torsions*, clinically (anteversion, tibial torsion, feet axes),
- examination of *capsular ligament laxity* (hyperextensibility of the knuckle joints beyond 90°, the elbow joints by more than 10°, and the knees by more than 5°, thumb-forearm distance of less than 2 cm),
- examination of *muscle contractions* (contraction of the hamstrings, triceps muscles, rectus femoris, iliopsoas),
- *neurological examination* (motor function, tendon reflexes, sensory function).

Growth, signs of maturation and physical development are described in ► chapter 2.2.2.

A full orthopaedic assessment can be waived for children who attend consultations more frequently than every six months. In such cases, the examination can be confined to the current problem.

**! Regardless of the problem, a pediatric orthopaedic mini-assessment should be conducted at least once a year (measurement of height, evaluation of leg lengths, examination of the spine for rib and lumbar prominences and hyperkyphosis).**

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## 2.1.2 Neurological examination

R. Brunner

### Objectives

In orthopaedics, the neurological examination has two objectives

1. To identify and document lesions of the nervous system (e.g. in injuries). The subsequent progress can then be assessed through comparative follow-up examinations.
2. To record the extent of a neurological disease and its effects on the musculoskeletal system (for example, plexus paresis, cerebral palsy).

### Examination procedure

A comprehensive neurological examination of a child is very extensive and time-consuming, and many details are of secondary importance for an orthopaedic assessment. Consequently, the type and extent of the various neurological examination options must be guided by the prevailing *orthopaedic problem*.

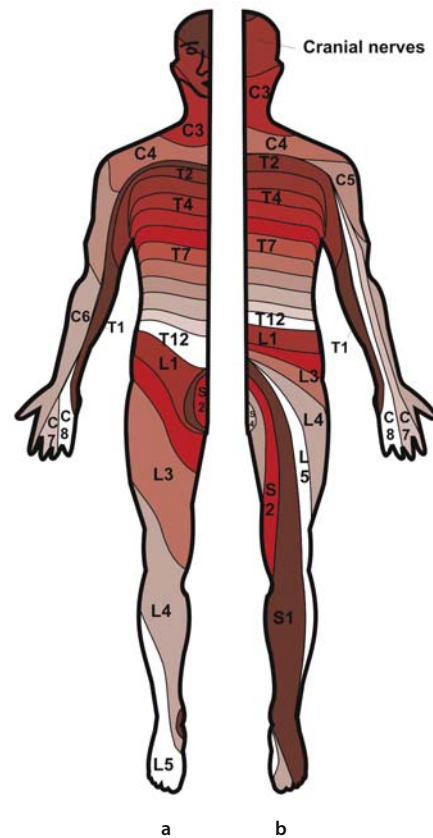
*Sensory deficits* or changes are identified by comparing sides or comparing with adjacent areas. The deficit areas of peripheral nerves or segments can be categorized (■ Fig. 2.1 and 2.2). With acute lesions, in particular, it can be useful for documentation purposes and subsequent comparison to mark and photograph the deficit areas on the skin.

*Motor strength* is assessed clinically and classified according to 5 categories (■ Table 2.2, Muscle status). The deficits, in turn, can be assigned to individual nerves or segments. The muscle reflexes are tested on both sides for comparison. In disorders of the central nervous system (CNS), bladder and bowel function are also investigated.

*Lesions of individual nerves* almost always affect sensory and motor function (flaccid pareses).

Lesions at the *spinal cord level* show segmentally structured deficits. The extent of the reduction in sensory function can vary, ranging from hypoesthesia to complete anesthesia. Motor deficits also follow a segmental pattern, with muscle activity being completely absent, diminished or even spastic.

Other *central deficits at the cerebral level* often result in a qualitative change in motor and sensory function. The motor symptoms manifest themselves in the form of spasticity, dystonia, muscle hypotonia or hypertonia, ataxia or



■ Fig. 2.2a, b. Sensory areas on the human body: a Anterior view, b Posterior view

■ Table 2.2. Evaluation of muscle strength

Grade	Muscle strength
5	Normal muscle strength (movement against heavy resistance of the investigator)
4	Movement against moderate resistance of the investigator
3	Movement against gravity
2	Movement with gravity eliminated
1	Signs of contraction with no movement in the joint
0	Complete paresis

athetosis. The qualitative change in sensory perception often appears in the form of hyperesthesia, paresthesia or hypoesthesia.

If spasticity is present, rapid movements during the clinical examination will elicit a spasm, causing the patient discomfort or even pain. This manipulation should be restricted to the neurological diagnosis of spasticity.



Since rapid movements hinder the orthopaedic examination of the range of motion, the latter should be conducted slowly, continuously and with patience. On the other hand, fast movements can help establish the extent of the spasticity. The degree of the joint movement at which the spasticity blocks another movement in the joint during this rapid investigation can be recorded as a measured parameter. Asking the patient to sit up from a supine position and to hop or stand on one leg is useful for investigating the tone, postural function and coordination of the muscles. The assessment of these parameters is also important for the analysis of the orthopaedic problem.

A lack of postural tone in the trunk and head area is expressed by poor control and flaccid caving in of the trunk.

### Symptoms

*Spasticity* refers to an exaggerated muscle reflex. In the typical case of *spinal spasticity* the higher-level inhibition of the intact reflex arc of the muscle reflex is lacking. Hyperexcitability of this reflex arc develops over time, causing even slight stretching of the muscles to elicit the muscle reflex and produce a tetanic contraction. A precondition for this outcome is an intact second motor neuron. This spasticity typically affects muscle groups that maintain posture against gravity (M. triceps surae, hamstrings).

On the one hand, spasticity can be triggered by sudden movements in a joint, on the other the muscle reflex is regularly exaggerated or clonic. The area over which the reflex can be elicited may be extended. The term spasticity is also used to describe a scenario of general, tenacious resistance, without cogwheel rigidity, as the joints are moved through their range of motion, the resistance being much weaker when the joints are moved very slowly. *Dystonia* involves a change in the muscle tone.

*Ataxia* occurs primarily during walking, and the patient's steps appear uncertain and of differing length. The feet are subject to constant stabilizing movements during the stance phase. As regards the differential diagnosis, the doctor should consider the possibility of *impaired vision* in a case of ataxia on its own, as this can lead to similar gait problems.

A typical *athetosis* can readily be identified by the spontaneous movements of the extremities and head or trunk. Frequently, however, only a single athetotic component is present. The athetotic movements may be slight and occur only occasionally, or may be completely absent while the patient is sitting calmly.

In the search for individual typical neurological signs and symptoms, the *coordinating functions* of the nervous system must also be checked. Clumsiness is often present, as are balance problems of varying se-

verity. A good overview of the orthopaedic situation can be obtained by asking the patient to hop or stand on one leg. Accompanying movements of the upper extremities may occur, for example during testing for diadochokinesia.

The *child's stage of development* is also important particularly in neurological disorders that result in severe disability. A rough indication can be obtained by asking when the child started walking unassisted (generally by the 18th month at the latest) or possibly sitting unassisted (approx. 11th month).

While it is often not possible to fully test for *sensory disturbances* in severely disabled or uncooperative patients, the history can provide clues to such disturbances: Thus, for example, a refusal to wear shoes or pulling the legs away from the floor may indicate problems such as dysesthesia or paresthesia.

### 2.1.3 Gait analysis

R. Brunner

In organizing our daily routine, most of us take the concept of locomotion for granted. But the function of walking is impaired in many orthopaedic conditions.

➤ **Walking is an extremely economical form of locomotion. The optimal use of external forces requires coordinated movements of limbs and trunk.**

#### The normal gait

The orthopaedist must be fully conversant with the normal gait before analyzing a patient's walk and identifying any pathologies. The various and, in some cases, complex movements during walking are designed to move the body's center of gravity forward as uniformly as possible and without deviating from the gait direction. At the same time, external forces (such as gravitation and mass movements) are controlled in such a way that the forward movement occurs with the minimum of expenditure. To this end, the leg must perform the chain of movements in an *orderly sequence*.

The *foot* moves largely in the sagittal plane in relation to the lower leg axis. Directly before the foot strikes the ground, the sole is perpendicular to the lower leg (plantigrade) or in very slight plantar flexion (approx. 5°), and the foot and toe levator muscles are active. After the heel strikes the ground, the foot is plantarflexed until the sole strikes the floor. This movement is termed the *»first rocker«*. Placing the sole of the foot on the floor is controlled by the foot levator muscles. The lower leg then moves forward over the foot that is now resting on the floor, resulting in a dorsal extension movement in the ankles, which is cushioned by the eccentric contraction of

### 2.1.3 · Gait analysis

the triceps surae muscle (»*second rocker*«). The full range of this movement is between 15° and 20°. At the end of this dorsal extension, the eccentric contraction of the triceps progresses to concentric contraction via an isometric phase. As a result, the heel is raised and the foot pushes the leg away from the ground (»*third rocker*«). After push-off, the foot once again returns to the plantigrade position preceding foot-strike by activation of the foot levator muscles [1, 2].

The *knee* flexes slightly after the foot strikes the ground, and the quadriceps muscle prevents the leg from buckling by eccentric activity. The deceleration of the lower leg's forward movement over the foot resting on the ground results in passive extension in the knee as the upper leg continues its forward progression. The ground reaction force, which can be presented as a vector between foot and ground, shifts from a position *behind* the knee to a position *in front of* the knee. While the force *behind* the knee can be subdivided into a component acting in the direction of the ground and a knee-flexing component (for which muscular compensation is needed), the ground reaction force occurring *in front of* the knee exerts an extending force on the knee. In other words, the knee is indirectly and passively extended primarily during the second part of the stance phase.

After push-off, the leg swings forward, after a short muscular acceleration, as a passive pendulum. The length of this passive pendulum and the weight of the leg determine the comfortable walking pace and step length, which differ slightly from one person to the next. During a step, and after being extended up to an approx. 5°–10° flexed position at the end of the swing phase, the knee initially undergoes slight additional flexion of approx. 15°–20°, followed by further extension up to around 5° or even 0° flexion. During the swing phase the knee is flexed by approx. 75°–85°, and the point of maximum flexion is reached when the knee passes the stance leg. The knee likewise moves only in the sagittal plane [1, 2].

The *hip joints* perform a flexion and an extension movement in the sagittal plane: During comfortable walking, the hip of an adolescent is flexed by approx. 30° as the foot strikes the floor. During the complete stance phase the joint is gradually extended up to an extension of 5°–10°. The hip then flexes again. Small children, in particular, are accustomed to a faster walking pace in relation to their height and therefore make relatively large strides. This increase in step length is expressed in increased flexion of the hip at footstrike.

In addition to flexion and extension, rotations at the hip are also required for the continuous forward movement of the body's center of gravity. The hip is rotated outwards concurrently with the flexion that occurs before footstrike. The pelvis then swings forward over the standing leg, resulting in an internal rotation. During the stance

phase, the hip joint is not only flexed but also externally rotated. At the same time, the leg is slightly adducted during the stance phase and slightly abducted during the swing phase. This abduction rotation and external rotation in the swing phase causes the whole leg to be slightly circumducted [1, 2].

#### Clinical gait analysis

➤ The clinical gait analysis can reveal gross pathologies in the movement sequence.

As preconditions for the analysis, the patient must be undressed down to the underpants and be capable of walking a sufficiently long distance (at least 3 meters). Ideally, the assessor should sit on a low stool so that the eyes can be kept roughly at the height of the patient's pelvis. Gait is assessed primarily from the front and back. Although it would be more productive to perform the examination from the side, this viewpoint is rarely possible for reasons of space.

The examiner notes the position of the feet while the patient is standing, the position of the knees during the swing and stance phases and the movements of the pelvis. A limp signifies a deviation from the normal movement sequence and usually occurs for compensation reasons, i.e. external forces generated via the displacement of the body's center of gravity are employed to replace muscle activity. A familiar type of limp is the exaggerated dropping of the pelvis on the side of the swing leg, known as a *Trendelenburg limp*, and the compensatory movement of the upper body towards the stance leg side, termed a *Duchenne limp*. Both are indicative of a functional deficit in the hip abductors on the stance leg side as they fail to provide adequate stabilization for the pelvis.

Forward leaning of the upper body relieves the load on the knee extensors. Active, full stretching of the knee after the foot strikes the ground is indicative of plantar flexor insufficiency. Other types of limp, e.g. an *antalgic limp* with a shortened stance phase for the painful leg, or a *shortening limp* with a highly asymmetrical movement of the pelvis, are also readily identified.

At the level of the feet, the examiner notes the position of the foot at footstrike (planta pedis or even equinus deformity) and the position of the heels in relation to the lower leg (varus or valgus deformity). The alignment of the foot in relation to the gait direction is another important factor. All of these points can be identified from an anterior or posterior viewpoint.

#### Instrumental gait analysis

Instrumental gait analysis is used to identify gait pathologies, particularly during faster movement phases, and record their functional impact.



A visually abnormal gait pattern does not necessarily always result in a functional impairment. However, this therapeutically relevant distinction is only possible with instrumental gait analysis which, even today, remains a highly elaborate method of investigation. Expensive and technically complex equipment measures various parameters, including forces, movements and electrical muscle stimulation (EMG), and it can take roughly a full working day to record and evaluate the data for a single patient. For these reasons, *instrumental gait analysis is reserved for complex problems.*

**Video analysis**

Video recordings allow the gait sequence to be viewed in slow motion.

Video analysis represents a simple form of gait analysis. In contrast with the clinical examination, the use of slow motion or single frames enables rapid movements to be analyzed or the position in several joints to be viewed simultaneously. The observing eye can be trained, and the significance of the assessment enhanced, if the gait pattern is viewed in a similarly consistent manner, hence the need for *standardized recording orientations*. Anterior, posterior and lateral views are suitable (■ Fig. 2.3). However, if the joints are not strictly aligned in these planes, the angles will be badly distorted and should therefore be evaluated with caution. Nor does the video analysis provide any information about the acting forces.

**Kinematics**

Kinematics reproduces the sequence of movements in the joints in 3 dimensions in relation to time.

An analytical system used in gait laboratories is needed to record the kinematic data. The system visually mea-

sures markers affixed to the patient's body, calculates the joint angles in 3 dimensions and represents these in relation to time. As a rule, the data is normalized to a single step for ease of comparison. The results are used to identify relationships between movements in movement sequences, for example in the lower limbs. The assessment is facilitated by graphics showing normal curves in the background.

**Kinetics**

Kinetics describes the torsional forces acting in the various joints and the physical work performed.

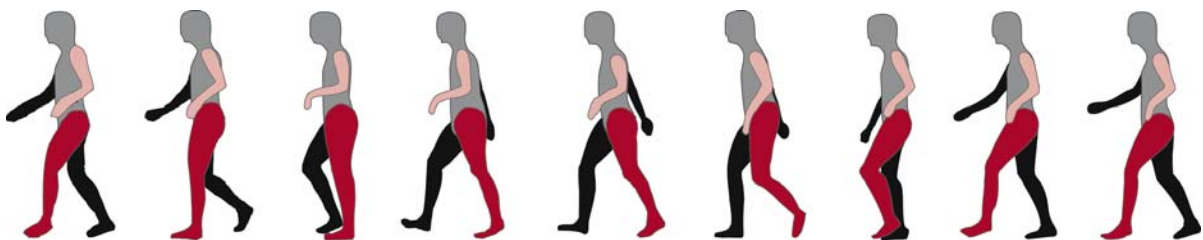
The forces act on segments, which rotate around centers of rotation. In physical terms they therefore constitute moments and can be calculated if both kinematic data and the ground reaction force are recorded simultaneously. These moments are either presented as externally acting forces or, on the basis of the principle »every action has an equal and opposite reaction«, as internal moments produced as a reaction. They provide information about muscle activity. In connection with the physical performance, kinetic analysis can also establish whether the muscles absorb or generate work, which is a crucial factor for the mechanics and efficiency of walking.

Force plate measurements are rarely used nowadays as raw data in gait analysis, since they are much more difficult to evaluate and the result is less reliable compared to the evaluation of kinetic data.

**EMG examination**

Dynamic EMG enables conclusions to be drawn about muscle control.

This examination can disclose co-contractions and muscle contractions with an incorrect onset, but does not



■ Fig. 2.3. Phases of a gait cycle (left and right steps) of a female patient with left-sided spastic hemiparesis, taken from a video recording: Only by analysis of the sequence in slow motion or single frames can the movement of the body as a whole be compared with that of the extremities. Also highlighted is the ground reaction force. If it pro-

ceeds on the flexion side from the hip or knee, the extensor muscles must actively counter any bending. If it occurs on the extension side, however, a passive extension moment comes into play and muscle force does not need to be expended

yield information about the exerted muscle force or the type of muscle contraction (eccentric or concentric). Recording this EMG data simultaneously with kinematic and kinetic data will, however, produce a comprehensive picture of the sequence of events during walking. The additional presentation of muscle lengths may provide further insights, and future software packages will include this tool.

#### Instrumental gait analysis

An EMG examination during walking is generally performed in particular before muscle transposition procedures, e.g. the transfer of the rectus femoris muscle, and in some cases before the transposition of foot muscles, in order to show the defective innervation and thus enable the surgical result to be predicted with greater accuracy.

Instrumental gait analysis can be used to demonstrate how gait pathologies lead to an increased expenditure of energy and to abnormal joint loading.

Instrumental gait analysis includes the simultaneous recording of kinematic, kinetic and EMG data. These examinations allow conclusions to be drawn about correction options and therapeutic goals. In complex functional problems gait analysis is a useful, and often necessary, basis for treatment planning. Only gait analysis can disclose whether excessive muscle force or muscular insufficiency prevails. The latter can also occur in spastic syndromes. However, the treatment will differ substantially, since a procedure to weaken the muscle will help in cases of incorrectly applied muscle force, while such a procedure would be contraindicated in cases of muscular insufficiency, for which the opposite measure of power training would be required.

#### Benefits of instrumental gait analysis

On the one hand, the benefits of gait analysis include the systematic recording of pathological gait patterns and the corresponding treatment options. Patient populations can then be investigated and evaluated in this context. On the other hand, gait analysis is also important for the individual patient, particularly since the time required for the evaluation has now been reduced to a few minutes. Without gait analysis, it would be almost impossible these days to draw up and monitor detailed treatment schedules with any reliability.

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## 2.1.4 Imaging diagnostics

Despite some spectacular advances in the field of imaging techniques, the *conventional x-ray* still forms the basis of imaging diagnostics in pediatric orthopaedics, since it is not only inexpensive, but also provides a very clear overview. What cannot be ignored, however, is the replacement of this method in certain areas of orthopaedic diagnostics, for example in the evaluation of the infant hip, where ultrasound has almost completely replaced conventional radiography. The order in which the imaging techniques are selected is crucially important in ensuring that a diagnosis is made as quickly and economically as possible. Since the clinician is usually responsible for making this decision, it is important that he should be aware of the possibilities offered by the various methods.

### 2.1.4.1 Imaging techniques Conventional x-ray

The general x-ray, or plain film, should always be recorded in two planes. X-rays of long bones should, if possible, include two adjacent joints. It is not useful, however, to x-ray the opposite, uninjured side for comparison purposes. Ignorance of the status of the epiphyseal plates does not justify an unnecessary exposure to radiation. It would be more appropriate in such cases to consult a radiology text book. Plain x-rays can be recorded using an analog or digital technique. Digital images offer the advantage of manipulability (e.g. by increasing or decreasing the contrast level). As a disadvantage for the orthopaedist, the proportions are more difficult to assess because digital images can be output in any arbitrary scale.

#### Stress x-rays

Stress x-rays can be useful in evaluating ligament tears. In this technique the joint is held in a (pathological) subluxed position. Although we hardly use this method any more for the ankle, such x-rays can be very effective in documenting the extent of the translation movement in ruptures of the anterior, and particularly the posterior, cruciate ligaments.

#### Functional x-rays

Functional x-rays document joint positions over their physiological range of motion. Such x-rays are recorded for the cervical spine (in reclination and inclination), the thoracic and lumbar spine in cases of scoliosis (in maximum lateral inclination) and the hip (in various rotational and adduction/abduction positions).

#### Sonography

The non-invasive imaging technique of sonography is based on the interaction between transmitted sound waves and the tissue interfaces in the body. When tar-

geted sound waves strike an interface between two tissue layers with differing acoustic impedance they are either reflected or refracted. High-frequency transducers operating at 7.5 MHz or 10 MHz offer good resolution and are ideally suited for use on the musculoskeletal system. In pediatric orthopaedics, sonography is primarily used for evaluating the infant hip (see ► chapter 3.2.4). It is also frequently used in swellings, protuberances and effusions in order to distinguish between solid tissue and fluid. Doppler sonography is used to investigate the circulation in an extremity.

### **Contrast-based imaging**

*Angiograms* are used to visualize vessels. The conventional angiogram is increasingly being superseded these days by the MR angiogram and by vascular imaging with spiral CT in the digital subtraction technique (DSA). *Myelography* is used to visualize intraspinal processes, particularly in cases of suspected mechanical impairment of the spinal cord. The images are either recorded by the conventional technique (rarely) or (increasingly nowadays) by computer tomograms (*myelo-CT*).

*Arthrograms* are images of the joint interior recorded after the intra-articular injection of contrast medium. Conventional arthrograms are rarely indicated nowadays, having been superseded by arthro-MRI, which is most commonly performed on the hip for evaluating lesions of the labrum, or on the knee for visualizing separation in osteochondrosis dissecans.

### **Scintigraphy and positron emission tomography (PET)**

Scintigraphy depicts the distribution of a radionuclide in the bone after intravascular injection. The main advantages of this technique arise firstly from the simultaneous imaging of the whole skeleton and, secondly, from the generation of a metabolic picture, i.e. metabolic activity is shown in comparison with that of adjacent normal bone. The technique is highly sensitive, but not very specific. Widely differing processes can produce a similar picture.

Scintigrams, or bone scans, offer a suitable screening method for tumors and infections. Technetium (Tc-99m-MDP) is the generally preferred radioisotope, and indium-111 is also used in some cases. In chronic infections, gallium-67-citrate is suitable for confirming therapeutic success or resistance. Bone scans can be evaluated by semi-quantitative analysis. If cortical bone appears to be thickened on a plain x-ray, the bone scan can, depending on the appearance in each case, indicate whether an osteoid osteoma (»hot spot«), osteomyelitis (diffuse, possibly irregular, uptake) or a stress fracture (localized, but not very »hot« uptake) is involved. For tumors, scintigraphy can be used on the one hand to assess the activity of the process and, on the other, to scan for metastases. The new

method of positron emission tomography (PET) is particularly sensitive in scanning for metastases (including in soft tissues).

### **Computer tomography(CT)**

A CT scanner consists of a gantry that incorporates a rotating x-ray tube and image sensors (detectors), the patient table and a computer for processing the data. The CT software converts the x-ray attenuation passing through the tissue into a CT density value (known as a Hounsfield unit, H.U.), by comparing it with the attenuation through water. The attenuation coefficient of water is 0 H.U., that of air -1000 H.U. and that of normal cortical bone +1000 H.U. The tissue types of bone, muscle, fat and water can very readily be distinguished from each other on the basis of their densities.

CT is an indispensable tool for visualizing bony structures, especially around joints and in the spine, e.g. in tumors, complicated fractures or deformities. The 3D reconstruction, in particular, facilitates the evaluation of structures in space (e.g. in complex spinal deformities, see ► chapter 3.1.7).

The newer *spiral CT technique* enables volumetric data to be acquired during continuous rotation of the x-ray tube and detectors. This technique substantially reduces the scanning times, enhances the resolution and facilitates the preparation of three-dimensional reconstructions. Used in conjunction with contrast administration, spiral CT can display the gastrointestinal tract or vessels in hitherto unmatched quality and three-dimensional vividness.

### **Single Photon Emission Computed Tomography (SPECT)**

This new technique has greatly improved the diagnostic accuracy in the evaluation of bone and joint abnormalities. SPECT offers enhanced contrast resolution compared to conventional tomograms.

### **Magnetic resonance imaging (MRI)**

Magnetic resonance imaging (MRI) is based on the re-emission of an absorbed high-frequency signal (RF signal) while the patient remains within a magnetic field generated by a magnet with a magnetic strength of 0.2–1.5 tesla (T). The system consists of the magnet, high-frequency coils (transmitter and receiver), gradient coils and a digital computer unit.

The ability to depict tissues is based on the intrinsic magnetic moment (spin) of atomic nuclei with an odd number of protons and neutrons (e.g. hydrogen) and a resulting net magnetic moment. The atomic nuclei then align themselves parallel to the magnetic field lines. The beamed pulses of high-frequency radio waves cause the atomic nuclei to absorb energy, inducing the nuclei to resonate and align themselves with the magnetic field.

When the high-frequency field is switched off, the emitted energy is recorded. The signal intensity corresponds to the strength of the high-frequency wave emitted by a tissue after its excitation [4].

There are two *relaxation times*:  $t_1$  and  $t_2$ . The spin echo (SE) produces a  $t_1$ -weighted image with a short repetition time (TR <500 ms) and short *echo time* (TE <40 ms), resulting in good high-resolution anatomical detail. The  $t_2$ -weighted image with a long TR (>1500 ms) and TE (>90 ms) produces high contrast. *Proton-weighted images* have a long repetition time and a short echo time (■ Table 2.3).

The IR sequences (»inversion recovery«) can be used to shorten the scanning times in multiplanar images. If a short inversion time (TI) in the range of 100–150 ms is used, the signal intensity of fatty tissues is suppressed. This is known as the STIR (»short tau inversion recovery«) technique. A similar result is also achieved with the CHES sequence (= chemical suppression). The main reason for suppressing the fatty tissues is to enable bone marrow edema to be evaluated (the fat of the bone marrow is suppressed thus producing a better view of the fluid). Recently, the fat suppression techniques have been combined with three-dimensional gradient echo imaging (GRE = gradient recalled echo), which provides an enhanced view of joint cartilage.

When the *contrast medium gadolinium* (Gd-DPTA) is administered intravenously it is taken up at sites of activity, e.g. at tumor borders, which can yield information suggesting whether a benign or malignant process is present. Tissue structures can also be seen more clearly after the intra-articular administration of gadolinium. The appearance of contrast medium flowing beneath a dissected flap in a case of osteochondrosis dissecans confirms the fact that separation has occurred. Arthro-MRI also facilitates better evaluation of a labrum lesion of the hip.

*Tissue characterization* is essentially achieved by the differing weighting of the MRI images (■ Table 2.4). Different weightings are obtained depending on the relationship between the two parameters of *repetition time* (TR) and *echo time* (TE). Comparing the images with differing weightings enables the *tissue type* to be evaluated on the basis of the signal intensity (or brightness) of the structures (■ Table 2.4). What is especially

important for practical purposes (particularly in tumor diagnosis, ► chapter 4.5.1) is the fact that cartilage-forming tumors can readily be distinguished from other neoplasms, that fluid accumulations are clearly identified and that nerves and tissues can be distinguished from other tissue types.

#### 2.1.4.2 Radiation protection

A crucial factor to consider in the application of diagnostic imaging is whether a method requires a high or low radiation dose compared to the available alternatives. As a matter of principle, therefore, ultrasound and MRI scans are preferred over conventional x-rays and CT scans if the diagnostic result is equivalent. This aspect of the smallest possible radiation dose has also resulted in monitoring investigations of thoracic scoliosis in which rastersterography (optical measurement of the back profile, ► chapter 3.1.3) has completely, or partially, replaced x-ray diagnosis.

All the basic rules of radiation protection apply in pediatric orthopaedic x-ray diagnosis, and each country has its own radiation protection regulations that form

■ Table 2.4. Tissue characterization based on the weighing on MRI images

Tissue type	$t_1$	Proton density (PD)	$t_2$
Fat	++	++	+
Bone marrow	++	++	+
Muscle	+–	+	+–
Hyaline cartilage	+	++	+
Fibrocartilage	–	–	–
Ligaments, tendons	–	–	–
Cortical bone	–	–	–
Synovial fluid	+–	+	++
Infiltrates, edema	+–	+	++
Nerve tissue	++	++	++

■ Table 2.3. Weighting of MRI images

Ratio of repetition/echo time	Repetition time (TR) (approx.) [ms]	Echo time (TE) (approx.) [ms]	Weighting
Short/long	500	15	$t_1$
Long/short	2000	15	Proton density (PD)
Long/long	2000	90	$t_2$

the legal basis. More important and more specific are the reports issued by the International Commission of Radiation Protection (ICRP) in 1977 and the WHO Study Group in 1987 [3]. A general principle applies: Doses of ionizing radiation used in humans should be kept as low as reasonably achievable (ICRP 1977). The specific radiation protection measures will be discussed in detail in the relevant x-ray investigations.

! **»The best radiation protection is the avoidance of unnecessary x-ray examinations and the restriction of the number of required x-rays to a diagnostically useful minimum« [2].**

A few generally valid guidelines of relevance to orthopaedic diagnostics are mentioned below.

### Field size

This is the most important dose-increasing factor in infants and small children. Every unnecessary increase in the radiation field produces a greater increase in dosage, the smaller the initial field [2]. Enlarging a field from a 13 × 18 format to an 18 × 24 format almost doubles the main entrance dose, e.g. for a general pelvic x-ray. Our radiology technicians always strive therefore to frame a prescribed x-ray with the smallest possible field size.

### Beam path

In many cases the PA projection, e.g. for recording spinal x-rays in adolescent girls, is a suitable way of reducing the dose in organs that are especially sensitive to radiation (mammary gland, gonads, lens, thyroid gland).

### Additional filtering on the x-ray tube

An additional 0.1 mm copper filter can reduce the proportion of »soft« radiation such that the absorbed dose not contributing to the imaging process is lowered by 50%. A disadvantage is a reduction in image contrast, although this is generally acceptable.

### Anti-scatter grid

The purpose of this grid is to reduce the scatter radiation, and thus improve image definition, when large body volumes are being exposed. In small body parts of small patients, however, the scatter radiation is so low that the image quality is hardly affected. Dispensing with grid x-rays for the infant pelvis reduces the dose by a factor of 2.5.

### Intensifying screens

These screens produce a change in the sensitivity of the film-foil system between 200 and 1,600 units. A system with a relative sensitivity of 400 units is generally recommended for pediatrics. Exceptions are detailed views of bone with a recommended 200 units and follow-up

x-rays of known abnormalities, e.g. scolioses with a recommended 800 units. An important consideration in this context is the coordination of the emitted light quality. Green-emitting screens will only achieve the desired effect in combination with green-sensitive films.

### Gonad protection

Gonad protection is always used provided it does not conceal any diagnostically important structures, i.e. in boys a testicular capsule is used if the testes have descended, and a roughly triangular shield is placed above the symphysis in girls.

Parts of this chapter (primarily concerning radiation protection) were incorporated from the 1st edition, for which I should like to thank the author C. Fliegel.

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## 2.1.5 School medical examination

### Objectives from the orthopaedic standpoint

» **An adolescent's worst lament is to suffer from a back that is bent.** «

In all developed countries, mass screening programs are conducted to detect abnormalities and illnesses at an early stage. In the past, such programs focused on the detection of infectious diseases such as tuberculosis, hence the routine use in many places of imaging investigations. In recent years these investigations have almost completely disappeared, and tuberculosis screening is only appropriate in regions where the disease is endemic [5]. Apart from eyesight and hearing tests, medical examinations in schools focus primarily on the detection of abnormalities of the musculoskeletal system.

! **The objective of medical screening programs in schools is always to detect problems at an early stage so that appropriate treatment can be initiated before they reach the stage where much more expensive therapy is required.**

As well as recording the public health status and attempting to avoid serious complications in later life, such programs must also consider not least the economic aspect. Overall, the school medical screening program should be cheaper than the subsequent expensive treatments.



### 2.1.5 · School medical examination

The following musculoskeletal abnormalities should be investigated in a school medical screening program:

- abnormal height (stunted growth or abnormally tall),
- gait disorders,
- congenital malformations,
- scolioses,
- hyperkyphoses,
- leg length discrepancies,
- axial deviation of the lower extremities,
- foot abnormalities.

All these problems can be investigated without sophisticated and expensive measuring instruments and they are all outwardly visible. Some may question the omission of *postural abnormalities* from the list of problems to be investigated. However, this problem is discussed extensively in ► chapter 3.1.3, where it is stated that »poor posture« in children and adolescents is, to a certain extent, physiological. A posture, can, by definition, be straightened and does not involve any structural change. Poor posture is primarily a muscular problem, and the borderline between poor and normal posture is completely arbitrary. Muscles can only be trained through exercise and it is usually non-sporting children that tend to exhibit a flaccid posture. The child will only exercise if it is motivated to do so, and whether this can be achieved by the introduction of special »*postural physical education lessons*« is extremely doubtful. Children and adolescents who are obliged to take part in such PE lessons refer to them as »hunchback PE« or »cripple PE« lessons. Their unaffected classmates also use similar expressions. *Since such PE lessons are demotivating for all those involved, they inevitably fail to achieve their objective.* It is much more useful to recommend a sport that such children can practice with a certain amount of enjoyment. Since these children tend to lack good motor skills, sports in which they have to compete with others are not particularly appropriate. They will always perform less effectively than their friends and therefore become demotivated. More suitable are sports in which they only have to compete with themselves and can determine their own improvement in performance, i.e. individual-based sports such as swimming, athletics, cycling, running, etc.

#### **What should be examined and how, and when is referral to a specialist indicated?**

**Height:** We first check the patient's *height* while standing. The height should be compared against nomograms or growth curves. If the child is below the 3rd percentile, the doctor must establish whether a hereditary condition is present. Above the 97th percentile, and especially in the case of girls, the doctor must consider whether steps should be taken to avoid an excessive final height, for

example by hormone replacement or epiphyseal closure at the distal femur and proximal tibia.

- Referral to specialists: Height below the 3rd or above the 97th percentile.

**Gait disorders:** A range of relatively simple tests is available for checking coarse motor function, for example walking along a line, hopping on one leg, etc. Clearly visible gait disorders such as an equine gait are also readily discernible. A more discriminating assessment is not possible, however, within the context of a mass screening program. Rather than the systematic investigation of all pupils, a much more efficient approach is to include the PE teacher in this examination. He knows those pupils who are always last and who seem particularly clumsy. The doctor then only has to examine these specific pupils more closely.

- Referral to specialists (neurologically oriented pediatricians or correspondingly trained pediatric orthopaedists): Assessment of walking, hopping, jumping or diadochokinesia suggests the presence of a coordination disorder.

**Malformations:** Malformations are rarely detected for the first time during a school medical examination. Parents usually notice these at the birth and seek medical advice at an early stage. Hand deformities requiring surgery must, as a rule, be treated during the first two years of life. Fairly serious foot deformities are also often treated during infancy. Major shortening of the upper and lower extremities is likewise detected at an early stage. Abnormalities detected primarily by the school doctor include the occasional case of pectus excavatum or carinatum.

- Referral to specialists: *Sternal abnormalities* (pectus excavatum or carinatum): indentation or protrusion of 2 cm or more above or below the thorax level.

**Leg length discrepancies:** Leg lengths are best measured during the school medical examination by the indirect method by assessing the iliac crest (see ► chapter 3.1.1).

- Referral to specialists: Leg length discrepancy of 1 cm or more.

**Back problems:** In purely quantitative terms, *scoliosis* is the commonest growth disorder that can be detected at an early stage by the school medical examination. The prevalence of scoliosis in Europe is 2–3%, and girls are around three times as likely to be affected than boys [3, 7, 8]. At the end of the 1970's and the start of the 1980's, screening programs using Moiré topography or the ISIS method and similar devices [2, 4] were developed (► Chapter 3.1.4). Over the years, however, these optical detection screening methods have proved to be no better than clinical examination, since they produce a large number of false-positive results. The forward-bending test has proved to



be sufficiently reliable [1, 6] and is described in detail in ► chapter 3.1.4.

- Referral to specialists: Patients with a rib or lumbar prominence of 5° or more.

In addition to the forward-bending test, we also observe *vertical alignment* while standing. For this we use a symmetrical weight hanging from a piece of cord and suspended from the vertebra prominens. The vertical alignment should be determined if boards are needed to offset leg length discrepancies.

- Referral to specialists: Vertical alignment is not in line with the anal cleft, but 1 fingerwidth or more to the side.

Another very common disorder is *fixed hyperkyphosis*. We observe whether the thoracic kyphosis evens out as the patient straightens up from a forward-bending posture (► Chapter 3.1.1).

- Referral to specialists: Fixed kyphosis in the thoracic spine area or (also including mild) kyphosis of the lumbar spine (usually identifiable by areas of pigmentation over the spinous processes in the upper lumbar spine area).

**Axial deviations of the lower extremities:** Up until the age of 10 years or so, cases of genu valgum are almost always physiological. In toddlers, cases of genu varum are invariably pathological, and a genu varum in children under 10 years of age needs to be investigated. Cases of genu valgum with an intermalleolar gap of up to 10 cm can readily be tolerated. They primarily give cause for concern if the child is obese. In adolescents, a genu varum with an intercondylar gap of up to 2 fingerwidths and a genu valgum with an intermalleolar gap of up to 3 fingerwidths can be tolerated. The degree of anteversion can readily be measured clinically (► Chapter 3.2.1).

- Referral to specialists: Anteversion of more than 40° measured clinically in an adolescent.

Torsion abnormalities are frequently observed in the lower leg. The foot axis is normally between 0° and 30° lateral to the femoral axis.

- Referral to specialists: Lower leg medial torsion of less than 0° or lateral torsion of more than 30°.

**Foot abnormalities:** Observing the callus formation on the feet is extremely useful. Provided the arch of the foot does not come into contact with the floor on the medial side beneath the navicular and provided no callosity is present, then the child probably has normal feet. So-called »flat feet« are physiological in children.

- Referral to specialists: Callus formation on the medial arch of the foot.

Occasionally the doctor will observe a case of *juvenile hallux valgus*.

- Referral to specialists: Redness over the first metatarsal head or a valgus deviation of the great toe of more than 20°.

### Equipment for the school medical examination

An adequate examination can be conducted with a few simple items:

- tape measure for measuring height,
- chair,
- boards for offsetting leg length discrepancies in thicknesses of 5 mm, 1 cm, 1.5 cm and 2 cm,
- protractor, possibly scoliometer (► Chapter 3.1.4),
- plumb line,
- examination table.

### At what age should school medical examinations take place?

From the orthopaedic standpoint, the aim of the examination is, firstly, to identify congenital disorders and, secondly, to detect growth disorders that develop during puberty. Since an annual school medical examination is not feasible for reasons of cost and organization, we recommend as a minimum that children be examined when they start compulsory education and again at puberty.

- All congenital deformities should be apparent soon after the start of compulsory education, i.e. around the age of 7 years. Ideally, the child should already have been attending school for 1 year so that the PE teacher can report whether any coordination disorder is present.

In later years, up until the start of puberty, orthopaedic conditions almost always manifest themselves via the symptom of pain, e.g. Legg-Calvé-Perthes disease or juvenile rheumatoid arthritis. The pain prompts the parents to take the child to the doctor even if no mass screening program is available.

*In both quantitative and qualitative terms, scoliosis represents the number one problem during the growth phase.* Scoliosis develops at the start of puberty and is known to start two years earlier in girls than in boys. The often used examination age of 14 years is clearly too late.

- The examination at the start of puberty should ideally take place at 12–13 years of age.

This is also the age at which epiphyseal separation is most frequently observed.

Of course, although we should not just consider orthopaedic criteria when establishing the time for the school medical examination, disorders of the musculoskeletal system form a major part of the examination and should therefore be taken into account when selecting the examination period.

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## 2.2 Development of the musculoskeletal system

### 2.2.1 Growth

#### Definition

Growth is a process that serves to enlarge or lengthen an organ. Growth processes take place primarily in children and adolescents. Skeletal growth has been studied extensively. Long bones possess their own growth system in the form of epiphyseal plates. But it is not just the skeletal system that possesses the ability to growth: Every other organ also grows, although we know relatively little to date about the growth processes in these organs. But growth processes do not stop once a person's final height has been reached. Anabolic and catabolic processes are constantly taking place even during adulthood in almost all organ systems, particularly in bone.

#### Historical background

- 1727: In an experiment S. Hales observed that the long bones only grow at their ends. He drilled two holes as markers in the femoral bone of a 2-month old chicken and, on observing that the distance between these holes remained unchanged, concluded that no growth occurred in the shaft area [6].
- 1770: J. Hunter discovers that apposition at the bone surface and resorption in the medullary cavity are necessary for growth in bone thickness [8].
- 1858: Description of the epiphyseal plate by H. Müller [13].
- 1873: A. Kölliker discovers osteoclasts [9].
- 1953: J. Trueta investigates the blood supply to the epiphyseal plate [16].

## Skeletal growth

The skeleton develops in the embryo from a primarily cartilaginous skeletal structure. The first ossification centers then form in the diaphyseal area. At birth, the diaphyses already appear to be largely ossified on x-rays, whereas the epiphyses still lack any ossification centers, which only appear during the first few years of life. The epiphyseal plate is located between the diaphyses and the epiphyseal ossification centers.

Bone growth takes place via two mechanisms

- endochondral (interstitial) linear growth,
- periosteal (appositional) thickness growth.

Growth, particularly the type of growth that leads to an increase in diameter, also involves constant *bone resorption*.

In *interstitial growth* bone is formed from the initial cartilaginous structure. Cartilage is a semi-solid, elastic tissue and extremely old in terms of evolutionary development. Before bone appeared, fish existed with a purely cartilaginous skeleton. Individual species that still possess a supporting organ consisting exclusively of cartilaginous tissue have survived to this day. e.g. sharks.

Growth can also occur via *periosteal apposition*. Growth in thickness particularly takes place by this method. But this type of appositional growth also occurs in the animal kingdom, for example in the shells of mussels and snails or in corals.

#### Endochondral growth

Endochondral growth involves bone formation from a cartilaginous precursor. These cartilaginous precursors are found at the following sites:

- Epiphyseal plates,
- Apophyseal plates,
- Articular cartilage.

Growth takes place in all 3 cartilaginous zones, although the most significant zone in quantitative terms is the growth in the epiphyseal plate. The localization of growth in this area has proved to be the most favorable naturally, since it facilitates the development of form-fitting joints, which would not be the case if the main growth zone were located at the ends of bones. On the other hand, if the growth zone were located in the center of the shaft of long bones, this would probably have mechanical drawbacks, resulting in the existence of potential fracture sites in the center of the shaft because the loading capacity of cartilage – particularly for bending forces – is lower than that of bone.

### Development of the epiphyseal plate

We can subdivide the long bones into diaphysis, metaphysis and epiphysis. The epiphyseal plate is located between the metaphysis and epiphysis and is demarcated from the cancellous bone of the epiphysis by a thin layer of compact bone. This bone plate is clearly visible on an x-ray and often persists into old age even after closure of the epiphysis.

In the epiphyseal plate itself, we can distinguish 4 zones histologically (■ Fig. 2.4 and 2.5):

- zone of resting cells, generative zone,
- layer of cartilage cell columns,
- layer of hypertrophic cells,
- zone of primary calcification and ossification.

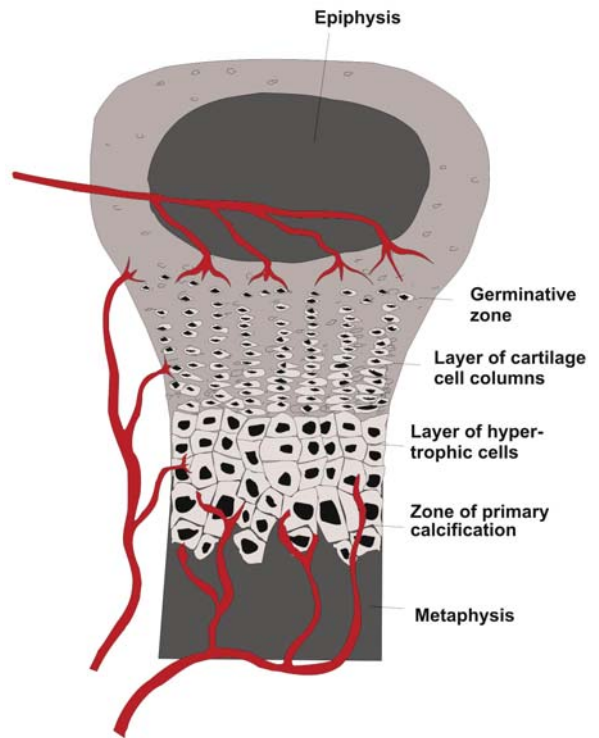
From the clinical or functional standpoint, we can distinguish between two plate sections

- the epiphyseal section, with the potential for proliferation,
- the metaphyseal section, without cell proliferation.

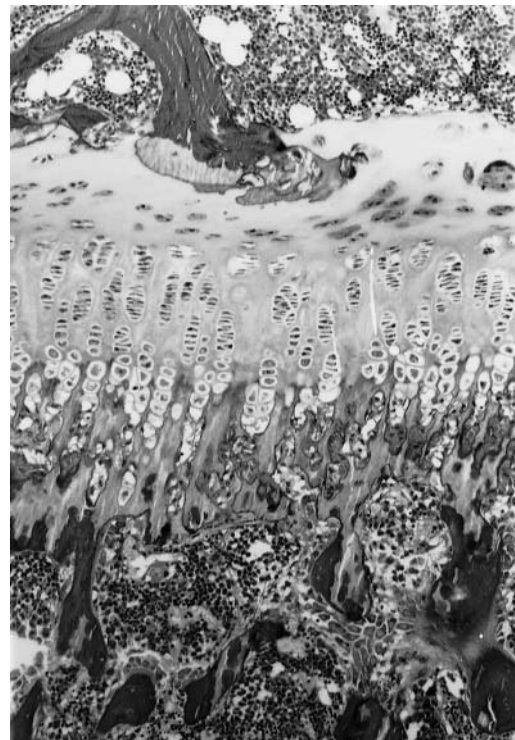
In the epiphyseal area, the matrix part predominates over the cellular elements. In the metaphyseal part of the plate, however, the cellular portion predominates in relation to the matrix. The mechanical strength of the cell-rich metaphyseal section is lower than that of the matrix-rich epiphyseal section. The plate is surrounded by perichondrium, which is responsible for the growth in thickness of the cartilaginous plate. The layer of chondrocyte columns accounts for roughly half of the total plate thickness. The cells in this zone are tightly packed together and flattened against each other.

Cell replication occurs in the section where these columns face the epiphysis, which thus constitutes the actual »energy source« for linear growth. The cells on the metaphyseal side grow increasingly larger and eventually hypertrophy. The intercellular substance declines, causing the individual cells to lie closer together. The cell nuclei are now located in the cell centers and no longer at the edges. These cells degenerate, producing the first bone trabeculae. The first signs of calcification are observed in the greatly reduced intercellular substance between the hypertrophied cells and the degenerating cells as calcium salts in the form of granules.

Three separate *vascular systems* supply the metaphysis, perichondrium and epiphysis and their respective plate sections. The metaphyseal and epiphyseal vascular systems communicate with each other via the perichondrial system. But individual small vessels also pass directly through the epiphyseal plate [4], particularly during infancy, although such vessels also appear to be present during adolescence. This explains why the epiphyseal plate, unlike articular cartilage, does not form an absolute tumor barrier, and why metaphyseal tumors can spread through the plate into the epiphysis. These vessels are



■ Fig. 2.4. Diagram of the structure of the epiphyseal plate. The cells are formed in the germinative cell layer and then align themselves in columns and hypertrophy before undergoing mineralization. The circulation is at its lowest and mechanical strength at its weakest in the zone of hypertrophic cells



■ Fig. 2.5. Histology of the epiphyseal plate

### 2.2.1 · Growth

still relatively large in infancy, allowing even a metaphyseal osteomyelitis to manifest itself very rapidly as a joint infection.

The perichondrial vascular system breaks down during puberty and is gradually replaced by vessels which, after closure of the epiphyseal plate, grow through the plate from the metaphyseal to the epiphyseal side. The vascular connection between the metaphysis and epiphysis is at its weakest, and the number of vessels penetrating the plate is at its lowest, during the pubertal growth spurt. Any disruption of the perichondrial vascular system at this time can have disastrous consequences for the epiphysis, possibly resulting in necrosis (■ Fig. 2.6).

#### Factors affecting endochondral growth

Growth is regulated by *hormones*. The most important hormone is somatotropin (STH). This growth hormone directly affects the activity of the epiphyseal plate and indirectly influences its mechanical strength [12]. The more active the plate, the lower its loading capacity. The sex hormones also influence growth. Testosterone promotes growth and reduces the mechanical strength of the growth plate, while estrogen promotes maturation and thus indirectly increases the loading capacity. Thyroxine produces a catabolic effect, inhibiting the activity of the growth plate and likewise increasing its strength.

A general reduction in the mechanical strength of the epiphyseal plate, particularly in the metaphyseal section, i.e. the zone of hypertrophic chondrocytes, occurs during the pubertal growth spurt under the influence of hormones. This plate section is where epiphyseal separations occur, not just after trauma, but also during the pubertal growth spurt as a result of chronic mechanical overload (e.g. in overweight individuals), particularly in those plates exposed to high shear forces because of their three-dimensional orientation. While this particularly applies at the proximal femoral epiphyseal plate, the ring apophyses in the spinal column are also weakened. Consequently, even Scheuermann disease, which involves the protrusion of disk tissue through the cartilaginous ring apophysis into the vertebral bodies, must be viewed as an expression of this mechanical weakening of the cartilage during puberty. If processes surrounding, and directly influencing, the plate result in an increased blood flow to the epiphyseal vessels – and thus to enhanced growth cartilage function – *stimulation of the epiphyseal plate* can be expected to occur. This increased perfusion can be induced by healing processes after fractures in the metaphyseal or epiphyseal region, but also in the diaphysis.

Growth can likewise be stimulated by osteomyelitis or a tumor, but also by continuous tensile forces [17]. Thus, plate stimulation can occur after division of the periosteum as a result of the reduction in pressure in the growth



■ **Fig. 2.6.** 14-year old girl with a trochanteric avulsion fracture (a), which healed without problems with conservative treatment. A femoral head necrosis developed 6 months later (b). Evidently the circumflex femoral artery had also been injured at the time of fracture. Since the vascular connection between the metaphysis and epiphysis is at its weakest and the number of plate-penetrating vessels is at its lowest at this age, the circulation to the femoral head is highly jeopardized. This complication would not occur in an adult with this type of fracture

plate [17]. However, the extent to which such processes are hormonally controlled [2] remains unclear.

*Growth may be inhibited* if the plate is injured. Growth is disrupted particularly if the germinative layer is damaged. While this is usually the case with so-called epiphyseal fractures, the germinative layer can also occasionally be affected in epiphyseal separations, which generally occur in the metaphyseal section of the plate. Similarly, a growth disorder, i.e. premature closure of the plate, can also result from a distraction epiphysiolysis, particularly if the distraction occurs too rapidly [3, 5, 10, 18]. But continuous pressure can also lead to a growth disorder. In cases of extensive diaphyseal lengthening, the abnormal pressure can induce diminished growth in the adjacent epiphyseal plates [7].

Endochondral growth is very strongly influenced by mechanical loading. The epiphyseal plate reacts to bending stresses by attempting to realign itself at right angles to the axis of the main force flow. Experiments and clinical experience have shown a correlation between cartilage growth and the degree of axial pressure. The application of excessive eccentric pressure to the epiphyseal plate stimulates the plate, which responds with additional growth to offset any axial deviation. This illustrates how the body reacts to a functional problem with functional adaptation. However, if the excessive pressure goes beyond a certain level, growth is suppressed, the axial deviation deteriorates and the mechanism of functional adaptation is interrupted [14].

The forces generated at the epiphyseal plate by the pressure of growth can be considerable, which explains



how even fairly trivial bone fractures within the epiphyseal plate can break open again as growth continues.

### Periosteal growth

➤ In the shaft area, apposition of bone substance proceeds directly from the periosteum (desmogenous), without taking the indirect route via a cartilage matrix. The osteoblasts originate from the periosteum.

The increase in bone thickness is primarily attributable to periosteal growth. Periosteal bone formation also plays an important role in fracture healing. The ability of the periosteum to form bone is also exploited in diaphyseal or metaphyseal bone lengthening [1].

A range of substances contained within bone that stimulate the formation of new bone has been identified in recent years: bone morphogenetic proteins (BMP), transforming growth factor  $\beta$  (TGF- $\beta$ ), insulin-like growth factors I and II (IGF-I and IGF-II), platelet derived growth factor (PDGF) and fibroblast growth factor (FGF) [15]. BMP is the only one of these factors capable of also stimulating the formation of new bone outside existing bone (heterotopic ossification). These substances are sometimes used therapeutically. Stem cell transplantation and gene therapy are also discussed as possible treatment options for stimulating bone healing [11].

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## 2.2.2 Physical development

### Stages of life

At birth, the average infant is 20 inches (50 cm) long and weighs 6–8 pounds (3–3.5 kg). By the time the infant reaches adulthood it will have grown by 350% and increased in weight by a factor of 20. Physical development occurs in stages (Table 2.5).

During *infancy* the child is completely dependent on its parents. Various development steps and abilities are typically reached at the following times.

#### Typical periods for development steps and abilities

2nd–5th month:	Smiles spontaneously
2nd–4th month:	Head control
3rd–6th month:	Turns from a prone to a supine position and vice versa
5th–8th month:	Sits independently
6th–9th month:	Pulls itself up using furniture, starts to stand
9th–13th month:	Says »mommy« and »daddy«
10th–16th month:	Drinks from a cup
11th–16th month:	Walks independently
14th–22nd month:	Climbs stairs
14th–30th month:	Combines 2 different words
20th–30th month:	Puts on clothing
22nd–30th month:	Hops on the spot

During *childhood* growth proceeds at a constant rate (and certainly does not occur in the »spurts« frequently perceived by parents).

*Adolescence* is characterized by the physical changes associated with sexual maturation and the pubertal growth spurt.

### Linear development

➤ The influence of growth must always be considered when evaluating orthopaedic problems in children and adolescents.

## 2.2.2 · Physical development

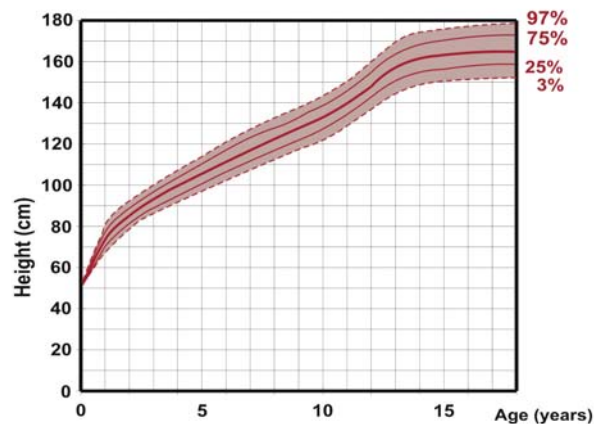
This is where pediatric orthopaedics differs substantially from adult orthopaedics, in that the former is based on knowledge of the natural growth processes. In addition to height, the relative proportions also play a significant role. It is particularly important to consider the ratio of trunk length to overall height. In certain types of dysostosis, the ratio of upper to lower leg length and of upper arm to forearm length is also altered.

Height is related to the stage of growth. The *growth rate* does not remain constant throughout physical development. While it largely follows a linear pattern during childhood, the growth rate increases markedly during two phases of life, namely during infancy and puberty (■ Fig. 2.7). The fastest growth occurs during pregnancy and in the first year of life. Growth then remains constant for several years until the onset of the pubertal growth spurt, which occurs some 2 years earlier, on average, in girls than in boys.

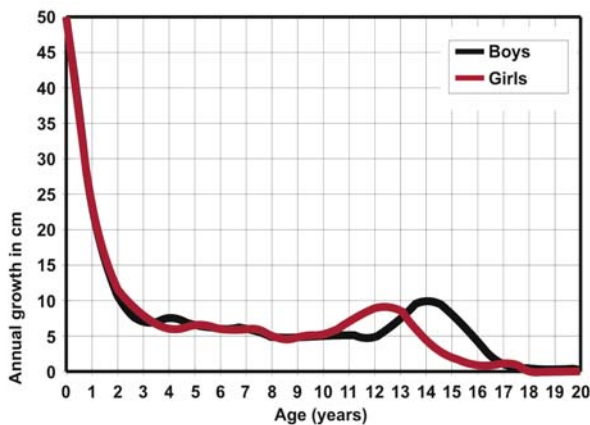
*Height* is dependent on the rate of growth and is compared against standardized values. These standardized values are based on the statistical analysis of very large populations. Unfortunately, most of the corresponding tables are already several years old and, given the phenomenon of *acceleration*, no longer accurately reflect the current situation, since children now grow taller, on average, than they did 20 years ago (► Chapter 1.3). However, the tables are still usable as approximations.

The *standardized values* for height are subject to a certain degree of *dispersion*. For everyday clinical use, the

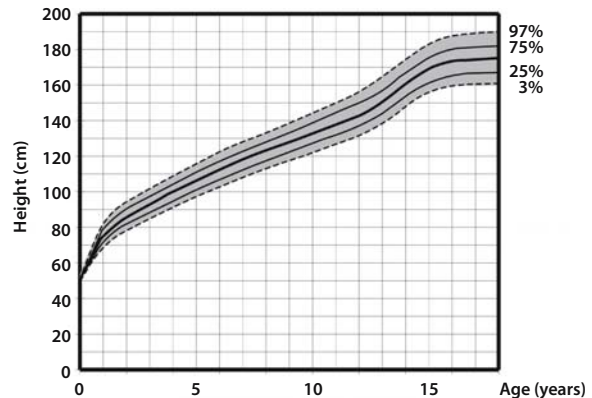
graphical presentation of the *mean values for height* has proved most effective. The means are supplemented by *percentiles*, which provide a measure of the dispersion. The percentile figure indicates the percentage of boys or girls that are *smaller* than the graph reading. Thus, only 3% of children are smaller than the figure for the 3rd percentile, 25% are smaller than the 25th percentile etc. Normally, the 3rd, 25th, 50th, 75th and 97th percentiles are shown (■ Fig. 2.8 and 2.9) [1]. *Stunted growth* is the term used to refer to children below the 3rd percentile, while *dwarfism* involves a (presumed) final height of less than 150 cm.



■ Fig. 2.8. Growth curve for girls: Height with mean and percentiles according to age. (After [3])



■ Fig. 2.7. Growth rate: Annual height gain in boys and girls. (After [3])



■ Fig. 2.9. Growth curve for boys: Height with mean and percentiles according to age. (After [3])

■ Table 2.5. Phases of physical development from birth to adulthood

Age group	Age	Subdivision	Age
Infancy	0 to 1.5 years		
Childhood	Girls: 1.5 to approx. 10 years Boys: 1.5 to approx. 12 years	Preschool age	1.5 to 7 years
		School age	7 to approx. 10 or 12 years
Adolescence	Girls: approx. 10 to 14 years Boys: approx. 12 to 16 years		



*Height* is measured with the child standing barefoot against a wall. The heels and head should be in contact with the wall and the child should look straight ahead. The *trunk length* is best determined as a *sitting height*. To this end, the patient sits on a (hard) chair with known seat height. The height while sitting is measured and the seat height subtracted. The percentage ratio of trunk length to height declines during childhood from 70% to 48% (■ Fig. 2.10) [2]. Between birth and 5 years of age, the trunk and leg length both increase in linear fashion, while between 5 years and puberty, leg growth is responsible for two-thirds of the increase in height and trunk growth for the other third. This ratio is then reversed after puberty [2]. The ratio of trunk length to height is altered in the disproportionate growth of dwarfism, in which the growth of the spinal column and head is less impaired than that of the extremities (e.g. in achondroplasia and hypochondroplasia).

*Arm span* correlates very closely with height. In 77% of individuals the difference is less than 2 inches (5 cm), in 22% it is between 2 and 4 inches (5 and 10 cm) and in only 1% of cases does it exceed 4 inches (10 cm) and arouse suspicion of a Marfan syndrome [2].

**Weight**

Weight gain does not follow the same pattern during development as the increase in height. Weight can likewise be compared against graphs showing means and percentiles. The 5th, 25th, 75th and 95th percentiles are usually shown for weight (■ Fig. 2.11 and 2.12) [1]. Children and adolescents above the 95th percentile are obese. Obesity has a considerable effect on growth disorders, and obese children and adolescents should be referred, with their parents, for dietary counseling. While no difficulties with orthopaedic problems would be expected in adolescents (particularly girls) below the 5th percentile, they may have an anorexic mindset and thus require psychological counseling.

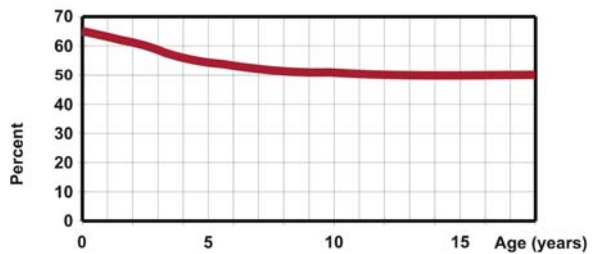
**Skeletal age**

The growth status, and thus the predicted final height, can be determined much more accurately if the actual skeletal maturation (the »skeletal age«) is used as the basis for calculation rather than the chronological age. Not all children mature at the same rate, and the physiological range incorporates differences of ± 2 years. The same phenomenon is also observed with hormonal maturation. Some girls experience the menarche as early as 10 years, while others have to wait until they are 15. In boys, the voice can break between the ages of 12 and 16 years of age. The chronological age can differ from the skeletal age by many years, particularly in severely disabled individuals.

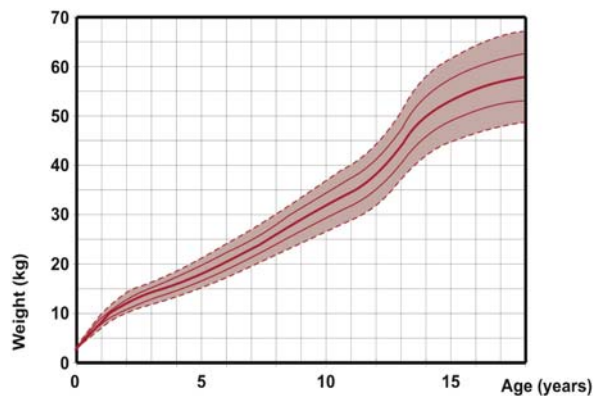
**Puberty**

The onset of the pubertal growth spurt occurs in girls at about aged 10 compared to around 12 years in boys

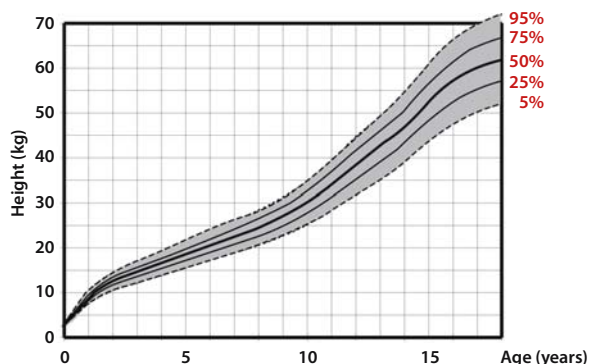
and lasts for 3–4 years. The rate of growth (particularly of the spinal column) is much faster during this period than during childhood (■ Fig. 2.7). After the growth spurt has peaked (or in the middle of the spurt), the menarche occurs in girls and the voice breaks in boys. Besides the menarche and the breaking of the voice, the state of maturation can also be evaluated on the basis of pubic hair, the development of the genitals(boys) and breast development(girls) [4] (■ Table 2.6). Both the menarche and the start of ossification of the iliac crest apophysis (Risser sign, ► chapter 3.1.4) occur around 1 year after the



■ Fig. 2.10. Ratio of sitting height to overall height. The sitting height is stated as a percentage of overall height. (After [1])



■ Fig. 2.11. Weight in girls: Average with mean and percentiles according to age. (After [3])



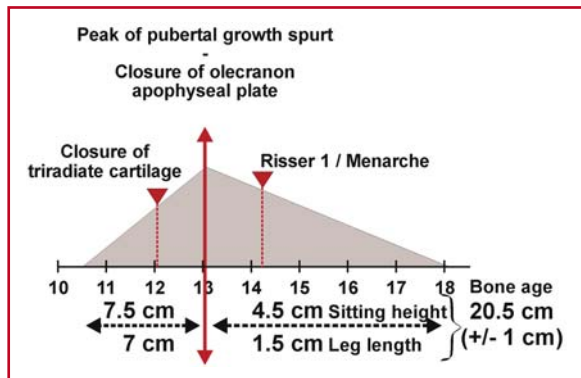
■ Fig. 2.12. Weight in boys: Average with mean and percentiles according to age. (After [3])

## 2.2.2 · Physical development

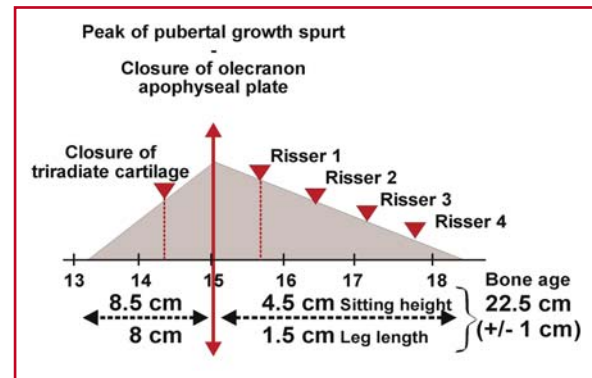
peak of the growth spurt, although a closer correlation exists with the closure of the olecranon apophyseal plate (■ Fig. 2.13 and 2.14) [2].

In *endocrinological* terms, puberty starts even before the outwardly visible changes with the secretion of gonadotropins by the anterior lobe of the pituitary gland at around age 8 in boys and 7 in girls. The gonadotropic

hormones stimulate the gonads to secrete sex hormones (testosterone and estrogen) which, in turn, stimulate the secretion of growth hormone (STH = somatotropic hormone) in the pituitary (prepubertal growth spurt). At fairly high concentrations, the sex hormones inhibit the secretion of growth hormone and maturation is promoted until growth is completed.



■ Fig. 2.13. Pubertal growth spurt in girls. (After [2])



■ Fig. 2.14. Pubertal growth spurt in boys. (After [2])

■ Table 2.6. Evaluation of the maturation status in boys and girls

Stage	Characteristics	Duration
<i>Development stages of the genitalia in boys</i>		
Stage 1	<i>Prepuberty:</i> Scrotum and penis remain the same size as during childhood	
Stage 2	Enlargement of scrotum and testes	1 year
Stage 3	Lengthening of penis	1 year
Stage 4	Penis becomes larger and thicker, the glans develops, the scrotal skin turns a darker color	2 years
Stage 5	Genitalia assume their adult form	
<i>Maturation stages of pubic hair in boys and girls</i>		
Stage 1	<i>Prepuberty:</i> Still without pubic hair	
Stage 2	Sparse growth of fine, light-colored, downy hair, which is straight or only slightly curly, primarily on the root of the penis and the labia	1 year
Stage 3	Hair becomes darker, coarser and more curly. Downy facial hair in boys	1 year
Stage 4	Hair growth resembles the adult pattern, but the area covered by the hair is smaller, hair growth also in the armpits. Facial growth more pronounced in boys	2 years
Stage 5	Pubic hair assumes its adult form	
<i>Development stages of the breast in girls</i>		
Stage 1	<i>Prepuberty:</i> Still without breasts, but nipples project	
Stage 2	<i>Budding breast:</i> Projection of the breast and nipple as a small protuberance	1 year
Stage 3	Further enlargement and swelling of the breast without demarcation of contours, the areola also grows	1 year
Stage 4	Separate swelling of the areola and nipple across the actual surface of the breast	2 years
Stage 5	Breast assumes its adult form. The areola recedes into the general contour of the breast, and only the nipple projects	

## Constitution

As in adults, a distinction can often be made in children, and always in adolescents, between athletic, leptosomic and pyknic physiques.

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### 2.2.3 The loading capacity of the musculoskeletal system

We have become accustomed to seeing teenagers winning Olympic medals in certain sports, for example in swimming, gymnastics or figure skating. But performance training frequently begins before the completion of growth in other disciplines as well.

**! Sporting activity almost invariably involves periods of acceleration and thus forces and torques. Potential energy is constantly being converted into kinetic energy and vice versa. If the forces exceed a certain level, the structure subjected to the greatest stress will fail at the point of weakest mechanical strength.**

The musculoskeletal system consists of bones, muscles, tendons, ligaments and articular cartilage. All these structures undergo major changes in terms of their mechanical strength between birth and adulthood. But the most important difference between children and adults is the presence of growth cartilage.

#### **Experimental investigations on the mechanical strength of various tissue types during growth**

##### **Growth cartilage**

The growth plate normally adopts a position at right angles to the force resultants. The plates are primarily subjected to compressive and shear forces and, more rarely, to tensile forces. Typical growth cartilage plates subjected to tension are those of the lesser trochanter and humeral epicondyle. The surface quality of the growth cartilage plays an important role in the strength of the connection between the epiphyseal plate and the metaphysis. Rather than forming a flat disk with a smooth surface, the epiphyseal plate possesses indentations and notches of varying depth. These provide a certain resistance to shear forces, but do not play a major role in resisting tensile forces [9].

Growth cartilage consists of cellular elements and intercellular substance. The rate of regeneration of the chondrocytes on the one hand and their hypertrophy during development on the other determine the rate of growth. The intercellular substance with its collagen fibers maintains the internal cohesion of the growth cartilage and is almost exclusively responsible for its mechanical strength.

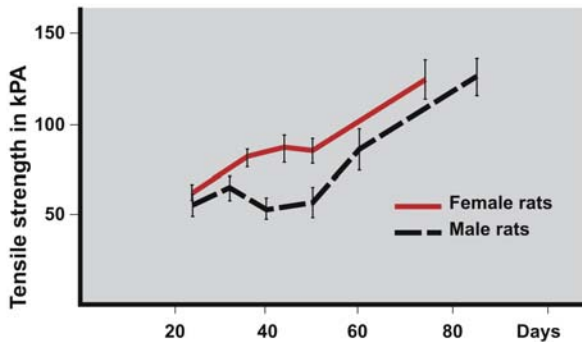
Within the epiphyseal plate we can distinguish between various zones that differ in terms of their organization, the shape and size of the chondrocytes and the quantitative relationship between cells and intercellular substance (► Chapter 2.2.1). Since the intercellular substance is primarily responsible for the mechanical loading capacity, the weakest zone is in the area of the hypertrophic cells close to the metaphysis, where the volumetric ratio of intercellular substance to cellular elements is at its lowest.

The growth rate in all mammals is at its greatest immediately after birth, and an acceleration of growth also occurs during puberty. Major changes in the mechanical properties of the growth plates occur during this phase, as has been shown by animal experiments employing tensile loads applied to the proximal epiphyseal plate of the tibia in rats [9]. Mechanical compression and distraction result in a reactive enlargement of the chondrocytes in the zone of hypertrophic columnar cartilage [8, 16].

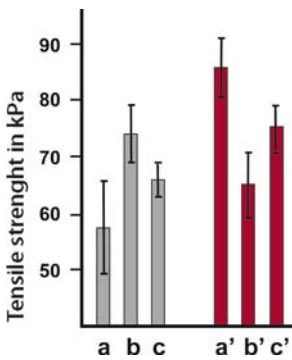
A reduction in the ability of the epiphyseal plate to withstand tensile forces is observed at the start of puberty, particularly in male rats (■ Fig. 2.15). Various hormones specifically affect the growth plate. *Growth hormone* secreted by the anterior pituitary is vitally important for normal physiology and thus also for the morphology of the epiphyseal plate. If this hormone is lacking, growth stops, the epiphyseal plate narrows and its mechanical strength increases. By contrast, the administration of growth hormone above a certain optimal level prolongs the growth period, slows down the physiological narrowing of the plate and causes the growth cartilage to remain mechanically weak for a longer period (■ Fig. 2.16).

Male and female *sex hormones* produce a wide variety of effects. During sexual maturation, the anabolic effect of the androgens predominates, while estrogens accelerate the actual maturation process. The anabolic effect of the testosterone is responsible both for the faster growth of male animals (and boys) and for the decline in the mechanical strength of the epiphyseal plate at the onset of puberty. Unlike androgens, estrogens do not have any obvious effect on protein synthesis. At low doses, they tend to produce an anabolic effect and, at high doses, a catabolic effect. During puberty estrogens slow down the activity of the epiphyseal plate, resulting in an acceleration of the maturation process of the plate. This probably explains why the phase of epiphyseal plate weakening lasts longer, and is more pronounced, in boys than in girls (■ Fig. 2.15).

### 2.2.3 · The loading capacity of the musculoskeletal system



■ Fig. 2.15. Average tensile strength of the proximal tibial epiphyseal plate in rats according to age and sex. This is markedly reduced during the pubertal growth spurt (between the 30th and 50th days of life), particularly in male animals. (After [9])

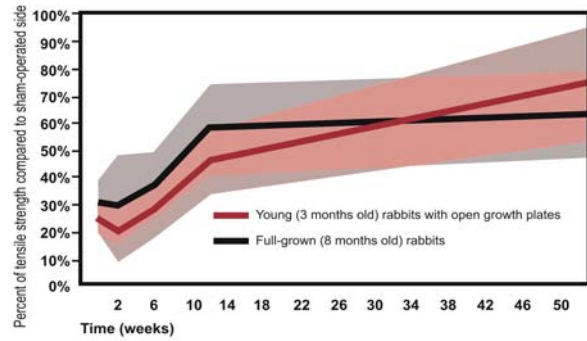


■ Fig. 2.16. Effect of sex hormones on the tensile strength of the proximal tibial epiphyseal plate in rats: a untreated normal male rats, b castrated male rats, c castrated male rats treated with testosterone, a' untreated female rats, b' castrated female rats, c' castrated female rats treated with estrogen

#### Bone

Compared to fully-developed bone, the bone in children possesses a lower elastic modulus (80 Pa at the age of 5 years, 150 Pa at the age of 40 years). The bone in small children under 6 years of age also has a lower bending strength than that of adults (150 Pa vs. 200 Pa). The immature bone bends to a greater extent when stressed and absorbs more energy before it breaks. In other words it is more plastic and less elastic than mature bone.

It follows therefore that the bone of small children fractures at lower loads compared to bone in adults, undergoes more extensive plastic deformation under the action of a given force and thus absorbs more energy. In clinical terms, this plastic deformation is demonstrated by the greenstick fractures typically seen in children. Unfortunately, since the above-mentioned study failed to measure the tensile strength of bone, no direct comparison is possible with epiphyseal cartilage. Our own tests have



■ Fig. 2.17. Ultimate tensile strength of the anterior cruciate ligament after partial division compared to the sham-operated opposite side in young and full-grown rabbits. Complete regeneration has not occurred even after a year. Recovery was slightly better in the young than the old rabbits. (After [7])

shown that the tensile strength of bone is some 25 times higher than that of cartilage. Loading is an important growth factor for bone. Experiments have demonstrated that just 15 minutes of loading is required to stimulate osteoblast growth by extracellular signal-regulated kinase (ERK) [5].

#### Ligaments

Stiffness, tensile strength and the collagen concentration of ligaments increase with age, whereas their water content decreases [20]. Our own tests have shown that the anterior cruciate ligament of juvenile rabbits has a lower tensile strength but a higher elasticity compared to that of full-grown animals [7] (■ Fig. 2.17).

Another study involving bone-ligament-bone preparations of the human anterior cruciate ligament showed that the values for elastic modulus, tensile strength and maximum load at rupture in young adults aged between 16 and 26 years were approximately two to three times higher than in older individuals in their 6th decade. We can therefore conclude that the tensile strength of ligaments increases steadily from birth until the end of puberty, but that the anchorage between ligament and bone is the critical point during this time, and that a significant reduction in tensile strength occurs in later life. The strength of ligaments and their anchorage in bone reaches a peak at the end of puberty. Strength and stiffness subsequently decline with increasing age.

#### Muscle tissue

Muscle tissue does not appear to represent a critical structure in respect of loading capacity during the growth phase. Muscles possess considerable functional adaptability and protect themselves against damage due to fatigue. They develop neurologically controlled voluntary forces and thus represent the active part of the

locomotor apparatus. But the greatest loads occur passively, e.g. as a ground reaction force on landing after a jump. These forces are neutralized in bones, ligaments and cartilaginous tissue and, to a lesser extent, in muscle tissue.

### Articular cartilage

Articular cartilage is subjected primarily to compressive and shear forces and, to a lesser extent, to tensile forces. Morphological studies have shown that articular cartilage in neonates possesses an undifferentiated structure, whereas young adults show a highly differentiated morphology, the extent of which is greatly dependent on load-bearing. In other words, the loading capacity of cartilage appears to be trainable, although this is a very gradual process. The strength of cartilage increases by up to 1000% during the course of growth and is matched by a concurrent increase of 100% in the proportion of collagen fibers [18].

### Clinical observations

#### Physiological adaptive processes

The most well-known adaptation process is the increase in muscle mass that occurs during regular training. We have already mentioned the changes in articular cartilage. Less familiar are the adaptive processes of bone. While no effect of the intensity of exercise on height has been demonstrated to date [1], growth in the width of bone does appear to occur, since measurements have shown that the bones of an adolescent undertaking sporting activity are thicker than those of inactive adolescents. Whether this represents a training effect or not is, however, by no means confirmed. It is equally possible that these differences are indicative of the average constitution of the sporting adolescent and that constitution type inherently involves a certain selection.

We also know that the menarche occurs rather later in female athletes than in their non-sporting counterparts, whereas male athletes tend to mature rather earlier on average than non-sporting boys. We have learned from surgical experience with osteotomies that the bone of sporting patients is much harder than that of non-sporting individuals (including obese patients). Evidently, the acceleration associated with sport is far more effective at strengthening bone than the force acting on the body as a result of weight.

#### Pathological changes

In *toddlers* symptoms of overload are relatively rare – primarily of course because performance training is not generally undertaken at this age. An interesting statistical analysis of skiing accidents has shown that children under 10 years of age are, on average, less likely to have an accident over a given distance traveled than adolescents and adults. Once an accident does occur, however, the

probability of suffering a shaft fracture of a long bone is significantly higher in toddlers than in older individuals. Epiphyseal fractures are much rarer at this age than during adolescence, and lesions exclusively involving the ligaments are practically non-existent. So the critical structure in toddlers is not the growth cartilage, and not even the ligamentous apparatus, but rather the bone.

The situation is completely different for *adolescents*. The growth cartilage is weakened during puberty, particularly in boys. This reduction in mechanical strength is the cause of several typical disorders and overload syndromes whose occurrence depends on the growth rate and the load. A corresponding example is *Scheuermann disease*, which involves a disparity between the actual loading and the loading capacity of the cartilaginous endplates of the vertebral bodies (► Chapter 3.1.5). This condition is much more common in boys than in girls and predominantly affects tall adolescents.

So what is the current view on the influence of sporting activity on the occurrence of this disease? Older statistical analyses show that the disease occurs more frequently in practitioners of certain sports, particularly athletes, ski racers, rowers and racing cyclists. It is difficult, however, to compare the various studies with each other since they are often based on different definitions of the condition. One study will consider the occurrence of a single Schmorl nodule sufficient to secure the diagnosis, while others refer to Scheuermann disease only in cases of fixed total kyphosis of more than 50°. Consequently some statistical analyses report the disease occurring in 50% of athletes, compared to 30% in the general population.

In an investigation conducted in our own hospital, where we apply stricter criteria, we observed the disease in 11–17% of athletes compared to 1–2% of the general population. The condition particularly affects athletes whose spine is loaded in the direction of the kyphosis. That Scheuermann disease does not occur more frequently in weight-lifters is due principally to the fact that performance training in this sport only tends to start after puberty.

But the clinical relevance of Scheuermann disease to the sufferers is the important question, and this is addressed in more detail in ► chapter 3.1.5. Overall the morbidity value of the thoracic form is fairly low, in contrast with the lumbar form. Another condition caused by the weakness of the epiphyseal cartilage during puberty is *slipped capital femoral epiphysis*. This disorder is known to occur primarily in obese and tall adolescents. But what about the influence of sporting activity? Undoubtedly, acute, obvious epiphyseal separation with distinct slippage of the femoral head is extremely rare in athletic adolescents. However, if we examine the x-rays of older patients with incipient coxarthrosis, we very frequently find that a milder form of epiphyseal separation has occurred in the past. In an investigation of the hip x-rays of



### 2.2.3 · The loading capacity of the musculoskeletal system

251 schoolchildren, tilt deformity was assessed on the basis of a ratio [11] (► Chapter 3.2.6). If this ratio was greater than 1.35, then the result was classed as a tilt deformity, suggesting that a mild form of epiphyseal separation had occurred in the past and thus constituting a type of pre-arthrosis.

This deformity was found in 9% of children who undertook little sporting activity, but in 24% of those that practiced sport very intensively. The authors concluded that this deformity was significantly more common in jumpers and long-distance runners. In our own investigation of 50 patients with coxarthrosis we observed that 58% of them had formerly been involved in competitive sports. The tilt deformity occurred much more frequently in athletes than in non-athletic types [14].

Another typical problem experienced by adolescents is the occurrence of *spondylolysis* and *spondylolisthesis*. The posterior elements of the vertebral bodies are primarily loaded in hyperextension. This applies above all to the vertebral arch of L 5, particularly when the inferior articular process of L 4 is very large and thus presses against the pars interarticularis during hyperextension. But the problem of spondylolysis is not a problem of cartilage growth, but rather one of reduced loading capacity of bone that is not yet fully developed. Spondylolysis is significantly more common in athletes, particularly track-and-field athletes, javelin throwers, gymnasts and ballet dancers (14–25% vs. 4–5% in the general population) [3, 4, 12]. But this substantial increase in spondylolysis is not matched by an increase incidence of spondylolisthesis. Evidently the well-trained muscles of the athlete prevent the forward slippage of the vertebral body despite the defect in the pars interarticularis. Again the question arises concerning the clinical relevance of spondylolysis. Most patients in the general population with a pars defect are asymptomatic, and this probably applies even more to sporting patients with good muscle function. Problems may occur at a rather later stage, after the muscle training stops or during pregnancy.

Another common back problem that can develop during adolescence is *scoliosis*. According to the latest findings, idiopathic scoliosis does not develop as a result of an asymmetrical muscle contraction, but rather a discrepancy between the growth of the vertebral bodies and that of the posterior elements – possibly as a result of an inadequate response of the posterior ligaments to the tensile loading associated with growth (► Chapter 3.1.4). The available statistics show that idiopathic scoliosis is no more common in athletes undergoing asymmetrical loading, e.g. javelin throwers, fencers and tennis players, than in the general population [12, 19]. By contrast, there is clear evidence to show that scoliosis is more common in those sports involving an extremely straight posture (rhythmic and standard gymnastics, figure skating and also ballet dancing). One excellent study found a 10-fold

increased incidence of scoliosis in practitioners of rhythmic gymnastics compared to the general population.

*Trauma* is, of course, more common in the active, sporting child than in the bookworm. At this point, we shall only highlight certain general differences from the adult situation: Compared to toddlers, adolescents suffer epiphyseal fractures slightly more frequently and shaft fractures slightly less frequently in relative terms. Another difference relates to ligament injuries: Although ligaments are more elastic and weaker in children and adolescents than in young adults, their strength is not a critical factor up to the period shortly before the completion of growth.

While we have seen a rise in recent years in the numbers of intraligamentous tears of the anterior cruciate ligament in adolescents, exclusively ligamentous injuries are rare up to the end of puberty. Much more common is avulsion of the ligament at the cartilaginous-bony attachment, probably because of the higher relative speed of the external force compared to adults [2]. Tendon attachment avulsions are also very typical in adolescents. Such *apophyseal injuries* typically involve the patella (30%), pelvis (23%), spinal column (20%) and proximal femur (19%) [15] (■ Fig. 2.18). *Chronic overloading of the apophyses* is also very widespread at this age, most typically in the form of Osgood-Schlatter disease (► Chapter 3.3.3.3).

A final comment concerns *vertebral fractures*. In contrast with adults, vertebral fractures in children occur more frequently in the upper and mid-thoracic area [13]. This zone is protected by the rigid rib cage in adults, who therefore tend to suffer fractures primarily at the thoracolumbar junction or in the lumbar or cervical spine.



■ Fig. 2.18. Typical injury in adolescents: Apophyseal avulsion (here in the anterior inferior iliac spine, attachment of the rectus femoris muscle) in a 15-year old boy

### Conclusions

The musculoskeletal system consists of tissues with widely differing characteristics. Overloading causes failure to occur at the weakest point. The mechanical properties of the various structures change during growth.

**! The critical structure in small children under 10 years of age is bone tissue, while this role is assumed by growth cartilage in adolescents. In young adults, the ligaments can ultimately be described as the weakest point in the tissue system of the musculoskeletal apparatus. In old age – because of osteoporosis – bone again becomes the tissue with the lowest loading tolerance [6].**

So what advice should we give to young athletes? When should they start performance training? Should adolescents wait until growth is completed and run the risk of no longer being competitive? Before we can answer these questions we need to know more about the long-term effects of the aforementioned illnesses.

**! Legitimate doubts exist as to whether a thoracic Scheuermann disease or spondylolysis actually represents a major problem in later life. This does not apply, however, to a tilt deformity, which leads to impingement in the hip and constitutes a distinct form of pre-arthrosis – and coxarthrosis does actually appear to be more common in former athletes than in the general population. Consequently, excessive loading should be avoided particularly during early puberty.**

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## Diseases and injuries by site

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### 3.1 Spine, trunk

#### 3.1.1 Examination of the back

##### History

- **Trauma history:** Has *trauma* occurred?  
If so:
  - When did the trauma occur?
  - What was the patient doing (sport, playing, normal routine)?
  - Direct or indirect trauma?
- **Pain history:**  
Where is the pain located (neck, upper thoracic spine, lower thoracic spine, lumbar spine, lumbosacral spine)? When does it occur? Is it related to loading or movement, or does it also occur at rest (e.g. while sitting) or even at night? If so, does the pain occur only while changing position, or does the pain cause the patient to wake up at night? Does the pain occur on bending down or straightening up again? Does the pain also radiate to the legs? Does the pain occur on coughing or sneezing?
- **Sports history**  
What sports does the patient practice outside school? If spondylolysis is suspected ask specifically about the following activities: gymnastics, figure skating, ballet, javelin-throwing. If Scheuermann disease is suspected ask specifically whether the patient is involved in cycle racing or rowing.
- **Neurological symptoms**  
Is a leg weakness present and, if so, since when? Are there problems of micturition or defecation?

##### Inspection

After the gait analysis (▶ Chapter 2.1.3), the standing patient's back is inspected from behind.

**!** To ensure that the patient's back is at eye-level, the examiner himself should not stand but preferably sit on a chair of the appropriate height (■ Fig. 3.1).

##### — Inspection from behind

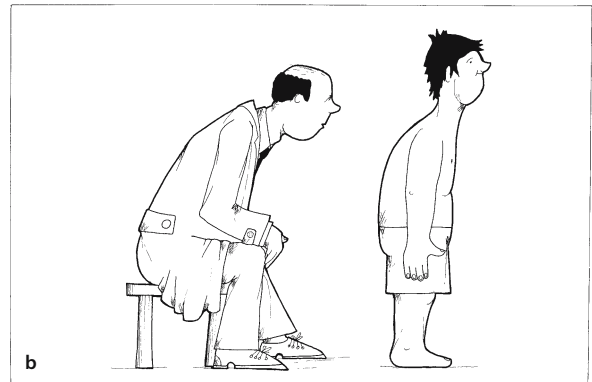
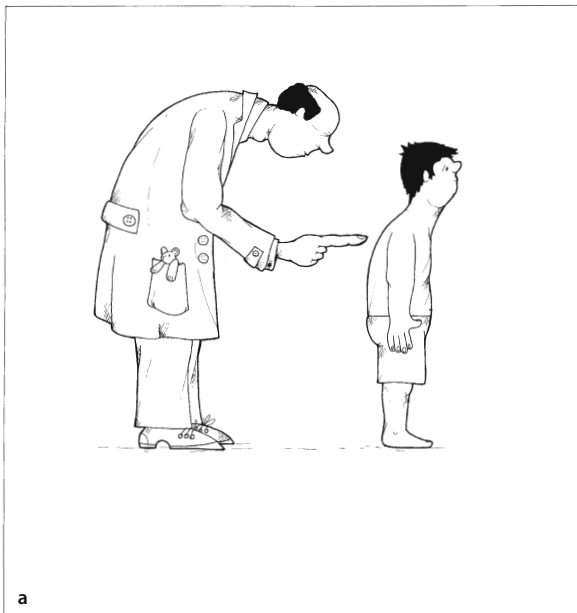
We observe the position of the shoulders, the height of the scapulae and particularly the symmetry of the waist triangles. We look for pigmentation over the spinous processes, especially over the lumbar spine, as this can be an indication of (usually pathological) kyphosis in this area. A (hairy) nevus in this area can be a sign of an intraspinal anomaly.

##### — Inspection from the side

We assess the sagittal curves and establish a postural type: normal (physiological) back, hollow back (increased thoracic kyphosis and lumbar lordosis), fully rounded back (kyphosis extending down to the lumbar area), hollow-flat back (hyperlordosis of the lumbar spine with reduced kyphosis of the thoracic spine, common in small children), flat back (reduced kyphosis of the thoracic spine and lordosis of the lumbar spine; ■ Fig. 3.2).

**!** If the sagittal curves can be corrected by bending backwards or forwards, then postural variants are involved rather than (fixed) pathological changes. N.B.: beware of overdiagnosis and overtreatment!

We observe whether a ventral or dorsal overhang is present (■ Fig. 3.3) and the extent of the pelvic tilt (■ Fig. 3.4).



**■ Fig. 3.1a.** Not like this! **b** During examination in the standing position the patient's back should be at the eye level of the examiner, who should therefore be seated. Small children may need to stand on a box so that the iliac crest is at the examiner's eye level. The child must be undressed down to the underpants. The dignity of the child or adolescent must be preserved. Girls who have reached puberty should also be allowed to wear their brassiere. Otherwise, all items of clothing, including socks, should be removed.

A vertical line from the center of the shoulders should pass through the center of the ankle. The forward and downward pelvic tilt is approx. 30° in relation to the horizontal. A reduction in this tilt is an indication of lumbar kyphosis (e.g. in lumbar Scheuermann disease) or of spondylolisthesis.

In order to assess posture-related muscle performance, Matthiass has proposed the arm-raising test. The child is asked to stand as straight as possible and raise his arms and keep them in a horizontal position. He should try and maintain this position for 30 seconds. A child or adolescent with normal postural capacity is able to maintain this position, in contrast with a child with postural weakness (Fig. 3.5).

We now ask the child to bend down as far as possible while keeping the knees perfectly straight. We now measure the finger-floor distance (FFD; Fig. 3.6). Normally, children and adolescents should be able to touch the floor with their fingertips or even place the whole palm of their hand on the floor. If this is not possible, we measure the distance from the fingertips to the floor in

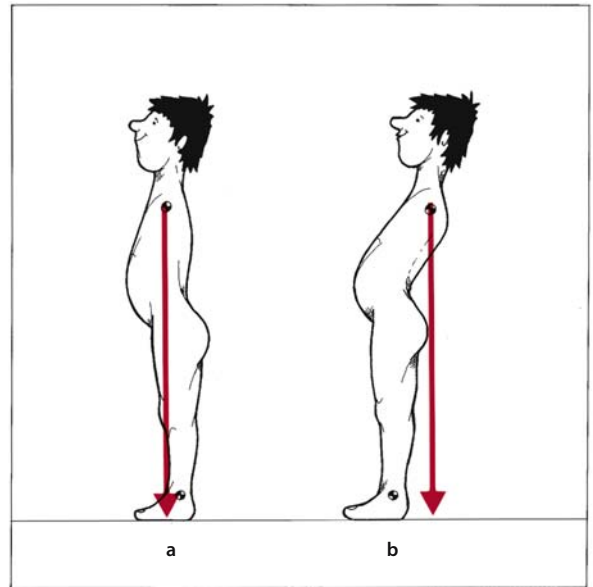


Fig. 3.3a. Ventral and b dorsal overhang: A vertical line from the center of the shoulders falls in front of or behind the center of the ankle

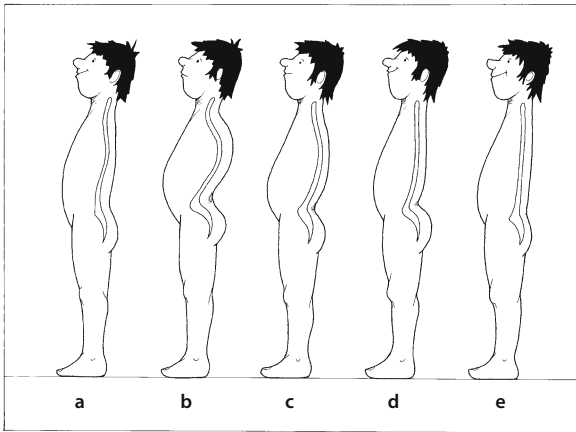


Fig. 3.2a–e. Postural types: a normal back, b hollow back, c rounded back, d hollow-flat back, e flat back

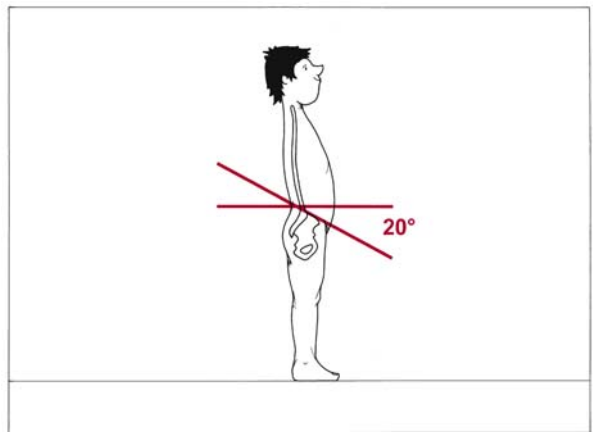


Fig. 3.4. Pelvic tilt: The forward and downward pelvic tilt in relation to the horizontal is normally approx. 20°–30°

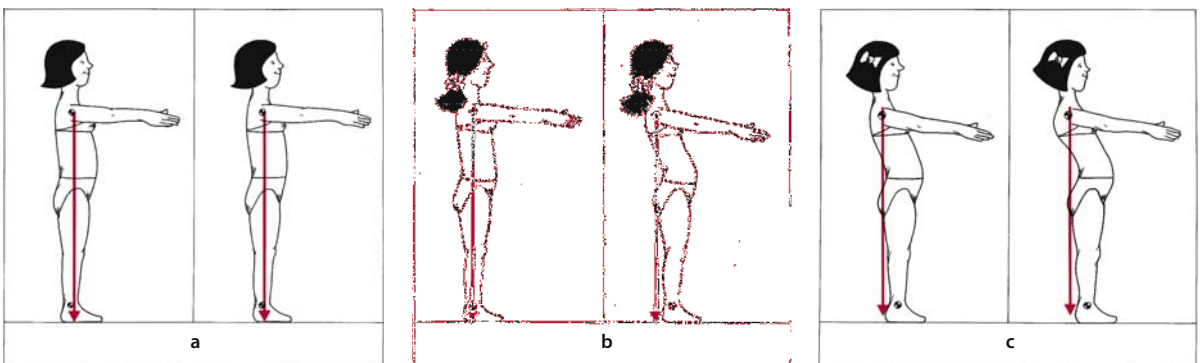
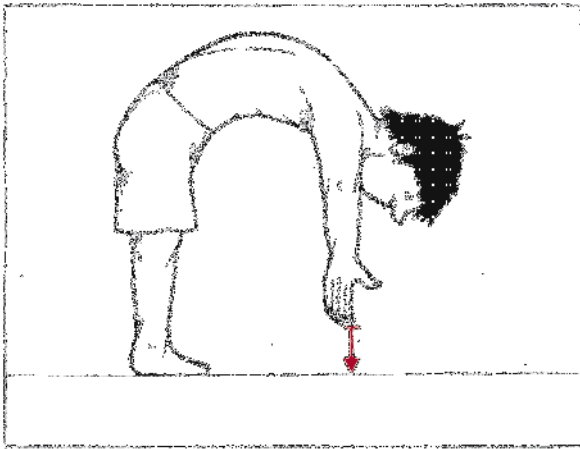


Fig. 3.5a–c. Arm-raising test according to Matthiass: The child is asked to stand as straight as possible and raise his arms and keep them in a horizontal position. He should try to maintain this position for 30 seconds. A child or adolescent with normal postural performance is able

to maintain this position (a), in the case of a postural weakness this posture is lost (b), while a child with extremely weak muscles cannot even adopt the upright posture (c)

## 3.1.1 · Examination of the back

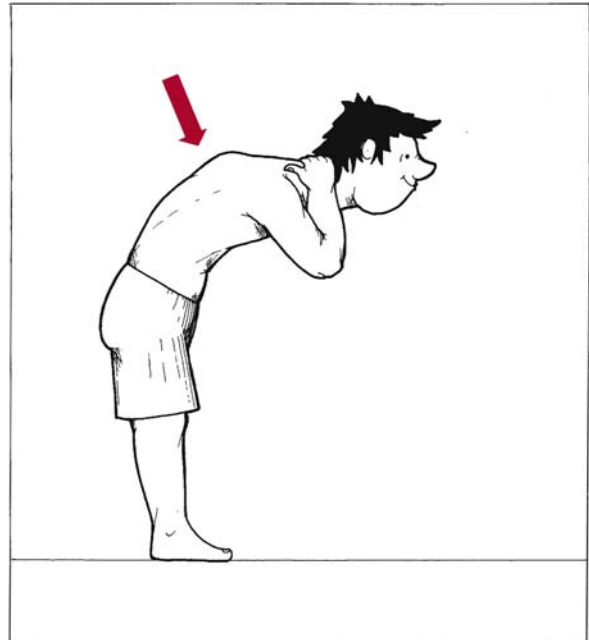


■ **Fig. 3.6.** *Finger-floor distance (FFD):* The patient bends down as far as possible without bending the knees. The distance between the floor and the fingertips is measured. Normal value = 0 cm

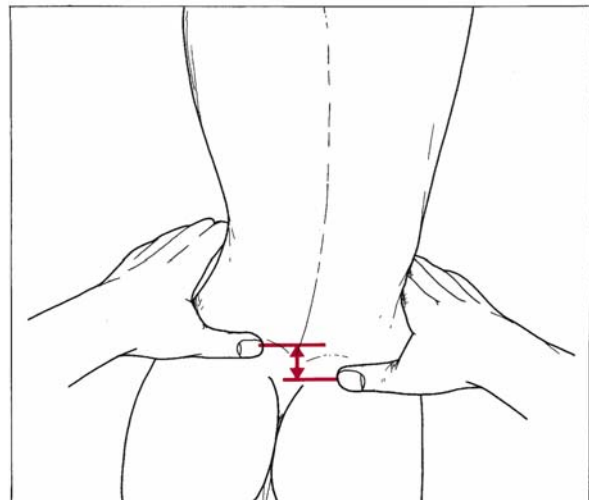
centimeters. However, this distance is less an indication of reduced mobility of the back than of contraction of the hamstrings. With the patient in a forward-bending position we observe whether the lumbar lordosis is corrected and whether the thoracic spine shows the right degree of kyphosis (correction of postural curvature in the case of a hollow or flat back). The patient is now asked to clasp his hands behind his neck (to prevent the shoulders from being pulled forward by the arms) and try to look up at the ceiling without changing the flexed position at the hip. Ideally, the patient is held in this position with a hand placed at the apex of the kyphosis and then asked to bend back (»look up at the ceiling«). We can then observe whether the thoracic kyphosis straightens out or whether a fixed kyphosis is present (e.g. as in a case of thoracic Scheuermann disease; ■ Fig. 3.7). If the latter is suspected, the condition of the pectoral muscles must also be assessed at the same time. To this end, the shoulders of the erect patient are pushed backwards by hand. If the pectoral muscle is contracted, the shoulder remains in front of the thoracic plane.

#### ■ Evaluation of the iliac crest

We place extended index fingers on both sides of the ilium and extend and abduct the thumbs at right angles, which then serve as pointers. We try to hold both thumbs horizontally (■ Fig. 3.8). If one iliac crest is lower than the other this will be reflected in the difference in the height of the thumbs. However, since it can be difficult to establish the precise difference, we place boards under the shorter leg until the iliac crests on both sides are at the same level and the two thumbs are likewise at the same height. The thickness of the boards corresponds to the leg length discrepancy in centimeters.



■ **Fig. 3.7.** *Straightening of the kyphosis:* While in a forward-bending position the patient clasps his hands behind his neck (to prevent the shoulders from being pulled forward by the arms) and tries to look up at the ceiling without changing this flexed position at the hip. Ideally, the patient is held in this position with a hand placed at the apex of the kyphosis and then asked to bend back (»look up at the ceiling«). We can then observe whether the thoracic kyphosis is straightened out or whether a fixed kyphosis is present



■ **Fig. 3.8.** *Height of the iliac crests:* Extended index fingers are positioned on both sides of the ilium. The thumbs are extended and abducted at right angles to serve as pointers. If one iliac crest is lower than the other this will be reflected in the difference in the height of the thumbs. Boards are placed under the shorter leg until the iliac crests on both sides are at the same level and the two thumbs are likewise at the same height.

❗ When measuring leg length indirectly it is extremely important to ensure that both the knee and hip joints are fully extended, unless this is rendered impossible because of flexion contractures.

#### Vertical alignment

A cord with a symmetrical weight is placed against the vertebra prominens, and we assess whether the weight is in line with the anal cleft or, if not, how many fingerwidths it deviates to the right or left (Fig. 3.9).

#### Examination of mobility

##### Examination of mobility from behind

We examine the maximum lateral inclination of the standing patient's spine from behind (Fig. 3.10). We observe whether the whole spinal column curves harmoniously to the side or whether individual segments are fixed and do not move with the rest of the spine (indication of fixed scoliosis). The pelvis must be fixed in order to evaluate trunk rotation. The rotation of the shoulder girdle in relation to the frontal plane is measured in degrees and is best observed from above (Fig. 3.11).

The patient is now asked to bend forward until the thoracic spine forms the horizon. The symmetry of the thorax is assessed. Protrusion of the rib cage on one side is termed a rib prominence. Using a protractor (or – if available – a scoliometer or inclinometer) we measure the angle between the rib prominence and the horizontal (the latter can be determined parallel to a door or window frame in the examination room; Fig. 3.12).

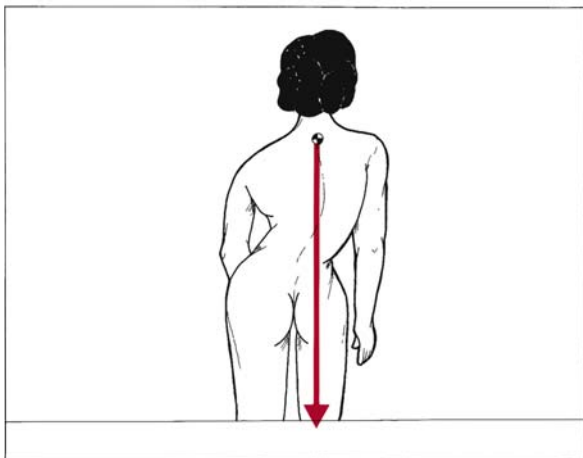


Fig. 3.9. Vertical alignment: A cord with a symmetrical weight is placed against the vertebra prominens and checked to see whether it is in line with the anal cleft or how many fingerwidths it deviates to the right or left

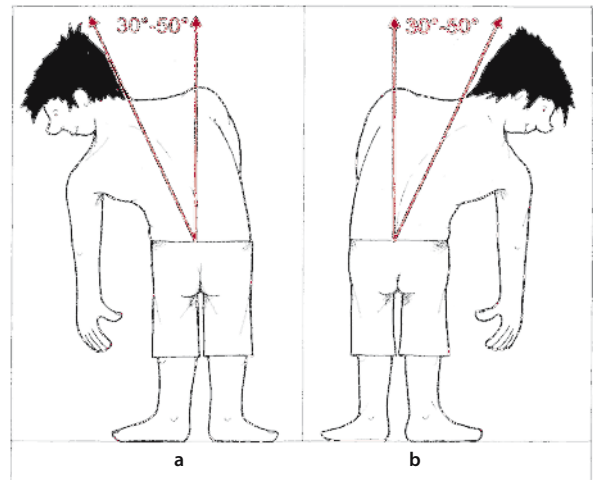


Fig. 3.10a, b. Lateral inclination of the trunk: The angle between the vertical and maximum lateral inclination of the spine is estimated in degrees from behind the standing patient (normal value: 30° – 50°). We observe whether the whole spinal column bends harmoniously to the side or whether individual segments are fixed and do not move with the rest of the spine

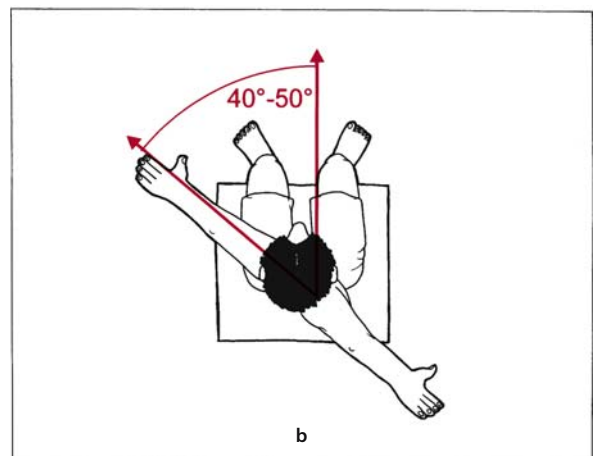
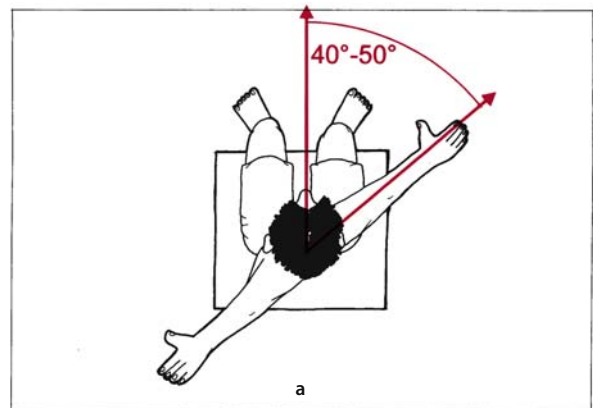
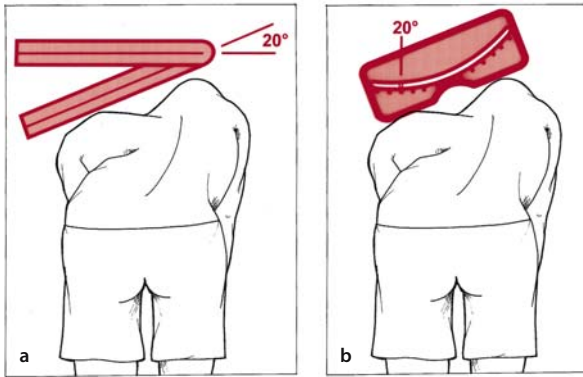


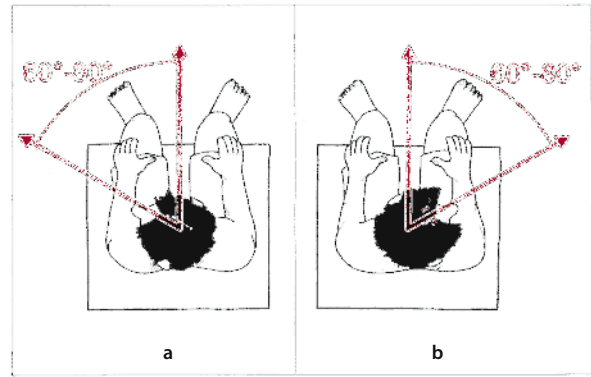
Fig. 3.11a, b. Rotation of the trunk: With the pelvis fixed, the rotation of the shoulder girdle in relation to the frontal plane is measured in degrees and is best observed from above. Normal value: 40° – 50°



## 3.1.1 · Examination of the back



■ **Fig. 3.12a, b.** *Measurement of rib prominence:* The patient bends forward until the thoracic spine forms the horizon. **a** With a protractor, the angle between the horizontal (i.e. parallel to the door or window frame) and the surface of the back is measured. **b** A simpler and more accurate measurement is obtained with an inclinometer with integrated spirit level and a notch in the center to avoid any distortion of the measurement caused by the projecting spinous process



■ **Fig. 3.13a, b.** *Head rotation:* Head rotation to both sides is measured from above with the patient in a sitting position. The rotation is stated in degrees measured from the midline. It can be measured actively (by asking the patient to turn his head) or passively (by holding the sides of the head with both hands and turning to either side). Normal value:  $60^\circ - 80^\circ$ . Observe any tensing of the sternocleidomastoid muscle at the same time

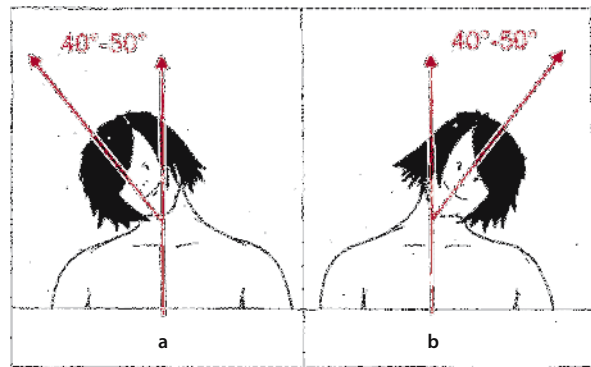
A rib prominence of more than  $2^\circ$  together with a horizontal pelvis is a reliable indication of a fixed rotation of the vertebral bodies. A rib prominence of  $5^\circ$  or more represents a serious case of scoliosis and requires radiographic investigation. The patient is now asked to continue bending forward until the lumbar spine forms the horizon so that we can then identify any lumbar prominence. Here, too, it is important that the pelvis is horizontal. If one leg is shorter than the other, the leg length discrepancy must be corrected using a board of appropriate thickness. The lumbar prominence is also measured with a protractor. An angle of  $5^\circ$  or more requires x-ray examination.

#### — Examination of the mobility of the cervical spine

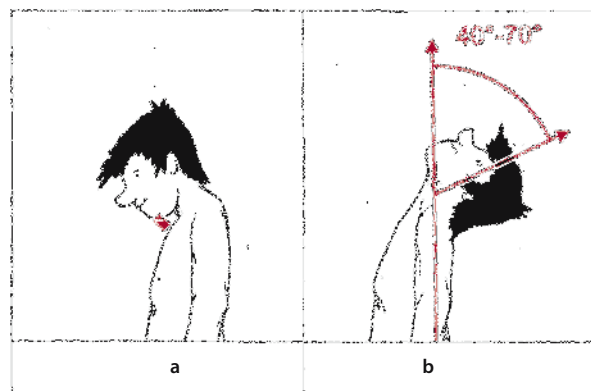
The head rotation to both sides is ideally measured from above with the patient in a sitting position (■ Fig. 3.13). The rotation can be actively (ask the patient to turn his head) or passively (hold the sides of the head with both hands and turn to either side). We can also observe any tensing of the sternocleidomastoid muscle during this maneuver. If a contracture due to muscular (congenital) torticollis is present, the muscle tenses on the side of the rotation movement.

We then check lateral inclination (■ Fig. 3.14), which can also be measured actively or passively. Here, too, the tensing of the sternocleidomastoid muscle is observed. If contracture is present, the muscle tenses when the head is inclined to the opposite side.

Finally, inclination and reclination are examined. With the head inclined forward the chin-sternum distance is measured. The patient then bends his head back and the angle with the axis of the body is estimated (■ Fig. 3.15).



■ **Fig. 3.14a, b.** *Lateral inclination of the head:* This can be measured actively or passively. The deviation from the midline is stated in degrees. Normal value:  $40^\circ - 50^\circ$ . Observe any tensing of the sternocleidomastoid muscle at the same time



■ **Fig. 3.15a.** *Inclination of the head:* The chin-sternum distance is measured (in centimeters or fingerwidths; normal value: 0 cm). **b** *Reclination:* Estimate the angle in relation to the axis of the body in degrees. Normal value:  $40^\circ - 60^\circ$

### — Schober measurement

The Schober test is used to determine the mobility of the spine in the sagittal plane and involves measurement of the stretching of the skin over the thoracic and lumbar spine. An initial mark is made over spinous process S1 and a second mark 10 cm above the first. The distance between these skin marks increases as the patient bends forward, reaching a maximum of 15–17 cm. Thoracic spine: A mark is made over spinous process C7, and a second mark is made 30 cm below this. As the patient bends forward the distance between the two increases by 2–3 cm (■ Fig. 3.16).

The maximum reclination of the spine is measured as shown in ■ Fig. 3.17. We observe whether the patient complains of pain around the lumbosacral junction (indication of spondylolysis).

### Palpation

We palpate the spinous processes and establish whether pain is elicited on pressure, percussion or vibration. To check pain on vibration we grasp the spinous processes between forefinger and thumb and move them back and forth. If the patient finds this painful, particularly around the lumbosacral junction, this is an important indication of possible spondylolysis.

We palpate the paravertebral muscles to assess whether these are strong, normal or weak, palpate any painful areas of muscle hardening (myogeloses) and check for tenderness over the muscle attachments. The transverse processes can also be felt by deep palpation.

During palpation, the skin moisture, temperature and elasticity of the skin are assessed and any dermatographic urticaria noted.

### — Heel-drop test

The patient is asked to stand on tiptoe and the examiner rests his hands on the patient's shoulders. The patient is now asked to drop onto his heels while the examiner simultaneously presses down on the shoulders. This maneuver will elicit any vibration-related pain in the spine caused by inflammation, tumors or herniated disks.

### — Iliosacral joints

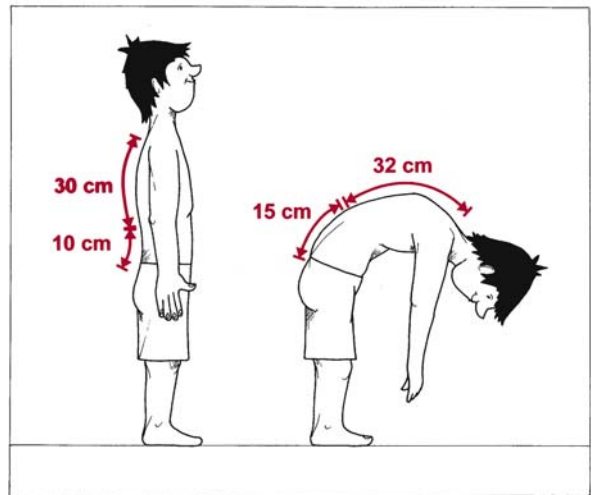
We check for pain on pressure or percussion and pain on compression from the side and sagittally. Mennell sign: In disorders of this joint, pain is elicited if the hip on the same side is overextended.

### Neurological examination

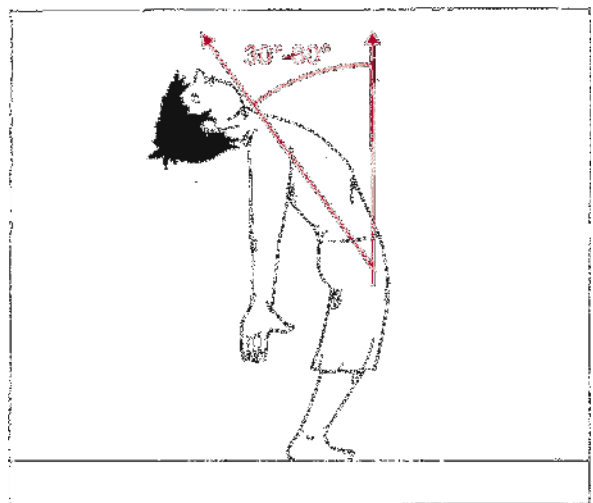
A complete examination of the back (■ Table 3.1) also includes at least a cursory investigation of the neurological status. A very rough (and quick) indication of a motor disorder can be obtained by checking the patient's ability to walk on tiptoes or on heels. The most important aspects of the neurological examination from the orthopaedic standpoint are described in ► chapter 2.1.2.

Brief overview of spinal status (e.g. in mass screening or if the child is being seen specifically for a back problem):

- Inspection from behind,
- Height of the iliac crests,
- Finger-floor distance,
- Rib prominence, lumbar prominence on forward bending?
- Walking on tiptoes and heels.



■ Fig. 3.16. *Schober sign.* Lumbar spine: Make an initial mark over spinous process S1 and a second mark 10 cm above this. The distance between these skin marks increases as the patient bends forward, reaching a maximum of 15–17 cm. Thoracic spine: A mark is made over spinous process C7, and a second mark is made 30 cm below this. As the patient bends forward the distance between the two increases by 2–3 cm



■ Fig. 3.17. *Reclination of the trunk:* The maximum reclination of the spine is measured as the angle between the upper body's vertical axis and the frontal plane. Normal value: 30°–60°

**Table 3.1.** Examination protocol for the back

Examination position	Examination	Questions
I. Walking	Movement pattern? Limping?	Ataxia? Neurological lesion?
II. Standing from behind	Position of the shoulders? Scapulae symmetrical? Spine straight? Iliac crests horizontal? Gluteal folds symmetrical? Waist triangles symmetrical? Plumbline in the center? Pigmentation over spinous processes? Hardening of paravertebral muscles? (if necessary examine on the lying patient as well) Pain on percussion/vibration of the vertebral bodies? (if necessary examine on the lying patient as well)	Scoliosis? Plexus paresis? Sprengel deformity? Winged scapula? Sprengel deformity? Scoliosis? Leg length discrepancy? Hip condition? Scoliosis? Severe scoliosis? Lumbar kyphosis? Myogelosis (muscle spasm)? Tumor? Infection? Spondylolysis?
III. Standing from the side	Shoulders pulled forward? Sagittal curves? Transition between front/back	Contracture of the pectoralis muscles? Scheuermann's disease? Contracture of psoas or hamstrings?
IV. Standing with flexed back from behind	Spine straight? Rib hump >5° Lumbar prominence >5°	Scoliosis? Thoracic scoliosis? Lumbar scoliosis?
From the side	FFD? Can the thoracic kyphosis be straightened out?	Contracture of hamstrings? Thoracic Scheuermann's disease?
V. Mobility	Lateral inclination of the head? Head rotation? Reclination/Inclination of head? Lumbar pain on reclination? (if necessary examine on the lying patient as well)	Torticollis? Torticollis? Klippel-Feil syndrome? Spondylolysis?

### 3.1.2 Radiography of the spine

The following standard spinal x-rays are recorded:

— *Cervical spine, AP and lateral:*

The patient can either stand or lie down for the AP x-ray of the cervical spine. The central x-ray beam is targeted on the 4th cervical vertebra (at the level of the Adam's apple) and is inclined towards the head at an angle of 15°–20°. (■ Fig. 3.18 left). For the lateral x-ray, the patient can either stand, sit or lie down, and hold his head up straight in a neutral position. The central beam is targeted horizontally on C4 (chin height; ■ Fig. 3.18 right).

— *Transbuccal x-ray of the dens:*

For the specialist dens x-ray the patient is placed on his back with the head in the neutral position. With the patient's mouth opened as wide as possible, the central beam is vertically aligned with the center of the open mouth (■ Fig. 3.19a). While the x-ray is recorded, the patient is asked to say »ah«, causing the tongue to press against the floor of the mouth thereby preventing its shadow from being projected onto vertebral bodies C1 and C2. The dens, axis, lateral masses

of the atlas and the atlantoaxial joints will be clearly visible on the resulting x-ray.

— *Functional x-rays of the cervical spine from the side during maximum inclination and reclination:*

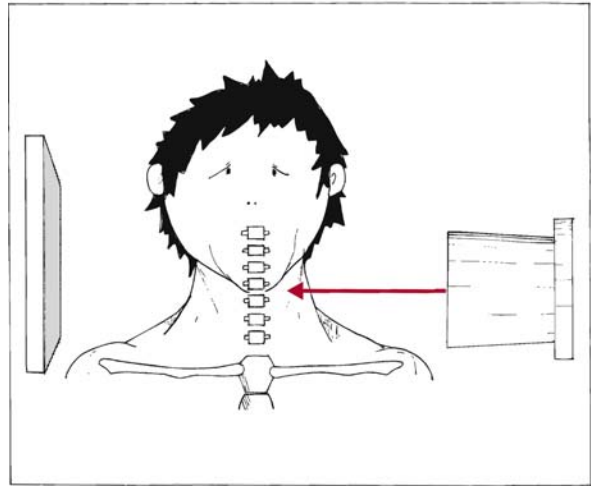
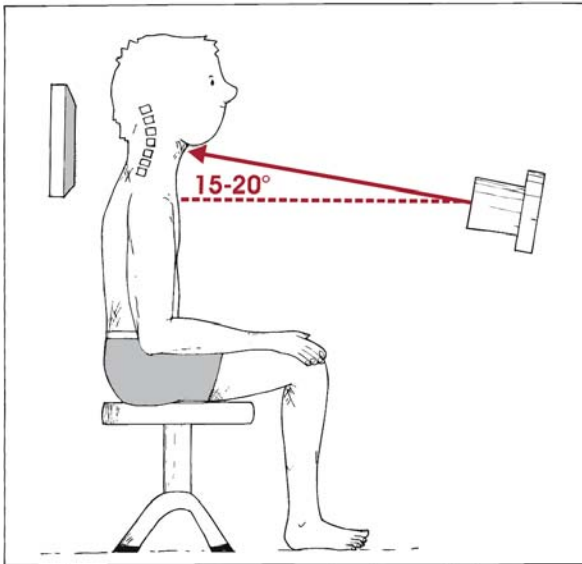
If instability or a ligamentous injury is suspected, the cervical spine is x-rayed (on the awake patient) from the side, while the patient is sitting up and during maximum inclination and reclination (■ Fig. 3.19b).

— *Thoracic spine, AP and lateral:*

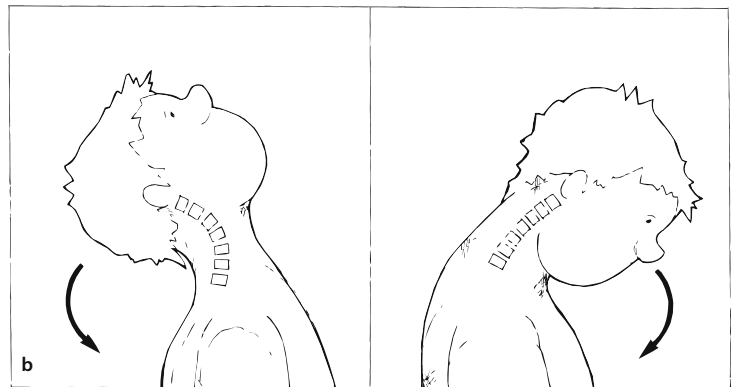
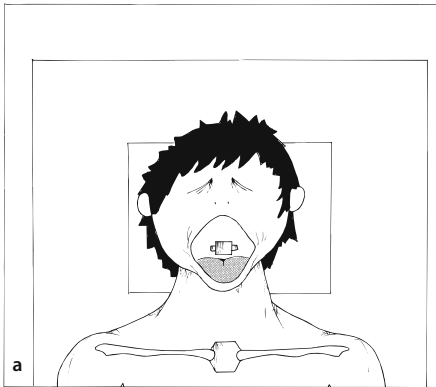
The AP and lateral x-rays of the thoracic spine should, if possible, be recorded while the patient is standing. For the AP view, the central beam is targeted perpendicularly onto a point approx. 3 cm above the xiphoid process of the sternum. For the lateral x-ray of the thoracic spine, the patient is asked to raise his arms. The central beam is targeted horizontally at the level of the 6th thoracic vertebra and tilted towards the head at an angle of about 10°. The resulting x-ray shows the vertebral bodies and the intervertebral disks viewed from the side (■ Fig. 3.20).

— *Lumbar spine, AP and lateral*

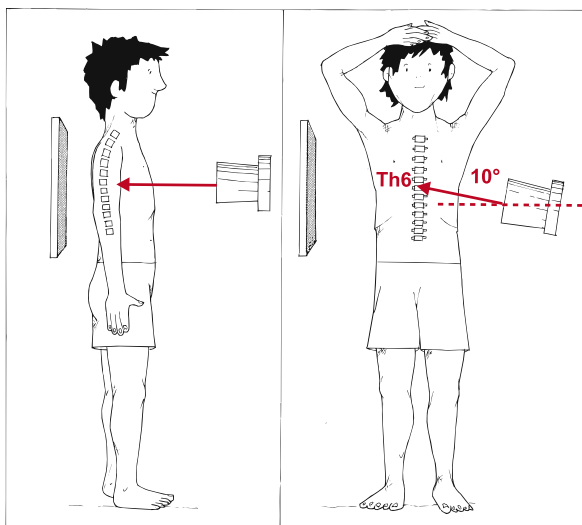
The AP and lateral x-rays of the lumbar spine should likewise be recorded while the patient is standing. For



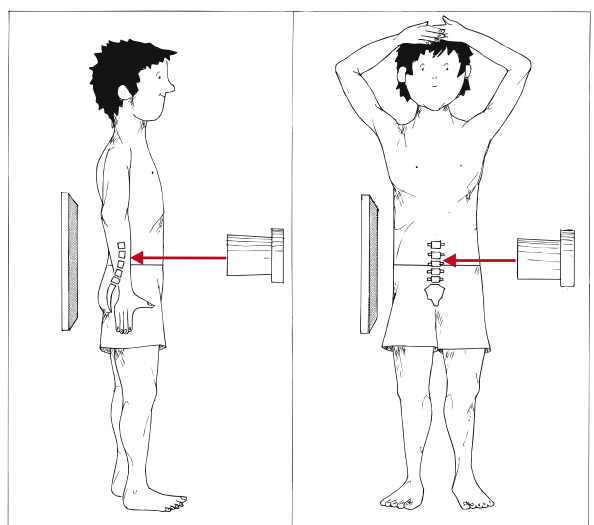
■ Fig. 3.18. Recording lateral and AP x-rays of the cervical spine. (after [1])



■ Fig. 3.19a, b. Recording cervical spine x-rays. a Radiographic technique for the transbuccal view of the dens, b Functional lateral x-rays of the cervical spine in maximum reclination (left) and inclination (right)



■ Fig. 3.20. Recording thoracic spine x-rays, lateral (left) and AP (right). (after [1])



■ Fig. 3.21. Recording lumbar spine x-rays, lateral (left) and AP (right). (after [1])

### 3.1.2 · Radiography of the spine

the AP view, the central beam points perpendicularly, at the level of the iliac crests, onto the center of the abdomen. For the lateral x-ray, the central beam is targeted on L3 at the patient's waist level (■ Fig. 3.21). In adolescents, wide cassettes should be used so that the iliac crest is included in the x-ray (so that the remaining growth potential can be assessed).

■ **Thoracolumbar junction, lateral:**

For this x-ray the central beam is targeted on T12.

■ **Lumbosacral junction, lateral:**

For this x-ray the lateral beam path is centered on L5.

■ **Oblique x-rays of the lumbosacral junction:**

For the oblique x-rays of the lumbar spine, the patient lies on his side on the examination table and then turns 45° to the right so that the small vertebral joints on the right are viewed (similarly, raising the left side will enable the facet joints on the left to be viewed). The central beam is targeted vertically onto the center of L3 (■ Fig. 3.22). See ■ Fig. 3.68 and 3.69 for examples and explanations.

■ **Whole spine, AP and lateral:**

With children and younger adolescents it is possible to depict the whole spine on a single normal cassette. The central beam points to T12. If deformities are present, this overview is more useful for evaluating the statics of the spine than individual images of the thoracic and lumbar spine. Here, too, wide cassettes should be used so that the iliac crest is included in the x-ray. For full-grown patients the spine must be x-rayed using combined films in special cassettes. Since the distance from the x-ray tube is considerable, this not only has an adverse effect on image quality, but also involves a high dose of radioactivity. We only record such x-rays in exceptional cases.

■ **CT of the spine:**

CT is extremely useful in fractures for revealing fragments in the spinal canal. They are also effective for identifying intraosseous tumors.

■ **Myelo-CT:**

Myelo-CT has largely superseded the conventional myelogram when it comes to viewing any impediment in the spinal canal resulting from a neurological lesion.

■ **Angiogram:**

Angiograms can be recorded conventionally, as MR angiograms or, using a more recent technique, as CT angiograms, which produce the best view of the blood vessels. Such images are required in certain tumors or for depicting the artery of Adamkiewicz prior to vertebrectomies.

■ **MRI of the spine:**

The MRI scan is used for cases of inflammation and tumors (primarily for the imaging of the soft tissue components) and for revealing intraspinal anomalies before scoliosis operations (particularly for congenital scolioses).

■ **Bone scan:**

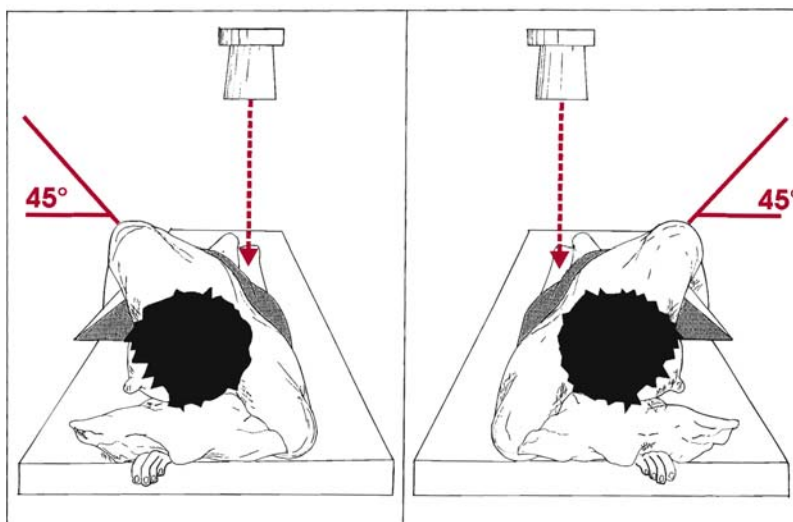
The technetium scan is useful for revealing small tumors that are not clearly depicted with conventional imaging techniques (e.g. osteoblastomas) or in the search for metastases.

■ **Ultrasound scans:**

Ultrasound scans are recorded in cases of a suspected spinal abscess or seroma.

### Reference

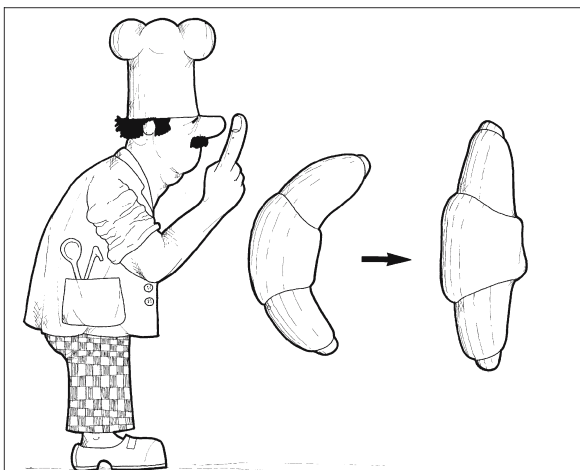
1. Greenspan (2003) Skelettradiologie. Urban & Fischer, Munich Jena



■ **Fig. 3.22.** Positioning of the patient and targeting of the central beam in oblique x-rays of the lumbosacral junction (after [1])



### 3.1.3 Can the »nut croissant«<sup>1</sup> be straightened out by admonitions? – or: To what extent is a bent back acceptable? – Postural problems in adolescents



» The body is the visible manifestation of the soul.  
(Christian Morgenstern, *Steps*) «

#### The back – a mirror of the soul?

Parents' concerns about the posture or the shape of the back of their offspring are one of the commonest reasons for a visit to the pediatrician or the orthopaedist. Their worries are essentially attributable to two main factors: On the one hand they are worried that a non-correctable deformity of the spine might result from the poor posture, as an expression of some sinister frame of mind. On the other hand, it is a generally known fact that back pain is one of the commonest conditions suffered in adulthood and one that might possibly be prevented by appropriate measures taken during childhood and adolescence.

But why are parents so worried about their child's appearance, particularly in relation to back problems, even though the back is usually covered by clothing and thus less exposed than, say, the face or the hands? – The back has special symbolic significance in linguistic usage and is, to a particularly great extent, the »visible manifestation of the soul«, as Christian Morgenstern puts it. A »good« posture for the spine is »upright«, just as a person's character can be described as »upright«. This also reflects the relationship between truth and dishonesty.

But terms associated with the back can also have other connotations. Politicians who adopt a particular standpoint and do not always change their opinion to match the prevailing mood are said to show »backbone«. But there are also others who are so thick-skinned that they can live without a backbone. Particularly strong-willed people are described as »unbending«. If their will is broken we say that it is »bent« to the will of another. People with a lot of problems are »weighed down by worries« until they eventually »collapse under the load«. Those who wish to ingratiate themselves with others »bow and scrape«. Those with huge debts are »laid low« and a person who refuses to take responsibility for his own mistakes and accept the consequences may try and »place all the blame on someone else's shoulders«.

So we can see how terms connected with the back and spine can also be used to describe emotion-provoking activities and properties that are closely related to a person's state of mind. Linguists are unable to explain whether the language actually creates this link between physical posture and mental outlook. We also find »crooked« characters in literature. Victor Hugo, in particular, made a hunchback the lead character in two of his works: Quasimodo in *Notre-Dame de Paris* and the court jester in *Le Roi s'amuse*. The latter play was used as the basis for Giuseppe Verdi's famous opera *Rigoletto*. And the French poet Paul Féval has a hunchback as the main character in *Le Bossu*. But in these literary examples the hunched back does not represent the manifestation of a sinister soul. Quite the opposite, since they are kind-hearted sensitive individuals who have been disadvantaged by nature and brutally exploited by others because of their inability to defend themselves.

But while the body is indisputably an expression of the soul, the connections are much more multilayered and complex than suggested by the vernacular language. Viewed at a superficial level, nature can also be at variance with linguistic usage. Thus, parents always want their child to adopt as straight a posture as possible. But the drooping and loutish posture of the adolescent is precisely an expression of the desire not to »bend« to the will of his parents.

#### Economic significance of back pain

Lumbar back pain is one of the commonest conditions suffered by adults and the number one reason for lost productivity. Thus, according to one epidemiological study, 66% of employees stated that they had suffered back pain in the previous 12 months [5]. And even a group of individuals in their twenties (Swiss recruits and soldiers) showed a prevalence of 69% for lumbar back pain [7]. An American study showed that 11% of men and 9.5% of women visited a general practitioner because of lumbar back pain [3]. In the USA, the loss of earnings is estimated

<sup>1</sup> Nut croissant: term used in Switzerland for a croissant filled with nuts. The expression »nut croissant figure« is commonly used in Switzerland to refer to a particularly drooping, kyphotic posture.

### 3.1.3 · Can the »nut croissant« be straightened out by admonitions?

at around 10 billion dollars [8]. In Switzerland, too, back pain is the second commonest cause of disability, after accidents. A high prevalence of lumbar back pain, at 48.2%, has been reported for industrial workers in Russia [9], indicating that back pain is not a specialty of the West, although it is clearly a much more serious problem in industrial nations than in the developing world. The significance of back pain evidently tends to parallel the degree of industrialization.

In Oman, the demand for back treatment has risen dramatically since the oil boom [2], a finding that is also of major economic significance. According to a Canadian statistical survey, approximately 30% of the total amount paid in 1981 as compensation for loss of earnings in the form of disability pensions was paid to back patients [1]. The pain frequently starts at a young age, and around half of adolescents complain of occasional back pain [10] (► Chapter 3.1.15).

For all of the reasons outlined above, it is perfectly understandable that parents are worried about what could happen to their children's backs in future.

#### **Evolution of upright walking and posture**

Humans are unique among all living creatures in exhibiting an erect posture. While primates evidently developed the mechanism for maintaining the trunk in an upright position at a very early stage, only humans are capable of standing and walking upright on two legs for prolonged periods. This species-specific bipedal, erect posture freed up the hands so that humans could use these for tasks other than locomotion. In fact, this discriminating use of the hand was probably the very first evolutionary step. A secondary consequence of the discovery that hands could be used not just for locomotion was the development of the brain and upright walking. The use of hands as tools and also the use of tools with the hands was therefore the first step in the evolution of man, some 5 million years ago, from primate to homo erectus, the precursor of today's homo sapiens.

This upright posture caused the eyes to be shifted forwards, thereby widening the field of vision and eventually producing binocular, stereoscopic vision. Compared to quadrupeds and the climbing anthropoid ape, humans have better visual, acoustic and tactile spatial orientation. From the phylogenetic standpoint, the adoption of an erect posture in humans did not simply involve a rotation of 90° at the hip, but primarily around the lumbosacral junction as a result of the cuneiform shape of the 5th lumbar and 1st sacral vertebrae. The sacrum is the resting point about which this erect posture is achieved.

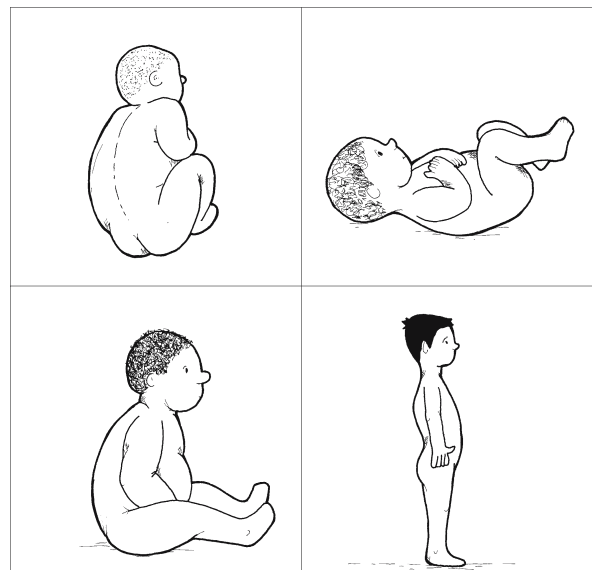
The development of the upright posture requires a specially-shaped spinal column. The double-S-shaped human spine differs from the single-S-shaped spine of the quadruped in its additional lumbar lordosis. Al-

though this lumbar lordosis is not absolutely essential for an upright posture, it came about primarily for functional reasons. The S shape of the spine is the optimal design for the corresponding dynamic loads. The cervical and lumbar lordosis, and also the thoracic kyphosis, act like linked elastic springs. Any major deviations from these functionally-adapted curves in the spine are mechanically inappropriate and result in adverse loading conditions.

The upright posture also has implications for other organs as well as the spine. Thus the iliac wing in humans is much wider than in quadrupeds, since it has to help carry the internal organs. The detorsion of the femoral neck during growth is another phenomenon specific to humans. In fact, humans have paid very dearly for this unique advantage of an upright posture and have evidently not yet completely come to terms with this evolutionary step. Man's unique erect posture not only contributes to his special dominant role in nature, at the same time it has become a direct potential disease factor whose implications cannot yet be fully grasped.

#### **Postural development in children**

The phylogenetic development of the back is imitated during maturation from the fetus to the child and then from the child to the adult. In the uterus, the fetus is in a flexed position and the spinal column is completely kyphotic. The neonate also holds the shoulders, elbows, hips and knees in flexion, causing the spine, apart from the cervical section, to be held in kyphosis, as is also the case



Postural development from the fetus, via the infant and toddler, to the child

with quadrupeds. Flexion contractures of up to 30° are physiological. At a later stage, the neck, back and femoral extensors are the first to be strengthened, providing the infant with head control. After a few months the baby is also capable of sitting up, albeit with total kyphosis of the back. At this stage the lumbar lordosis is still lacking, which is a physiological finding during this period before the start of walking.

Once the baby starts walking, the lumbar lordosis itself starts to develop. But this process does not fully parallel the strengthening of the muscles, and a hyperlordosis usually forms at this stage as a result of gravity acting on the ventral side. In toddlers this hyperlordosis is often not compensated by a hyperkyphosis of the thoracic spine, resulting in the scenario of the »hollow back«. This type of posture in the toddler is characterized by the physiological weakness of the muscles and the general laxity of the ligaments that is typical of the constitution at this stage. The toddler's back shape only develops into the adult shape shortly before puberty, although this shape is still dependent on the state of the muscles. In the elderly, the spine again resembles the kyphotic picture of the infant (■ Fig. 3.23).

An important characteristic feature of the infant is the asymmetrical tonic neck reflex. The persistence of this reflex can lead to an asymmetrical development of the muscles and the condition known as *resolving infantile scoliosis*. Resolving infantile scoliosis is a single arch-shaped curvature of the whole spine resulting from the asymmetrical tone of the muscles. The curvature is associated with little rotation and occurs with a left- or right-sided convex curve with equal frequency. If the child is

held by the head and feet, the opposite side can be made to curve. Resolving infantile scoliosis used to be much more common in the past, and is rarely encountered nowadays. This is possibly attributable to the trend (after 1970) of placing the infant in the prone position. More recently (since approximately 1992), the prone position is being abandoned following the discovery that sudden infant death syndrome occurs more frequently in the prone position than the supine position. We have, however, not seen an increase in resolving infantile scoliosis since then. Therefore there must be other etiological factors (e.g. genetic intermixture?).

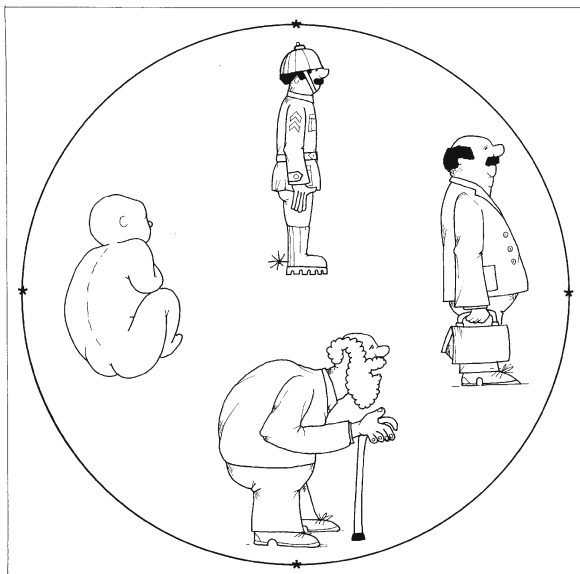
The prognosis for resolving infantile scoliosis is very good, as almost all of these curvatures disappear during the first year of life. This did not always used to be the case. Some cases of apparently resolving infantile scoliosis persisted and developed into progressive idiopathic infantile scoliosis, a condition that used to be particularly common in Great Britain [6]. The observation that the difference between the angle made by the ribs and the spine when seen from the side is greater in the progressive forms than in cases that spontaneously resolve themselves means that the progressive forms can be detected at an early stage (► Chapter 3.1.4).

The condition of progressive infantile scoliosis has almost disappeared even in Scotland, where the condition was particularly common. While the progressive form of the disease has an extremely poor prognosis, resolving infantile scoliosis is not associated with any long-term sequelae. It is completely unrelated to idiopathic adolescent scoliosis, and patients with a history of resolving infantile scoliosis show no increased risk of developing idiopathic adolescent scoliosis in later life.

### Postural types in the adolescent

Posture is influenced by the following factors:

- *The shape of the bony skeleton*  
The shape is determined by genetic factors (the mother: »His father has exactly the same crooked back«). The position of the sacrum, which in turn is dependent on the pelvic tilt, also plays an important role. The steeper the sacrum, the less pronounced the sagittal curvatures (lordosis and kyphosis).
- *Ligamentous apparatus*  
Posture can be active or passive. If our muscles are not activated, then we simply »hang« from our ligaments. Such a posture can best be adopted by overstretching the hips, sticking out the tummy, positioning the lumbar spine in hyperlordosis and tilting the upper body backward to offset the forward shifting of the center of gravity. If the center of gravity is shifted forward or backward we talk of a ventral or dorsal overhang (► Chapter 3.1.1). This posture cannot be adopted passively, however, since it is unstable and must be compensated for by muscle activity.



■ Fig. 3.23. Postural cycle (the old man returns to the kyphotic posture of the fetus)

### 3.1.3 · Can the »nut croissant« be straightened out by admonitions?

#### — Muscles

The state of the muscles has a considerable influence on our posture. Strong muscles with good tone can maintain an actively erect posture throughout the day. The condition of the muscles depends partly on constitutional factors and partly on the training status. But one other factor needs to be taken into account in relation to the growing body: The muscles, together with the skeleton, undergo substantial length growth but are unable to increase in width to the same extent. Consequently, a certain muscle weakness is physiological in the growing child. Only on completion of the growth phase can the »muscle corset« be trained and built up in the optimal way. Postural insufficiency is frequently associated with an intoeing gait and reduced hip flexion [4].

#### — Pelvic tilt

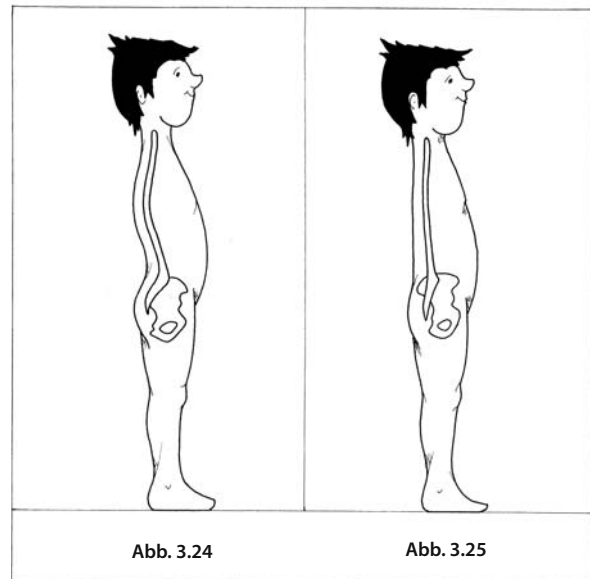
The pelvic tilt is closely related to the steepness of the sacrum. Straightening the pelvis reduces the lumbar lordosis and thus the thoracic kyphosis as well (■ Fig. 3.24, 3.25).

#### — Influence of the psyche

Posture is not a constant anatomical feature of an individual. Apart from constitutional factors, posture represents a snapshot that depends not only on muscular activity but, to a very great extent, on psychological status. As previously mentioned, linguistic usage also highlights this link. A state of mind characterized by joy, happiness, success, self-confidence, trust and optimism tends to affect the erect posture and the associated efficient postural pattern. By contrast, worries, conflicts, depression, failures and feelings of inferiority produce precisely the opposite effect and promote poor postural patterns.

Another special factor comes into play in adolescents: Puberty is a stage of life marked by internal conflicts associated with finding one's own personality. Since an important element in this process is the loosening of the bond with the parents, a certain protesting posture in respect of the parents can be considered physiological.

Since a straight posture is usually considered the ideal by parents, the internal protest against the parents' world manifests itself in the form of an – often ostentatiously – poor posture (particularly while sitting). The poor posture resulting from the physiological muscle weakness of the growing body is further emphasized by »casual« sitting. The more frequently the parents admonish their child with »sit up straight«, the quicker he or she resumes the »nut croissant« position. It is striking to observe how children with a very pronounced kyphotic posture are very frequently withdrawn and have one very dominating parent. When such adolescents are questioned about their symptoms or problems during the consultation, the



■ Fig. 3.24. Normal pelvic tilt with forward/caudal inclination of the pelvis by approx. 20°

■ Fig. 3.25. Cancellation of the pelvic tilt and consequent reduction of the lumbar lordosis and the thoracic kyphosis



Adolescents often deliberately adopt a seated posture that goes against their parents' ideas about good posture...

mother or father will constantly reply on their behalf. It is noticeable that the child is clearly overwhelmed by the mother or father.

But other problems can also cause adolescents to adopt a very kyphotic posture, e.g. if a female unconsciously tries to conceal her breasts by hunching her shoulders forward and folding her arms in front of her. Some girls are unable to accept the growth of

their own breasts. This is particularly apparent if the girl has a very dominant mother who herself has large breasts. But also a funnel or keeled chest can cause the girl to adopt a permanently kyphotic posture in the unconscious desire to conceal this part of her body.

#### — Social aspects

Not every social class or era has the same conception of the ideal posture. Since ancient times, statues and paintings have tended to present the ideal of an upright posture. In European royal dynasties, a stiff posture was often promoted by constraining the individual in a brace. But the social notions of the ideal posture have changed since then, and the ideals of the modern age are frequently characterized by a markedly »casual« posture.

As already mentioned, posture represents a »snapshot«. Every individual can adopt a variety of postures.

The standing posture can be subdivided into the following stages (■ Fig. 3.26–3.28):

- habitual posture,
- passive posture,
- actively straightened posture.

We can also distinguish between *constitutional postural types* (normal back, hollow back, rounded back, flat back, hollow-flat back, ► chapter 3.1.1).

The classification of the first 4 back shapes dates back to the 19th century (Staffel 1889 [2]). These are physiological variants with essentially no pathological significance. We have added the 5th back shape since it is a relatively common physiological variant, particularly in children. Instead of a »normal back« perhaps we should rather refer

to a harmonious back. Using the term »normal back« can easily give the impression that the other back shapes are abnormal, which is certainly not the case by definition, since these are, after all, types of posture. We only speak of a pathological shape if there is fixed hyperkyphosis of the thoracic spine, a permanent absence of lumbar lordosis or even a kyphosis in this area. The investigation of the correctability or fixation of individual segments is described in ► chapter 3.1.1.

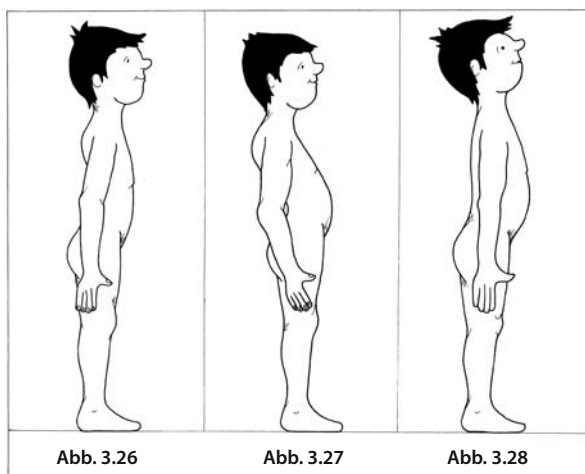
#### Pathological significance of poor posture

Whether »postural damage« actually exists is a matter of considerable dispute. Since back symptoms are common in adults and have also increased over the past few decades, the discussion of this subject is highly topical. Unfortunately there is a scarcity of scientifically-established hard facts and, on the other hand, widely diverging opinions based on subjective impressions. However, a number of factors in recent years have thrown some light on the subject.

Various widely-held traditional views first need to be corrected somewhat:

- *The development of structural scoliosis has nothing to do with posture.* A poor posture cannot induce idiopathic adolescent scoliosis. Scoliosis is known to result from a discrepancy between the growth of the vertebral body anteriorly and the growth of the posterior elements, resulting primarily in lordosis. Adolescents with scoliosis are therefore conspicuously straight and erect, and also often very keen on sport. The lateral curvature develops as a result of the rotation of the vertebral bodies and has nothing to do with posture (► Chapter 3.1.4). A leg length discrepancy may possibly promote lumbar scoliosis. This is definitely the case with uncompensated differences of more than 2 cm. Whether it applies for differences of less than 2 cm is controversial, and it is possible that the leg length discrepancy only influences the direction of the scoliosis rather than its development.

- *Of the physiological postural types, apart from the harmonious posture, the hollow back has a much better prognosis than the flat back.* Although the flat back is the esthetic ideal, the future prospects in terms of subsequent symptoms are much worse for the flat back than for a back with markedly sagittal curves, given the poorer shock-absorbing properties of the former. Lumbar disk damage occurs more frequently with this back shape and is also often associated with pain. The problem arises primarily from the kyphosing of the lumbar spine. The lack of lordosis shifts the center of gravity forward, which means that the lumbar paravertebral muscles have to work harder to maintain posture. The kyphosing of the lumbar spine is also often very pronounced during sitting.



- Fig. 3.26. Habitual posture posture
- Fig. 3.27. Passive posture
- Fig. 3.28. Actively straightened posture



### 3.1.3 · Can the »nut croissant« be straightened out by admonitions?

— *The development of a fixed kyphosis can be influenced by posture.* A permanent kyphotic posture can trigger Scheuermann disease during puberty. Although the prognosis in terms of symptoms is not bad in Scheuermann disease involving the thoracic region, it becomes increasingly worse the further down one goes, and lumbar Scheuermann disease is associated with a very high risk of subsequent chronic lumbar back pain. Usually the condition results in elimination of the lumbar lordosis, or even kyphosing in this area. This is extremely undesirable from the mechanical standpoint because of the forward-shifting of the center of gravity. It has to be offset by lordosing of the thoracic spine and considerable postural work by the paravertebral muscles in the lumbar area. The shock-absorbing properties of this type of spine are also poor.

#### Therapeutic options

Of the factors that determine posture, we can influence two in particular:

- the status of the muscles,
- possibly the psychological factors.

All other parameters are given and we have no way of influencing them.

As regards the *muscles*, we should always bear in mind that a certain amount of physiological muscle weakness is associated with growth.

**! Muscles can only be strengthened by activity. Such activity must be undertaken by the child or adolescent and cannot be imbued into the child from the outside. Consequently, the crucial factor in determining whether activity takes place or not is the child's motivation. The surest way of demotivating the child is to compel it to undertake an activity against its will.**

Since physical therapy is not an attractive type of activity, it is pointless to prescribe months, or even years, of physical therapy, at the expense of health insurance funds, when the child is not remotely motivated. The outcome will be a complete lack of any effect on the muscles. Equally questionable in my view are the »postural physical education lessons« provided in many schools. Since all students attending such lessons are labeled as those with »poor posture« the participants are stigmatized from the outset. Since it is self evident that such lessons are unlikely to motivate the students to keep active, it would be much more useful to encourage the adolescent to exercise within the context of a sport that affords him or her a certain amount of pleasure. Although the type of sport selected is not ultimately important, activities in which the arms are also used are preferable. Swimming is best, of course, although other ball-based sports such as

baseball, basketball or volleyball are extremely beneficial. Sports that exercise the muscles on one side of the body, e.g. tennis, are also perfectly appropriate since, as already mentioned, there is no need to worry at all about the possibility of scoliosis developing as a result of the unilateral muscle tension. Even scoliosis patients should be allowed to play tennis. The important thing is the pleasure gained from the sport. Passive and non-athletic children do not like taking part in ball-based sports because they invariably lose. However, perhaps such children can be motivated to take up swimming or possibly attend a fitness center on a regular basis. This avoids the problem of their having to constantly measure themselves against their peers.

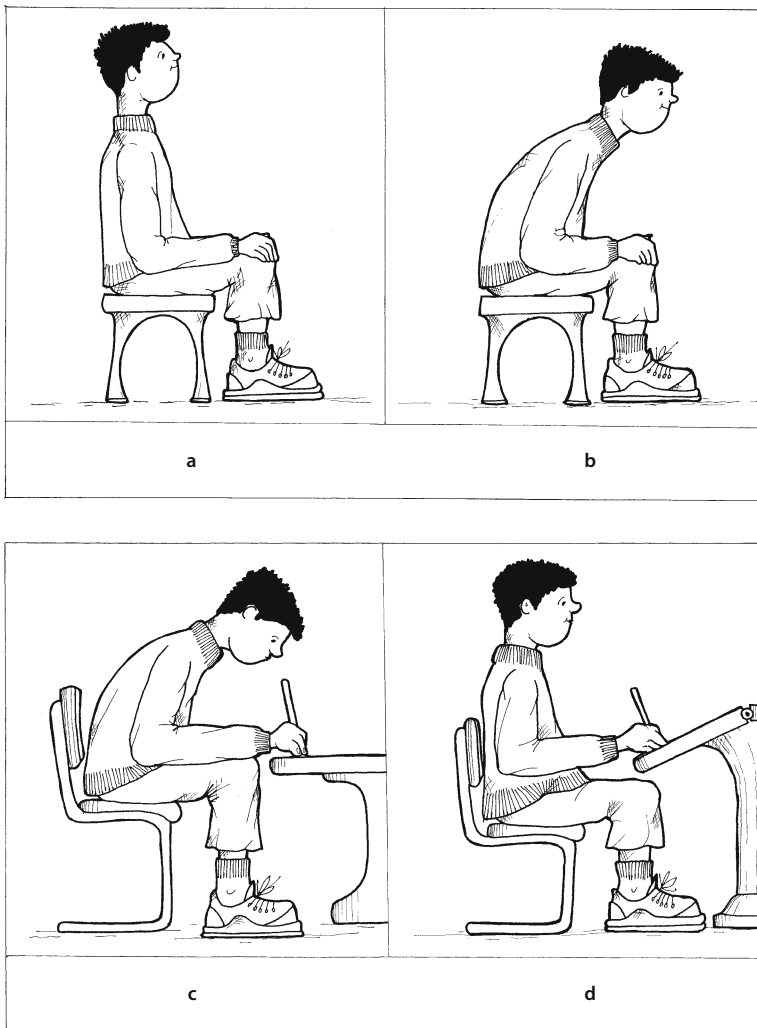
One particular factor that promotes passivity is the considerable amount of time spent sitting at school or in the home. The lumbar spine tends to kyphose during passive sitting. Certain useful measures can be taken to counter this tendency, even though these are implemented in only a very small proportion of schools: An inclined writing surface reduces the kyphosing of the lumbar spine during writing; the writing surface should be positioned sufficiently high; a ball chair also promotes lordotic sitting and stimulates the sitter to constantly perform slight compensation movements; a kneeling chair with support for the lower leg also promotes lordotic sitting (■ Fig. 3.29). Such aids promote a habitual lordotic sitting posture that produces positive effects in the long term.

In theory, *psychological factors* can also be influenced, although this is much more difficult. Since fixed hyperkyphosis of the thoracic spine is often indicative of a conflict between the adolescent and a parent, the doctor must proceed very cautiously. Psychological counseling can prove worthwhile on occasion however. Another potentially fruitful strategy in motivating the adolescent to take up sport is for him or her to meet other relevant individuals who could serve as new positive models. In most cases, however, it can be very difficult to explore often deep-seated conflicts, particularly since both sides (parents and child) frequently adopt a highly defensive attitude. What is certain, however, is that constant admonitions to sit up straight are counterproductive.

**! In other words, the question posed at the start, i.e. whether the »nut croissant« posture can be straightened out by cajoling, can be answered resoundingly in the negative. A permanent improvement in posture will only be achieved if the adolescent is motivated to take part in enjoyable activities.**

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■ Fig. 3.29a–e. Seated postures and sitting aids:

- a upright seated posture;
- b drooping seated posture;
- c kyphotic seated posture;
- d influence of writing height and slope of the writing surface on seated posture;
- e ball chair

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### 3.1.4 Idiopathic scolioses

» While her elegance in ballet may appeal, the risk of scoliosis is very real. «

#### ► Definition

Condition involving lateral bending of the spine of  $>10^\circ$  of unknown origin. There are two basic clinical pictures of scoliosis:

- A rare form in which the deformity starts as early as infancy or childhood (*infantile* or *juvenile scoliosis*). Boys and girls are equally affected by this type. Scolioses at the thoracic level frequently have their convexity to the left and are associated with kyphosis.
- The more common *adolescent form* starts during puberty. Girls are mainly affected and the thoracic form is always right convex. This type of scoliosis is usually associated with lordosis.

### Classification

Classification by age at onset (according to the American Scoliosis Research Society):

- Infantile: 0–3 years
- Juvenile: 4–10 years
- Adolescent over 10 years old

Because juvenile scolioses are extremely rare (and do not behave according to a typical pattern), the *British Scoliosis Society* classifies only two entities:

- Early onset: 0–7 years
- Late onset over 7 years old

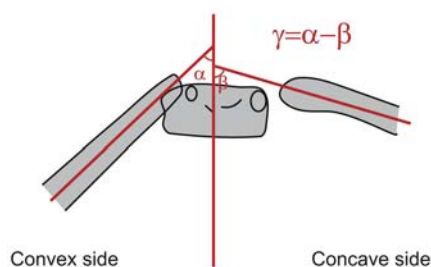
The condition known as *resolving infantile scoliosis* is not classed as an idiopathic scoliosis but is a special type of scoliotic posture. However, since it can progress to infantile idiopathic scoliosis it is discussed here.

#### Resolving infantile scoliosis

Resolving infantile scoliosis occurs at the age of a few months, but has become relatively rare in the west as a result of the frequent use of the prone position. Resolving infantile scoliosis is characterized by a long, usually left-convex, thoracolumbar, C-shaped arch with little rotation. The rib vertebral angle difference (RVAD) according to Mehta [68] is measured to distinguish it from progressive infantile scoliosis (see Fig. 3.30). The prognosis is good and a spontaneous recovery can be expected in over 96% of cases. Isolated cases can progress to infantile idiopathic scoliosis.

#### Infantile (early onset) scoliosis

This rare type is located in the thoracic area in 98% of cases and occurs 1.5 times more frequently in boys than in girls. In 76% of cases the scoliosis is left convex and often associated with a kyphosis. In the infant, an rib vertebral angle difference according to Mehta of more than 20° [68] indicates that the condition is not the benign resolving



■ Fig. 3.30. RVAD according to Mehta [64]. In scolioses in the infant, the angle between a vertical line passing through the vertebral body and the axis of the rib is measured on both the convex and concave sides. If the difference ( $\gamma$ ) between the two angles is 20° or more, the scoliosis is very likely to be the progressive form rather than a spontaneously correcting scoliosis

infantile scoliosis, but rather a progressive form of infantile idiopathic scoliosis (■ Fig. 3.30). The characteristic features of infantile scoliosis differ from those of the adolescent form to such an extent that it can clearly be considered as a different disease. The prognosis for infantile scoliosis is very poor. Despite brace treatment, it will often undergo substantial progression, resulting in the need for surgery even at an early age in many cases.

#### Juvenile scoliosis

If the scoliosis occurs between the ages of 4 and 10, the juvenile form is considered to be present. Girls are only slightly more frequently affected than boys. In addition to the thoracic location, lumbar and S-shaped curves also occur. The prognosis is poor. Only 5% of scolioses are non-progressive, while the rest increase annually by 1–3° up to aged 10, and by 5–10° a year during the pubertal growth spurt [88].

#### Adolescent (late onset) idiopathic scoliosis

This is by far the commonest form of scoliosis and is characterized by the following features:

- It is usually located at the *thoracic* level and almost without exception involves a *right-convex* curve.
- It occurs less commonly at the thoracolumbar and lumbar levels, and such cases show a marked tendency to go out of alignment. Sometimes these scolioses are not truly idiopathic but occur secondarily to leg length discrepancies or a lumbosacral junction anomaly.
- In around 10% of cases, adolescent scoliosis is *S-shaped*, i.e. there are 2 primary curves: Since the lumbar curve is usually more rotated than the thoracic curve, S-shaped scolioses are less conspicuous in cosmetic respects than *C-shaped* thoracic scolioses of the same severity.
- It is almost always associated with relative *lordosis* (for the thoracic level, an overall kyphotic angle of less than 20° is considered to be relative lordosis).
- It always involves *rotation*, whereby the posterior parts of the vertebral bodies are always *rotated* towards the concave side of the curve (if this is not the case then a structural idiopathic scoliosis is not present); for a given degree of curvature, the rotation is always more pronounced at the lumbar level than the thoracic level.
- Adolescent scoliosis probably develops as the result of a disparity between the *growth* of the posterior and anterior vertebral body sections; the diminished growth of the posterior sections forces the vertebral bodies to deviate laterally and to rotate. Instead of a scoliosis, one might describe this as a *rotational lordosis*.

### Etiology

By definition, the cause of idiopathic scoliosis is unknown. In adolescent scoliosis there is a disparity between the growth of the vertebral bodies anteriorly and that of the posterior elements. The vertebral bodies grow faster than the posterior elements, resulting primarily in a lordosis. The diminished dorsal growth impedes the ventrally located vertebral bodies from increasing in height, forcing them to become distorted, i.e. rotate, in order to create space for themselves. This produces a *rotational lordosis*. This principle is illustrated in Fig. 3.31 and 3.32. The idea of looking at in this way dates back to Somerville, and many more recent studies have confirmed this theory [32, 59, 78, 93]. Lordosis is almost always present in adolescent scoliosis, even when the spine appears kyphotic on the x-ray in a particular projection. The cause of the lordosis is unknown. The diminished growth occurs in the area of the spinal canal [78]. It is conceivable that the spinal cord is protecting itself against the stretching stimulus of growth. Several investigations in recent years have reported the existence of *intraspinal anomalies* or *neurological problems* in a certain proportion of »idiopathic« scolioses. MRI studies have shown an intraspinal syrinx in 8% of typical idiopathic thoracic adolescent scolioses [23, 91]. Such findings seem to occur much more frequently in atypical (i.e. not right-convex and thoracic) scolioses [77]. Other investigators have found pathological somatosensory potentials in over 50 percent of cases of idiopathic adolescent scoliosis [66]. The side on which the scoliotic convexity occurs does not appear to be relevant to such findings. Nor is »handedness« responsible for the direction of the lateral

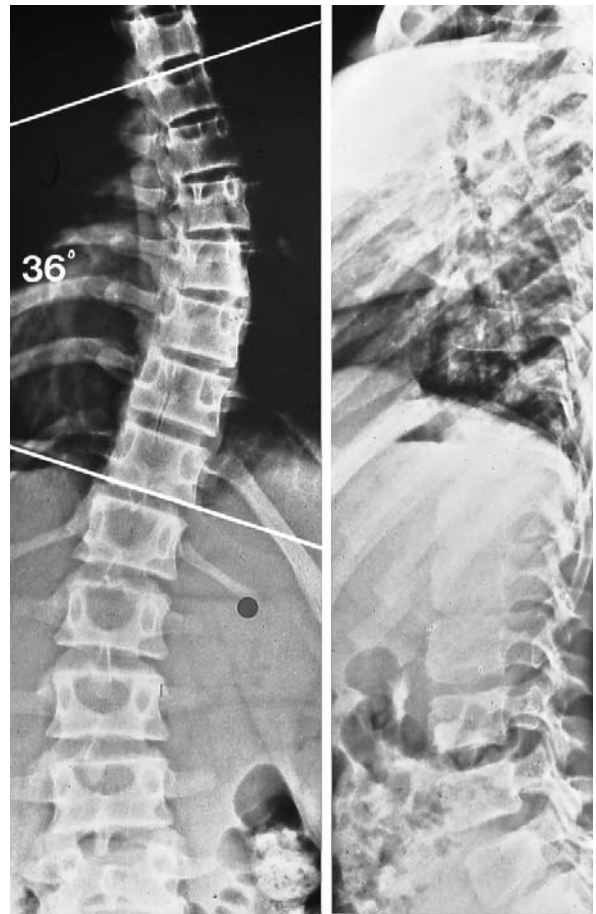


Fig. 3.32. Anteroposterior and lateral x-ray of an idiopathic thoracic adolescent scoliosis in a 13-year old girl. Note the pronounced *thoracic lordosis*

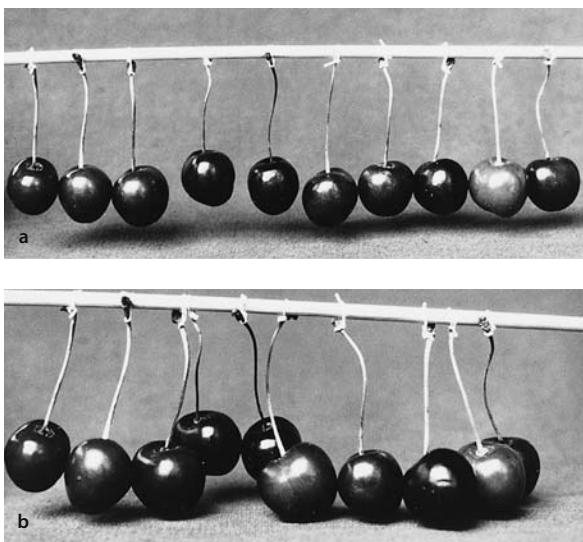
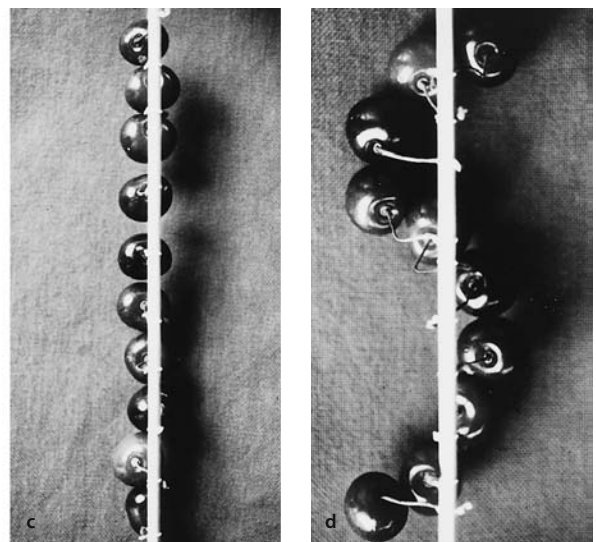


Fig. 3.31a–d. Principle of the development of idiopathic adolescent scoliosis. The cherries and the stems represent the vertebral bodies and vertebral arches respectively. They are suspended next to each other



from a rod (a, c). If the gap between the stems (i.e. the dorsal elements) is reduced, the cherries make space for themselves by rotating (b, d). The lateral curvature is the ultimate consequence of this rotation



### 3.1.4 · Idiopathic scolioses

curvature. The fact that idiopathic thoracic adolescent scoliosis tends to have a right-sided convex curve is rather attributable to the site of the mediastinal organs. Since the condition is caused not by the asymmetry of the muscles but rather by a (symmetrical) problem in the sagittal plane, the distorted side is essentially dependent on the anatomical configuration. As confirmation of this theory we have found a *left* convex thoracic scoliosis in 2 patients with situs inversus. Thoracic scolioses that are not right convex must therefore be investigated by MRI for the possibility of intraspinal anomalies before surgery (we now routinely arrange an MRI scan of the spine before every scoliosis operation).

The causes of early onset scoliosis likewise remain unknown. Some cases of juvenile scoliosis show a progression similar to that of adolescent scoliosis, while others, primarily cases with a very early onset, behave differently – as described above. *Neurogenic factors* probably play a certain role in terms of the etiology. Following the introduction of the practice of placing the infant in the prone position this formerly common condition has now become very rare. Since the 1990's, however, pediatricians have been advising parents to place their babies on their back again for the first 6 months because of the increasing incidence of sudden infant death syndrome observed for the prone position. However, since resolving infantile scoliosis remains extremely rare, the prone position cannot be considered solely responsible for the decline of this disease. Increased genetic intermixing may play a positive role in this respect. Recent studies involving MRI scans have shown that the proportion of intraspinal anomalies is very high in this patient group [29].

#### Other discussed etiological factors

- **Growth:** A premature pubertal growth spurt has been observed in scoliosis patients. Elevated levels of somatotrophic hormone have also been measured [33, 62], and patients with scoliosis are taller than normal adolescents of the same age [56, 111].
- **Osteoporosis:** Reduced bone metabolism was measured in one study of scoliosis patients undergoing iliac crest biopsy [11]. Measurements of bone mineral density have shown that osteopenia may be an important risk factor in curve progression [46].
- **Genetics:** Scoliosis occurs more frequently in patients with a family history of the condition. Whereas the normal incidence is approx. 1.8%, frequencies of 7–11% have been observed in first-degree relatives, and there is evidence of the involvement of a dominant gene on the x-chromosome [49].
- **Leg length discrepancies:** While there is no doubt that clinically relevant pelvic obliquity can promote the development of a scoliosis [100, 101], it is not clear at what point a leg length discrepancy becomes meaningful. Leg length discrepancies up to 1 cm

are extremely common and are of no relevance to the formation of scoliosis. Differences of 2 cm and more can, if not corrected, lead to structural scoliosis, specifically a lumbar scoliosis in which the convexity points towards the side of the shorter leg. However, the correlation between leg length discrepancy and scoliosis is not very close [44, 100], nor is the clinical measurement of leg length discrepancy very reliable.

- Several studies have investigated whether an anomaly or *asymmetry of the muscles* is present [48]. In most cases, biopsies were taken from scoliosis patients at operation and the muscle samples were examined histologically and histochemically. All authors found an increased proportion of type 1 muscle fibers on the convex side of the scoliosis compared to the concave side. It is generally agreed, however, that these are *secondary changes* and do not involve a primary asymmetry of the muscles. A corresponding investigation *before* the onset of scoliosis has not been implemented for obvious reasons.
- Scoliosis does not occur spontaneously in animals, but has been induced in *animal experiments* by a wide variety of manipulations, e.g. by interfering with the circulation or growth [20]. Unfortunately, none of these experiments has provided any significant findings concerning the etiology of idiopathic scoliosis.

**! Asymmetrical sporting activity, frequent sitting or standing in a scoliotic position [10] and handedness are of no etiological significance.**

#### Incidence

Scolioses occur 3.5 times more frequently in girls than in boys. At the age of 14 years, 1.2% of adolescents (1.9% of all girls) have a scoliosis of more than 10°, while 0.5% of this overall group (i.e. 0.8% of the females) have a scoliosis exceeding 20° [9]. Scoliosis appears to be more common in the white population than in other ethnic groups [82]. The incidence of scoliosis has remained fairly constant over the past few decades [70].

#### Clinical features, diagnosis

##### Clinical examination

The onset of the menarche is a particularly important factor in a girl's medical history since it occurs at the height of the pubertal growth spurt. A period of relatively strong growth (5–10 cm a year) can be expected for around 2 years from this point. Although growth will still not be complete by the end of this period, this is no longer particularly relevant as regards the progression of scoliosis. In boys, on the other hand, there is no corresponding sign of sexual maturation that can be established as reliably as the menarche in girls.

The most important clinical examination is the forward-bending test. The examiner sits behind the patient



so that the iliac crest is at eye level. The patient is then asked to bend forward. Any unilateral bulging of the rib cage («rib hump») at the thoracic level or of a «lumbar prominence» at the lumbar level now becomes apparent. If one of the patient's legs is shorter than the other, it is important during this examination to equalize the leg lengths by placing a board under the shorter leg (► Chapter 3.1.1).

A special instrument, the «inclinometer» (► Chapter 3.1.1), is used to produce an objective measurement of the extent of rib and lumbar prominences [8], although a sufficiently accurate measurement can also be obtained with a protractor (► Chapter 3.1.1).

❗ **A clinically relevant rib hump or lumbar prominence is considered to exist if the angle is 5° or more.**

The following relationships were calculated in one study [54]:

- Thoracic Cobb angle  
= (rib hump angle x 1.64) + 6.3
- Lumbar Cobb angle  
= (lumbar prominence angle x 1.58) + 7.6

The average error of the Cobb angle calculated by this method is just ±5.5°.

We also consider the *vertical alignment*. A plumbline suspended from the vertebra prominens must pass exactly through the anal cleft otherwise decompensation is considered to be present. With the patient in *lateral inclination* we observe whether the curvature of the spine is harmonious or whether an abnormally fixed position is present. We also establish whether the *waist triangles* are symmetrical. Examination from the side allows us to determine the presence of harmonious *sagittal curves*, relative thoracic lordosis (which is extremely common in idiopathic thoracic adolescent scoliosis; ► Fig. 3.33), hyperkyphosis or any cancellation of the lumbar lordosis (► Chapter 3.1.1).

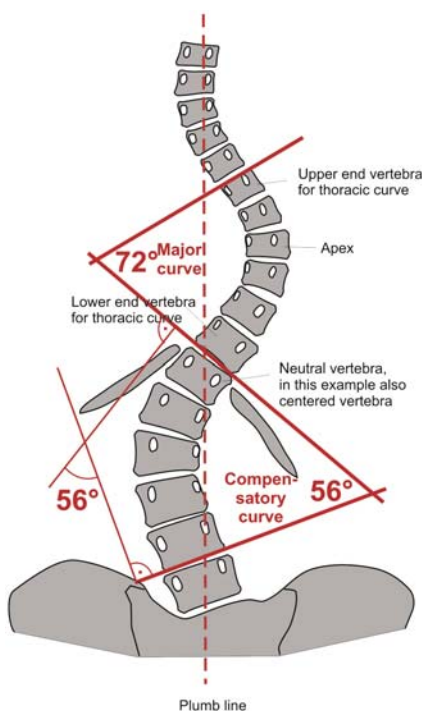
### X-rays

AP and lateral x-rays of the full thoracic and lumbar spine are required for a proper assessment of any scoliosis. The following *measurements* can be taken from the resulting images:

- On the AP x-rays we measure the *extent* of the primary scoliotic curve and that of the compensatory secondary curve. The method specified by *Cobb* is ideal for recording this measurement (► Fig. 3.34) [13] (*Cobb angle*). In this method two lines are drawn through the upper and lower endplates that are most severely tilted towards each other. The angle between these two lines (or the corresponding perpendicular lines) is the scoliosis angle. These two vertebral bodies are termed *end vertebrae*. Other measuring methods have failed to catch on. The *neutral vertebra* is the



► Fig. 3.33. 13-year old patient with *idiopathic adolescent scoliosis*. In addition to the asymmetry of the waist triangles and the projecting scapula, note the clearly visible thoracic lordosis

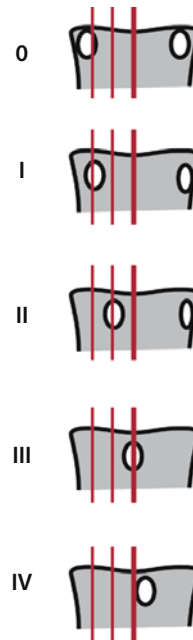


► Fig. 3.34. *Measurement of the Cobb angle with highlighted characteristic elements of scoliosis* [13]: Two lines are drawn through the upper and lower endplates that are most severely tilted towards each other. The corresponding vertebral bodies are termed «end vertebrae». The angle between these two lines (or the corresponding perpendicular lines) is the scoliosis angle. The more pronounced and less pronounced curves are respectively termed the primary and secondary curves. The «neutral vertebra» is the vertebral body between the primary and secondary curves that is least rotated. Centered vertebrae are the vertebral bodies whose center is in vertical alignment with the center of the sacrum. The apex of a curve is located at the level of the vertebral body with the greatest lateral deviation

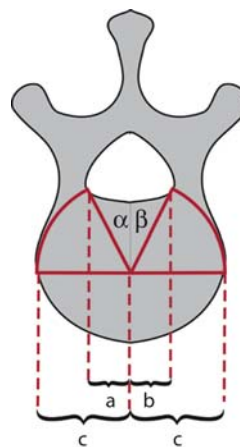
### 3.1.4 · Idiopathic scolioses

neutrally rotated vertebra and is not usually the same as the end vertebra, but located 1–2 segments beyond the apex. The *centered vertebra* is the vertebral body below the apex of the (thoracic) scoliosis that is in vertical alignment with the center of the sacrum.

- The *rotation* of the vertebral bodies can be estimated or measured. An approximation can be obtained by the evaluation method according to *Nash and Moe* [73]. To this end, the apical vertebral body on the AP x-ray is subdivided into 6 sections. The severity of the rotation can be estimated according to where the pedicle shadow is located on the convex side of the scoliosis (■ Fig. 3.35). The rotation can be determined more precisely by a mathematical method developed by ourselves [39], in which the rotation can be calculated in degrees from the width of the vertebral body and the distance between the pedicle and the edge of the vertebral body (■ Fig. 3.36). One elegant option is to use the rotation measurement template according to Perdriolle [76, 84] (■ Fig. 3.37). Although none of these methods is very accurate [84], they do provide an indication in routine clinical practice of the extent of the rotation.
- The extent of thoracic kyphosis and lumbar lordosis can also be measured in a similar manner on the *lateral x-rays* (▶ Chapter 3.1.5).
- The *skeletal age* is very important in assessing the prognosis. Since this can readily be evaluated if the iliac crest is visible on the AP x-rays, excessively narrow films should be avoided for the lumbar spine view. The extent of skeletal maturation can be evaluated according to the width of the ossified part of the iliac crest apophysis. The apophysis starts to ossify from the lateral edge. Risser has subdivided maturation into 5 stages («*Risser sign*», ■ Fig. 3.38) [86]. The start of ossification («*Risser I*») occurs approximately 4 months after the menarche and the peak of the pubertal growth spurt [21]. The skeletal sign of the height of pubertal growth is the closure of the elbow apophyses. From this point onward, a further 2 years or so of continuing spinal growth can be expected (overall approx. 5–7 cm), and no further growth of the extremities occurs. The growth spurt is concluded in stage IV and only minimal growth takes place until definitive ossification (stage V) is achieved. The spine can continue to grow until the age of 20, although the additional length is only 1–2 cm at this time. A *hand plate* must be prepared in order to be able to determine the skeletal age more accurately.
- *Functional x-rays* (with AP projection) with maximal lateral inclination to the right and left show the correctability of the primary and secondary curves (■ Fig. 3.39). The most reproducible results can be obtained by bending the spinal area to be investigated over a padded roll [63].



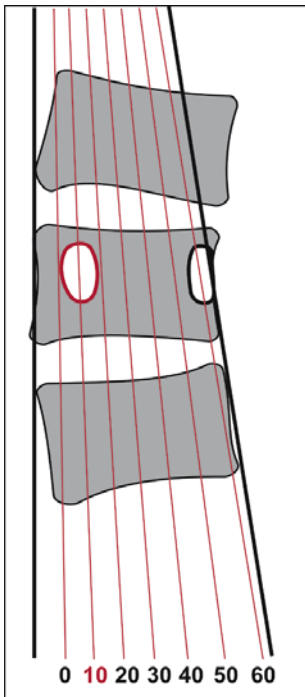
■ Fig. 3.35. *Estimation of rotation according to Nash and Moe* [69]: The apical vertebral body on the AP x-ray is subdivided into 6 sections. The severity of the rotation can be estimated according to where the pedicle shadow is located on the convex side of the scoliosis. *I* corresponds to a rotation of approx. 5°, *II* approx. 15°, *III* approx. 30° and *IV* approx. 40°



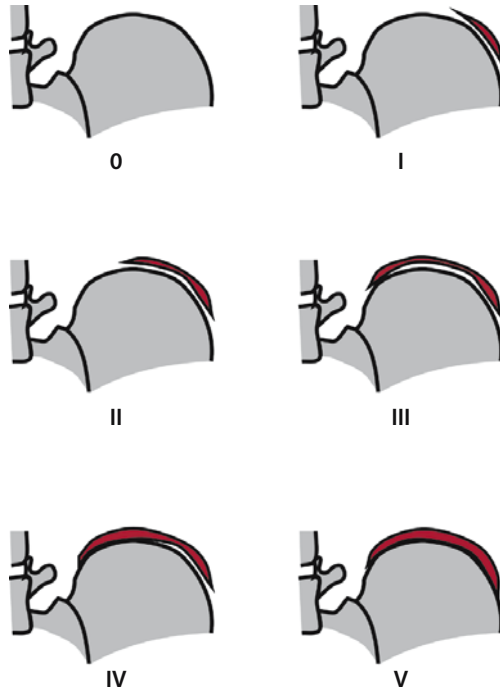
■ Fig. 3.36. *Calculation of rotation* [39]: If both pedicles of a vertebral body are visible on the AP x-ray, the rotation can be determined very precisely on the basis of the distance between the inner edge of the shadow and the center of the vertebra and the diameter (or radius) of the vertebral body according to the formula:  $Rotation\ angle = (a-b)/2$

#### Visual presentation of the surface of the back

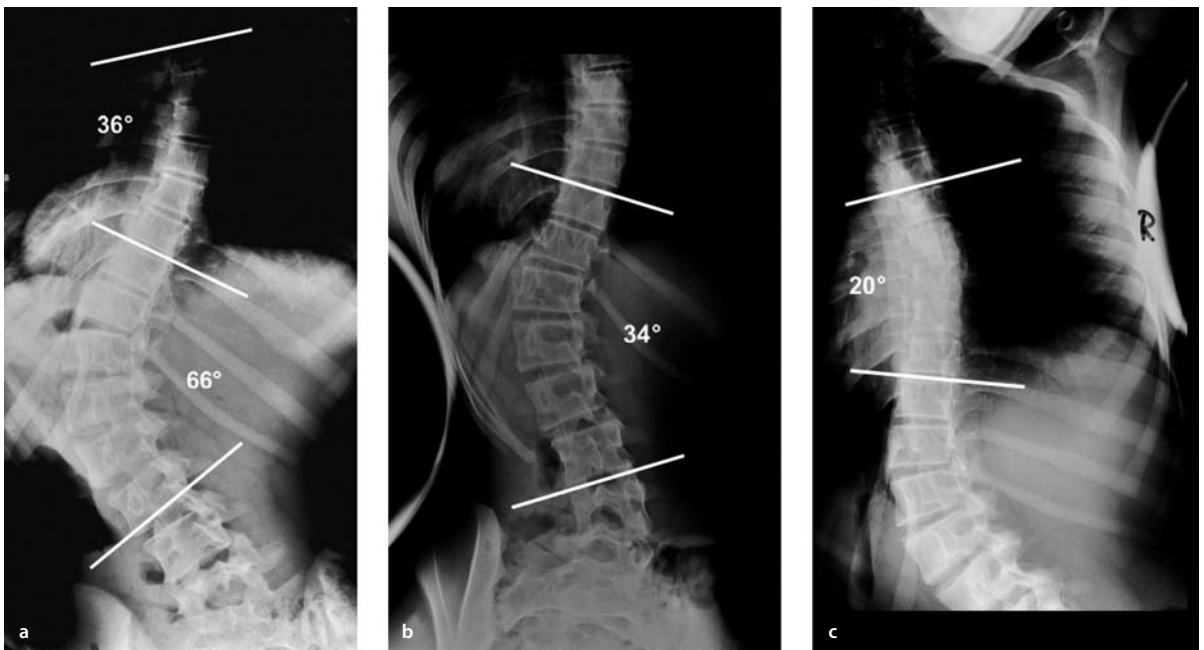
The need to document and measure the surface of the back arose mainly from the problem of radiation exposure during x-ray examination. Developments started with the introduction of moiré photogrammetry by Takasaki 1970 in Japan [97], which was followed by other new photographic and videographic techniques.



**Fig. 3.37.** Rotation measurement template according to Perdriolle [76]: This template on transparent film can be used to determine the rotation of a vertebral body on the AP x-ray. The template is placed over the vertebral body and aligned with the edges. The extent of the rotation is read off the scale via the line that passes through the center of the pedicle on the convex side of the scoliosis (shown at the bottom as an angle between 0° and 60°)



**Fig. 3.38.** Risser sign [87]: The stage of skeletal maturation (0–V) can be evaluated according to the ossification of the iliac crest apophysis. The ossification starts on the lateral side at the peak of the pubertal growth spurt (roughly contemporaneously with the menarche in girls) (Risser stage I). The pubertal growth spurt is concluded with Risser stage IV, and ossification of the apophysis (stage V) takes a further 2 years to complete



**Fig. 3.39.** Functional x-rays with maximal lateral inclination to the left (b) and right (c) are needed to evaluate the correctability of a scoliosis (a). For correction purposes, we bend the spinal section to be investigated over a roll (visible in b left and c right), to obtain

as reproducible results as possible [63]. In a lumbar scoliosis, the correction of the lumbar curve with the VDS instrumentation may not go beyond the straightening of the thoracic countercurve in the functional x-ray

### Moiré topography

Expressed in rather simplified terms, this technique involves the projection of focused light through a grating with parallel opaque lines of a defined width spaced apart at specified intervals. On a three-dimensional object behind the grating, the light is blocked in places depending on the distance from the grating (and also depending on the angle of the incident light). This produces shadows on the three-dimensional surface like the contours of a geographical map. The contours with corresponding depressions and projections are visually displayed on the surface of the back. Since the contours are so clearly displayed, the examiner can quickly assess whether the surface is symmetrical or asymmetrical. However, the resulting image is greatly dependent on the positioning of the patient. Nor is it easy to quantify the extent of the asymmetry.

### Other methods of surface measurement

Since it is difficult to assess the extent of any asymmetry using moiré curves, other methods for the three-dimensional calculation of the back surface have been developed. We ourselves have designed a technique in which dots of light are projected onto the surface of the back and the position of these dots in space is calculated by stereophotography [39]. A similar method using video cameras was developed in the UK («ISIS method» [98]). We used this method for 15 years.

A less posture-dependent technique of surface measurement is raster stereography [24]. A grid pattern is projected onto the surface of the back and a virtual »plaster cast« calculated. Conclusions about the skeletal geometry can be drawn by obtaining unchanging shape parameters and determining the concavity or convexity of the individual surface areas. Scolioses, kyphoses and lordoses can be evaluated very reliably by computer and any subsequent changes over time documented. This technique reduces the number of x-ray examinations required and is currently used in our scoliosis clinic.

### Additional imaging methods

#### Computer tomography

CT is suitable for checking rotation and has been used in a number of clinical studies. However, this method does not provide much more insight than a conventional x-ray given the lack of reference parameters. In particular, it is not possible to assess any rotation of the pelvis at the same time.

#### Magnetic resonance imaging (MRI)

MRI is a non-invasive, but expensive, method that can be used for identifying intraspinal problems (anomalies, tumors). MRI is indicated if neurological symptoms are present or if surgery is planned. It is not suitable for monitoring the progress of a condition as

the investigation must be performed with the patient lying down and, on the one hand, the layers do not match the plane of curvature while, on the other, the curved spinal column projects beyond the standard planes, making angle measurements almost impossible. The MRI is useful, however, for excluding intraspinal abnormalities [23].

### Natural history, prognosis

The diagnosis of idiopathic adolescent scoliosis in puberty then raises the question of the likelihood of the subsequent progression of the condition.

! In an investigation of over 700 patients, the following factors of relevance to prognosis were determined [60]:

- Cobb angle
- Age
- Risser stage
- Menarche.

No other factors, including for example the extent of rotation, lordosis, family history, sex, etc., showed any positive correlation with progression. The bar chart in Fig. 3.40 shows the risk of progression in relation to the initial angle and chronological age. An example of severe progression is shown in Fig. 3.41.

In a recent study the progression velocity of scoliosis was studied for 535 consecutive girls with untreated adolescent idiopathic scoliosis. The progression was most notable with a growth velocity of  $\geq 2$  cm/year, at ages between 9 and 13 years, bone ages between 9 and 14 years, Risser signs 0-1, and between 0.5-2 years before menarche [112].

The extent of rotation only has low prognostic significance. In boys, scolioses appear to progress at a later stage than in girls, even taking into account the skeletal age [51].

The prognosis for infantile and juvenile idiopathic scoliosis is much worse. If a scoliosis of  $>20^\circ$  persists beyond infancy, progression is inevitable.

### Progression in adulthood

In scolioses with a Cobb angle of *over*  $50^\circ$  on completion of growth, a further *progression of*  $0.5^\circ-1^\circ$  per year can be expected (this applies to thoracic and lumbar scolioses) [3, 102].

### Consequences of scoliosis

Thanks to investigations involving substantial numbers of patients and observation periods of up to 50 years we now know a great deal about the consequences of scolioses [3, 102, 103]. Overall, the degree of impairment in untreated scolioses is moderate after 50 years. Although the incidence of back pain is increased, this is rarely serious [103].



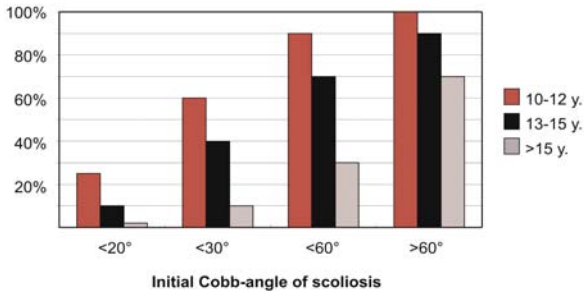


Fig. 3.40. Risk of progression in relation to age and the initial angle of the scoliosis. (after [60])

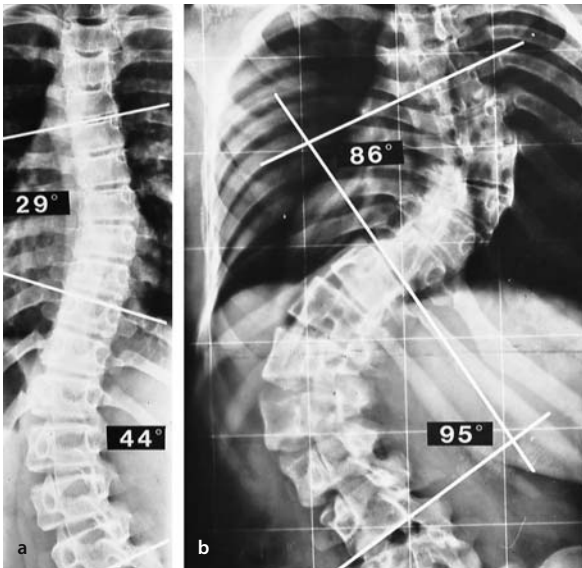


Fig. 3.41a, b. Example of rapid progression of a scoliosis in a female patient between the ages of (a) 11 years and (b) 17 years

### Reduction in life expectancy

In serious cases (particularly thoracic scolioses, from approx. 80° [103]) the impaired cardiopulmonary function can reduce life expectancy. Contributory factors are as follows:

- Volume restriction: this occurs both as a result of the deformity of the thorax and the relative lordosis and can lead to atelectases on the concave side and emphysema on the convex side.
- Thoracic rigidity: thorax fixed in the expiration position.
- Complications: chronic bronchitis, pneumonia, pleuritis.
- A direct link exists between the reduction in vital capacity and the scoliosis angle: for each 10° of scoliosis angle, the vital capacity is reduced by around 10%.

### Cosmetic impairment

An inconvenient rib hump, produced by rotation, occurs particularly in C-shaped thoracic scolioses, and starts to

become clearly visible from a rib prominence angle of 12° (corresponding to a Cobb angle of approx. 40°). In the lumbar area, the outwardly visible cosmetic impairment only starts to appear from a lumbar prominence angle of 15° (corresponding to a Cobb angle of approx. 60°), unless decompensation is present. Asymmetry of the waist, however, can have a more detrimental effect on the cosmetic appearance.

### Pain

The probability of back pain is increased in:

- lumbar and thoracolumbar scolioses [47], and in
- decompensating scolioses.

The probability of back pain is only slightly or moderately increased in patients with thoracic scolioses [3, 103].

### Paralysis

Spontaneously occurring paralysis does not occur in idiopathic scoliosis (paralysis only occurs in cases of congenital kyphosis and secondary scoliosis resulting from a tumor).

### Curve types

In 1983, King et al. [53] proposed a classification system with 5 curve types. Although commonly used for many years it only provides an incomplete description of the possible types. A new classification was proposed by Lenke et al. [58] in 2001. This distinguishes 6 types of scoliosis, each of which can be subdivided according to the extent of lumbar deviation (Fig. 3.42). This classification is more reliable than that of King [74] and is suitable for establishing the indication for modern surgical procedures. It also takes account of sagittal curvatures. Structural scolioses are defined by the measurement of a minimum scoliosis angle of 25° in lateral inclination on the functional x-ray. A vertical line extending upwards from the center of the sacrum is drawn on the AP x-ray of the standing patient in order to determine the extent of lumbar deviation.

### Treatment

#### Treatment objectives



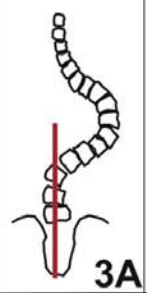

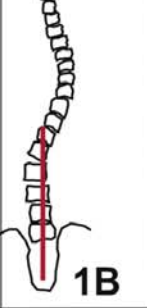
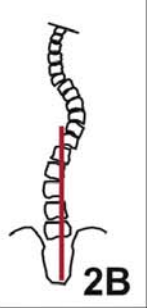
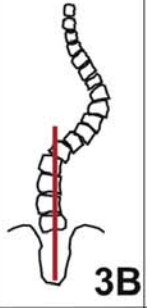
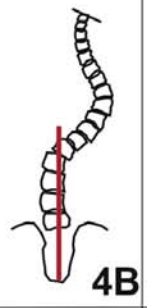

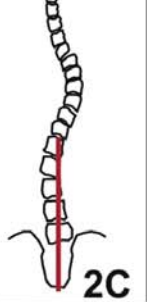
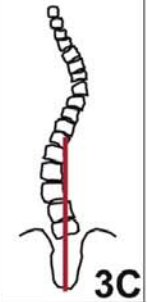
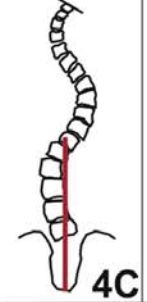
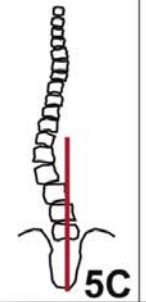
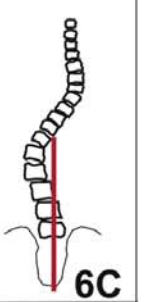
- Prevent progression
- Correct the existing curve
- Preserve the corrected position

#### Available options

- Exercises
- Brace treatment
- Electrical stimulation
- Operation



### Curve type (1-6)

<b>Lumbar deviation (A-C)</b>  <b>A</b> minimal	<b>Type 1</b> Single thoracic	<b>Type 2</b> Double thoracic	<b>Type 3</b> Double major	<b>Type 4</b> Triple curve	<b>Type 5</b> Thor.-lumbar or lumbar	<b>Type 6</b> Thor.-lumbar or lumbar
<b>B</b> moderate	 1A	 2A	 3A	 4A		
<b>C</b> severe	 1B	 2B	 3B	 4B		
<b>Sagittal plane</b>	 1C	 2C	 3C	 4C	 5C	 6C
	Normal	Cerv.-thor. kyphosis >20°	Thor.-lumb. kyphosis >20°	Cerv.-thor+ thor.-lumb. kyphosis >20°		

■ Fig. 3.42. Classification of scolioses according to Lenke [57]. The various types (1–6, A–C) must be managed by differing surgical approaches. In the sagittal plane, the kyphotic angle is additionally measured between T5 and T12. The sagittal modifier »N« signifies a kyphosis

between 10° and 40°, »–« refers to a kyphosis of <10° and »1« a kyphosis of >40°. Hypokyphoses in this area are typical of idiopathic adolescent scolioses, i.e. the sagittal modifier is »–«

### Physical therapy

Whether physical therapy by itself can prevent the progression of the scoliosis or even improve the condition, is a matter of dispute. The effect of exercises on the extent of the curvature has largely been rejected in the relevant English literature. Doctors in German-speaking countries are familiar with the treatment developed by *Katharina Schroth* in the 1920's, which consists of a combination of intensive in-patient rehabilitation and outpatient physical therapy. A recently published controlled study showed, for the first time, that the scoliosis of patients treated according to the Schroth method progressed significantly less than that of untreated patients [105]. Ideally, however, these results should be confirmed by a study conducted independently of the Schroth Clinic. Nevertheless, it is encouraging to discover that the efficacy of this well-known traditional treatment has now been scientifically documented. Compliance and the motivation of the patient definitely play a crucial role, more so than for other (passive) treatments. Given that neither the brace treatment nor surgery are attractive options for young people, this enhances the importance of physical therapy. Nevertheless, controversy still exists concerning the optimal type of treatment.

Based on our current knowledge of the pathogenesis of scoliosis and also given the substantially increased incidence of scoliosis among female gymnasts and ballet dancers, it is doubtful whether stretching exercises or strengthening of the erector trunci muscle are the appropriate gymnastic measures. Perhaps kyphosing exercises and the practice of corresponding sports (e.g. rowing, cycling with drop handlebars) would be more suitable.

#### Other objectives of physical therapy

- Improve the general posture
- Strengthen the muscles
- Flatten the lordosis
- Improve cardiopulmonary function

Both brace treatment and surgery must always be supplemented by exercises.

#### Clinical and radiographic follow-up

Provided a scoliosis has not exceeded 20°, annual checks (without an x-ray) are sufficient prior to puberty. Radiographic follow-up is only necessary if the scoliosis is thought to be increasing on the basis of rib prominence measurement with the inclinometer or raster stereography. The clinical checks should be increased to half-yearly during the pubertal growth spurt, or even every 3 months

for scolioses over 20°. Even for this age group we only record an x-ray if the clinical parameters suggest the occurrence of progression.

#### Plaster cast and brace treatment

Plaster cast and brace treatment is a non-surgical option whose efficacy has been scientifically proven.

#### History

In 1579, Ambroise Paré fitted 2 metal plates to the front and back of the body to straighten a crooked spine. In 1650, Glisson introduced traction treatment, primarily for the correction of rachitic scoliosis. Plaster techniques were developed in the 19th century. In 1895, Friedrich Hessing used a brace with an additional neck ring for extension. In 1927, the »turnbuckle cast« was introduced by Risser. In this technique the patient was extended by traction applied to the head and legs and then placed in a plaster cast. In 1952, Risser presented the »localizer cast«, a more sophisticated form of the »turnbuckle cast« that applied additional lateral corrective pressure. This treatment was perfected by Cotrel. In 1947, Blount et al. presented the Milwaukee brace [6], which combined the extending effect of a neck ring with the corrective effect of pads. Stagnara introduced a brace with a compressive action while, in 1975, Hall developed the Boston brace as a purely corrective orthosis [35].

#### Mode of operation of modern braces

Modern braces for the treatment of scolioses work according to one of the following 4 principles:

- active or passive extension with the aid of a neck ring and correction by lateral pads (example: Milwaukee brace),
- correction by lateral pressure according to the 3-point principle (examples: Boston brace, Cheneau brace),
- correction by compression (examples: Stagnara brace, Wilmington brace),
- correction by bending the trunk towards the opposite side (example: Charleston brace).
- active braces such as the SpineCor correct by means of pressure exerted by bands during movement.

The Milwaukee brace was primarily designed as a passively extending brace [5]. A chin and neck ring was connected to a pelvic girdle by rods, and the rods were elongated to produce passive extension. However, this resulted in pressure points and deformities of the jaw. In a later development, the chin and neck ring only produced an admonitory effect and sought to compel the patient actively to straighten up. We now know that the extending principle for the treatment of idiopathic adolescent scoliosis is basically incorrect. The extension and any as-

### 3.1.4 · Idiopathic scolioses

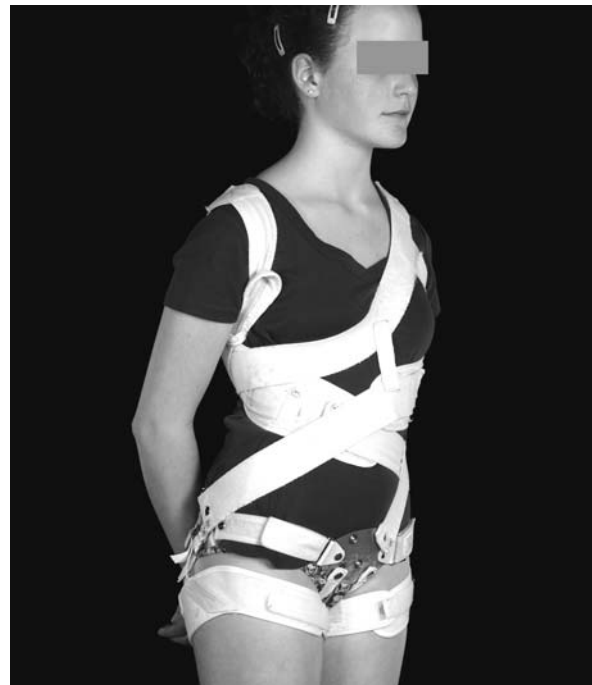
sociated lordosing effect are not desirable as the scoliosis is primarily a rotational lordosis.

We do not have any experience with compressing braces (Stagnara or Wilmington brace) because of concerns about the possible impairment of lung function.

We use braces that produce a corrective action by pads and also have a derotational effect, for example the Cheneau brace [104] (■ Fig. 3.43) and the Boston brace [35]. These correct the scoliosis according to the 3-point principle:

1. support for the pelvis with a pelvic girdle,
2. support under the axilla on the concave side,
3. correction below the apex of the scoliosis on the convex side. Such braces have a derotational effect, although this tends to affect the rib hump rather than the actual scoliosis [106].

Recently we have also used the dynamic brace (SpineCor, [14]). Fabric bands tensioned to a computer-calculated force produce a mainly corrective action during movements of the patient (■ Fig. 3.44). However, the efficacy of such braces has yet to be adequately proven in scientific studies.



■ Fig. 3.44. The *SpineCor* brace is a dynamic brace consisting of fabric bands with a computer-controlled tensile force



■ Fig. 3.43. The *Cheneau* brace is a modern derotation brace whose primary objective is not extension but lateral correction according to the 3-point principle and derotation

### Indication for brace treatment

We consider that brace treatment is indicated if the following apply

- idiopathic scolioses with a Cobb angle of  $>20^\circ$ ,
- proven progression (more than  $5^\circ$  difference between 2 check x-rays),
- growth potential still present (Risser grade III or lower).

For scolioses with a Cobb angle of  $>30^\circ$  and the existence of further growth potential we consider that a brace is indicated even if progression is not proven.

The wearing of a brace during puberty is not without its problems in adolescents. At this age there is a great need *not* to appear different from their peers if possible. This is expressed in their clothing and also in musical, sporting and other leisure interests. A female scoliosis patient will often be the only person in her class, or even in the whole school, wearing a brace. She therefore feels much more isolated than, say, a wearer of dental braces. In fact, dental braces are prescribed so frequently nowadays that over half the students in the class will be wearing a brace, and dentists are occasionally confronted with young patients with normal teeth asking to be fitted with braces just so they don't differ from their classmates.

The psychological effects of brace treatment have been investigated in several studies. Although there were hardly any serious long-term adverse effects, for most adolescents the brace treatment was a psychological burden [2, 50]. This also explains why compliance with the treatment is not very good. Actual wear periods of 65% are reported in follow-up surveys [22]. In an interesting study involving 50 patients with a Boston brace, silver platelets were incorporated in the brace without the knowledge of the patients. These platelets oxidized on contact and were thus able to show the contact time very accurately. The brace was only actually worn for 17% of the prescribed time (Houghton 1987, personal communication; unfortunately the author died before he was able to publish this interesting study). While this investigation calls into question the efficacy of brace treatment, a more recent study showed that a brace worn for only part of the time is almost just as efficient as when worn for the whole day. A significant advantage of the dynamic SpineCor brace is the much better acceptance by the patients and thus better compliance.

Brace treatment is even more important for lumbar scolioses, however, than for thoracic scolioses, since surgery does not represent a satisfactory alternative for the lumbar form.

### Results of brace treatment

Although brace treatment can halt the progression of the scoliosis it cannot correct the condition in the long term. The initial reduction in curvature is lost again when treat-

ment is discontinued after the completion of growth. If the brace is consistently worn, however, the patient can be reassured that the scoliosis will continue to remain exactly as it is. On the other hand, braces are only this effective up to a curve angle of approx.  $40^\circ$ .

A meta-analysis by the Scoliosis Research Society involving a total of 1910 patients (1459 of whom were treated with a brace, 322 with electrical stimulation and 129 were only observed) produced the following results: The weighted success rates were 0.39 for electrical stimulation, 0.49 for observation and, for brace wearing, 0.6 for 8 hours, 0.62 for 16 hours and 0.93 for 23 hours. Electrical stimulation was therefore ineffective, while the brace treatment was more efficient the longer it was worn each day [90].

In a recent study compliance was measured electronically by embedding a temperature sensor and logger in the Wilmington scoliosis brace in 34 patients. The compliance rate for the patients whose curve progressed ( $>5^\circ$ ) was 62%; the compliance rate for the patients who did not progress was 85%. In the group that had high compliance ( $>90\%$ ), one of the 9 subjects' curves progressed (11%). In the group with low compliance ( $<90\%$ ), 14 of the 25 subjects' curves progressed (56%) [81].

In order to prove successful, the brace actually needs to be worn for 23 hours each day [72, 90]. Only the Charleston brace appears to be effective when worn exclusively at night [31].

The following factors should also be taken into account during brace treatment:

- The brace impairs lung function while it is worn, though this recovers very quickly when the brace is discontinued [83]. This effect may be avoided with the new dynamic braces (SpineCor), although corresponding study results are still awaited.
- The brace cannot correct the rotation of the vertebral bodies [106] or the lordosis.
- The surface of the back (i.e. the rib prominence) is corrected to a slightly greater extent than the scoliosis itself.
- Brace-wearing period:

The brace must be worn consistently day and night until growth is complete (Risser IV). Since the psychological strain associated with brace treatment is considerable, the patient needs good support. Only if everyone involved (doctor, parents, physical therapist, teachers) is convinced of the purpose of the treatment will there be any prospect of success. The brace must be worn for 22–23 hours a day, and may be removed only for sports activities or during physical therapy. Sport may be practiced without restriction. Brace treatment must *always* be backed up by physical therapy and exercise since wearing of the brace results in atrophy of the paravertebral muscles, which have less postural work to do. This atrophy must be countered by swimming, sport and postural exercises. As the

### 3.1.4 · Idiopathic scolioses

patient is weaned off the brace we do not consider that it must continue to be worn during the night.

#### Clinical and radiographic follow-up

AP and lateral x-rays of the complete spine are needed to decide whether brace treatment is indicated. Images produced by digital methods are also suitable. When the brace is ready, its effect must be checked radiographically by AP views. Clinical checks are then arranged every 3 months. If raster stereography is not available, x-rays should be recorded every 6 months (AP only, without the brace), and otherwise only if progression is suspected. Regular check-ups should continue at this rate until the patient is weaned off the brace.

#### Electrical stimulation

In the 1970's and 1980's, electrical stimulation raised hopes of an alternative to the brace. However, it has now been shown to be ineffective [3, 90]. Surgery

Surgery can not only halt the progression of scoliosis, but can (to a certain extent) straighten the curvature and essentially maintain the correction after the spinal fusion has stabilized. We consider that surgery is indicated in thoracic scolioses from a Cobb angle of 40° and, in thoracolumbar or lumbar scolioses, from a Cobb angle of 50° or if decompensation is present. The disadvantage of all existing surgical procedures is the need to stiffen the corresponding section of the spinal column.

#### History

The era of the surgical treatment of scoliosis began with *Hibbs* back in 1911. He described a technique of posterior vertebral fusion [43], which he subsequently used in cases of scoliosis. The correction of the curvature was achieved with plaster casts. Subsequent refinements in the plaster cast technique produced such corrective casts as the »*turnbuckle cast*« and the »*localizer cast*«, which were proposed by *Risser* [87].

In 1962, *Harrington* reported on the correction of scolioses by instrumentation [38]. This primarily involved a distraction rod that was used on the concave side of the scoliosis. Subsequently, *Harrington* supplemented this instrumentation with a compression rod used on the convex side of the scoliosis.

In 1969, and unaware of the *Harrington* procedure, *Dwyer* in Australia described a correction method for scoliosis with an anterior approach [26]. He used screws inserted from the front into the vertebral bodies on the convex side and linked with each other by a cable. The correction was achieved by pulling on the cable to produce compression. The main disadvantage of this method was the kyphosing resulting from the resection of the intervertebral disks, which is highly undesirable in the lumbar area.

#### Development of existing commonly used instrumentations

The kyphosing problem was resolved in 1975 by a modification of the *Dwyer* procedure described by *Zielke* and known as *ventral derotation spondylodesis (VDS)* [113] (■ Fig. 3.45 and 3.46). With the *Zielke* instrumentation, a derotation can be performed while at the same time preserving the lordosis. Instead of a cable, *Zielke* used a threaded rod that can be rotated by a special derotation instrument. Kyphosing is avoided by the insertion of allogenic bone grafts in the spaces between the disks.

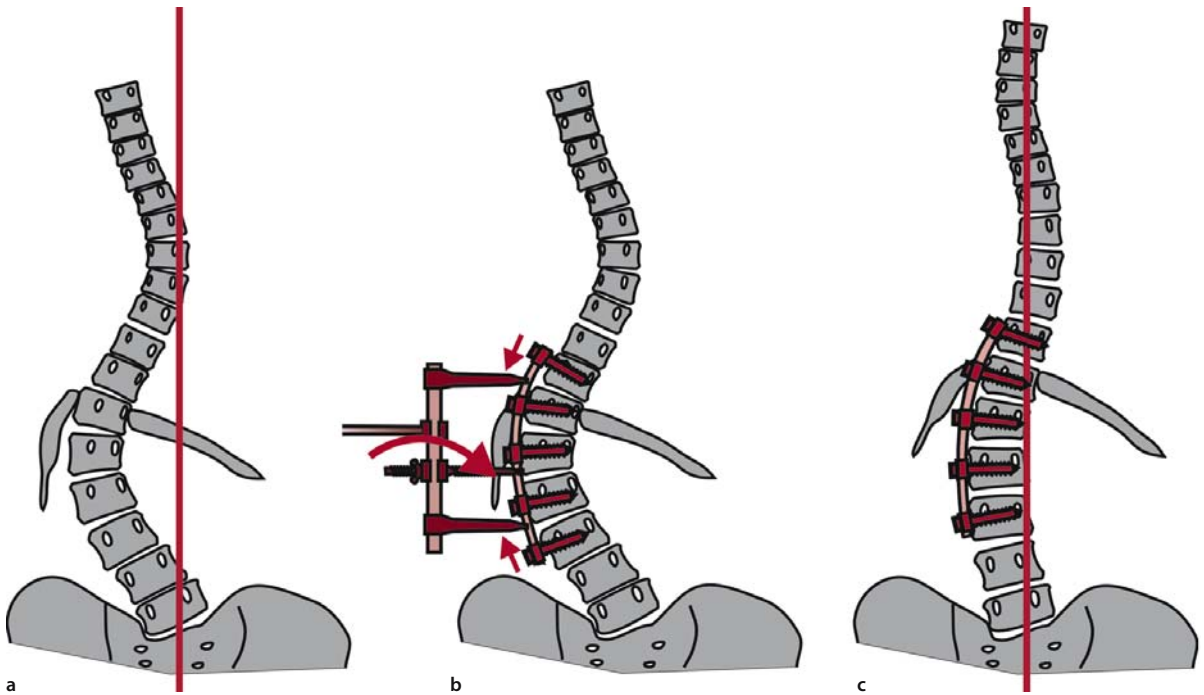
In the 1970's *Luque* introduced the rods named for him (which are anchored without hooks) and the technique of *segmental wiring* [65]. The principal advantages of segmental wiring: the correction is produced not just via longitudinal but also via transverse forces; a certain amount of derotation also occurs, thereby increasing stability. This technique still has an important role to play in neuromuscular scolioses.

At the start of the 1980's an instrumentation system that introduced new elements in the surgical treatment of scoliosis was developed by *Cotrel* and *Dubousset* in France [16]. This system allows the curvature to be corrected in three dimensions and provides stable fixation with a frame construction. Hooks and screws can be fixed to bendable rods at any desired point and in any desired position. The treatment of thoracic scoliosis in this system is based on the principle of inserting several hooks (usually 4) at certain points on the concave side. A rod that imitates the curve of the scoliosis is then inserted. The hooks are fixed with C rings, i.e. their spacing is fixed but they are able to rotate. The rod is then rotated through 90° in the dorsal direction, i.e. from scoliosis to kyphosis. Thus, at one and the same time, the scoliosis is reduced, the kyphosis increased and the spine derotated. The surgeon then applies distraction and secures the hooks in place. Another rod that exerts a compressive force is inserted on the convex side (■ Fig. 3.47).

In lumbar scolioses the rod on the concave side is rotated to a position of lordosis rather than kyphosis. A whole series of instrumentations has since appeared on the market, all of which are based on the principle of *Cotrel* and *Dubousset* and each offering its own particular advantages (e.g. easier removal, more flexible attachment of hooks and screws, more economical), for example the »*Isola*« or »*Texas Scottish Rite Hospital*« instrumentation systems.

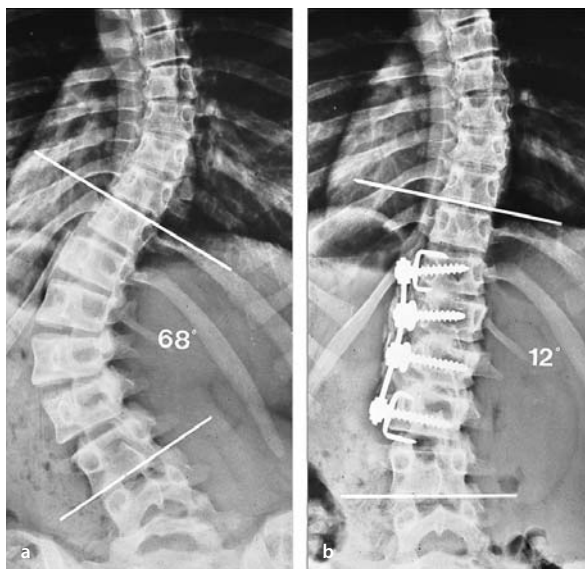
The segmental principle of *Luque* wiring is further perfected in the »*Universal Spinal System*« [4, 78]. A rod is inserted from the concave side and placed in the planned position of kyphosis or lordosis, and hooks and screws are inserted into the laminae or pedicles at various levels and then pulled towards the rod (■ Fig. 3.48 to 3.50). The advantage of this system over the *Cotrel-Dubousset* technique is the absence of any increase in the rotation of the countercurve.



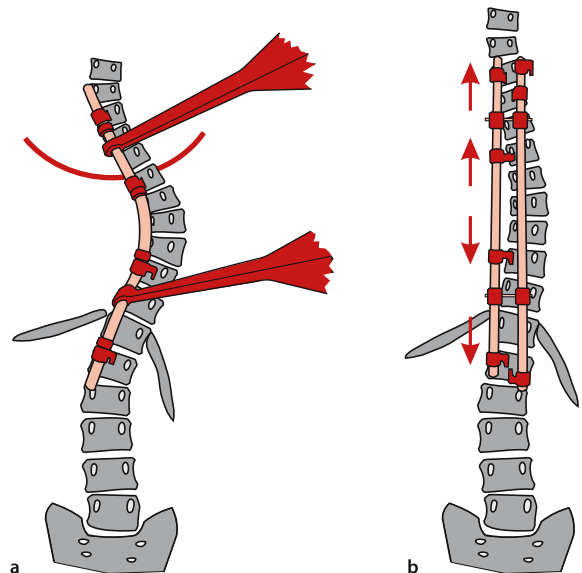


**Fig. 3.45a–c.** Schematic presentation of the ventral correction of lumbar and thoracolumbar scolioses with the VDS system according to Zielke: **a** Preoperative status. **b** Instrumentation and use of the derotation rod. In contrast with the Cotrel-Dubousset procedure, the rod is not rotated as a whole but is rotated more strongly (since it is flexible) in the center than at the ends. As a result, the derotation is transferred to a lesser extent (as additional rotation) to the countercurve. **c** Postop-

erative status. It usually proves possible to realign the spine, although it is almost always slightly out of alignment on the convex side of the lumbar curve. For (purely two-dimensional) geometrical reasons, one would expect it to go even further out of alignment if the curve is extended. However, since the end vertebra at the base of the spine is aligned horizontally the opposite is the case



**Fig. 3.46a, b.** AP x-rays of a 12-year old female patient with thoracolumbar scoliosis preoperatively (**a**) and 1 year postoperatively (**b**) after surgical correction with the VDS system according to Zielke



**Fig. 3.47a, b.** Schematic presentation of the Cotrel-Dubousset procedure. **a** The rod on the concave side is inserted through the prepared hooks and then turned to change the scoliosis into a kyphosis. The spine is slightly extended and derotated during this procedure. **b** Final situation

### 3.1.4 · Idiopathic scolioses

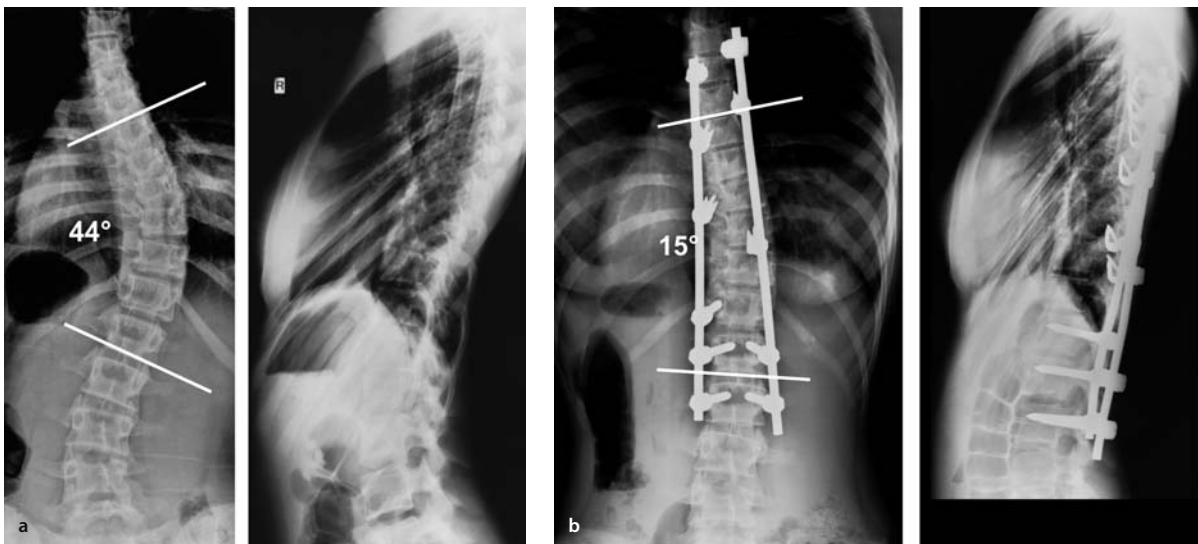
The *anterior techniques* were also perfected. The insertion of 2 rods, or the use of a *double-rod system* [36, 45], produced primary stability, thus dispensing with the need for a brace and involving fewer correction losses. Ideally, derotation is produced with a flexible rod (= less risk of pull-out at the rod ends) and stabilization with a rigid implant (= stronger result; ■ Fig. 3.51).

*Thoracoscopic techniques* for anterior epiphyseodesis (in young patients to avoid the so-called crankshaft phenomenon; see below) and for anterior release in severe scolioses are now considered to be standard treatment [27], although the instrumentation for minimally-invasive thoracoscopic techniques is still the subject of ongoing research and development [28].

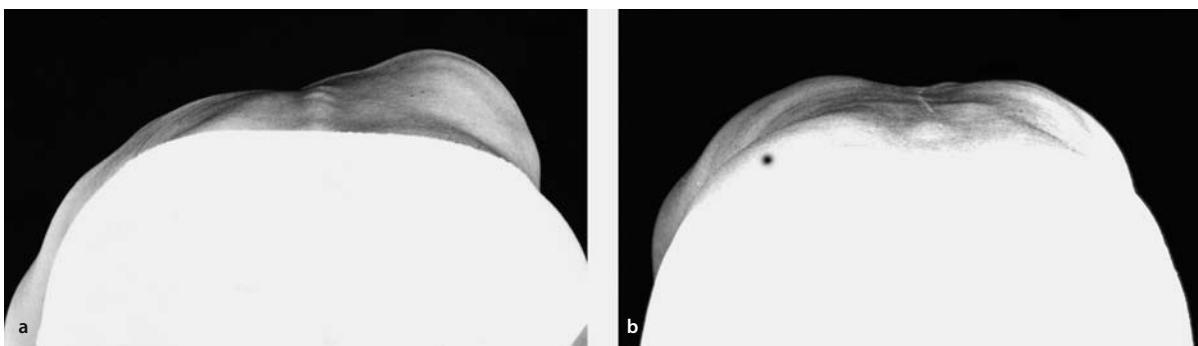
#### Preoperative investigations and measures

Preoperatively we record *standard AP and lateral x-rays* of the whole spine, as well as *functional x-rays* with the

patient in lateral inclination (see above) for all curves. The scoliosis is classified according to the Lenke system and the surgical procedure established on the basis of these radiographs. To minimize the risk of disease transmission by blood transfusions, we recommend *autologous blood donation* before surgery wherever possible. This is usually an option in patients aged 5 years and over. 3–4 units of blood are taken over a maximum period of 4 weeks for subsequent reinfusion during or after the operation. Thanks to the use of other blood-saving measures (blood dilution/hemodilution, return of blood lost during the operation via the »cell saver«, controlled hypotension etc.), we almost never require homologous blood for idiopathic scoliosis. As a precaution we record an *MRI scan* preoperatively to rule out any intraspinal abnormalities. *Sensory and motor potentials* are recorded on the day before surgery in order to determine the baseline levels in the alert state.



■ Fig. 3.48a, b. 14-year old female patient with thoracic idiopathic adolescent scoliosis, Lenke type 1B. a Preoperative AP and lateral x-rays of the spine, b result 1 year postoperatively after correction with USS (Universal Spinal System) instrumentation

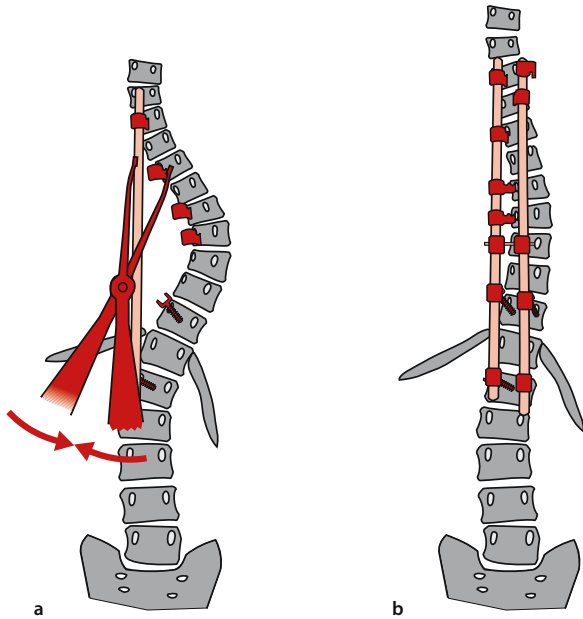


■ Fig. 3.49a, b. Clinical photograph of a forward-bending position preoperatively (a) and 1 year postoperatively (b) in the same patient as in Fig. 3.48. The correction of the rib prominence is clearly visible

### Surgical methods

#### Purely posterior methods with two rods

We use purely posterior methods exclusively for thoracic scoliosis with curve types 1 and 2 according to Lenke. The lumbar curvature in these cases is not structural (as can be established on the basis of the functional x-rays). We use the USS instrumentation (Universal Spinal System), which primarily applies transverse forces (see above;

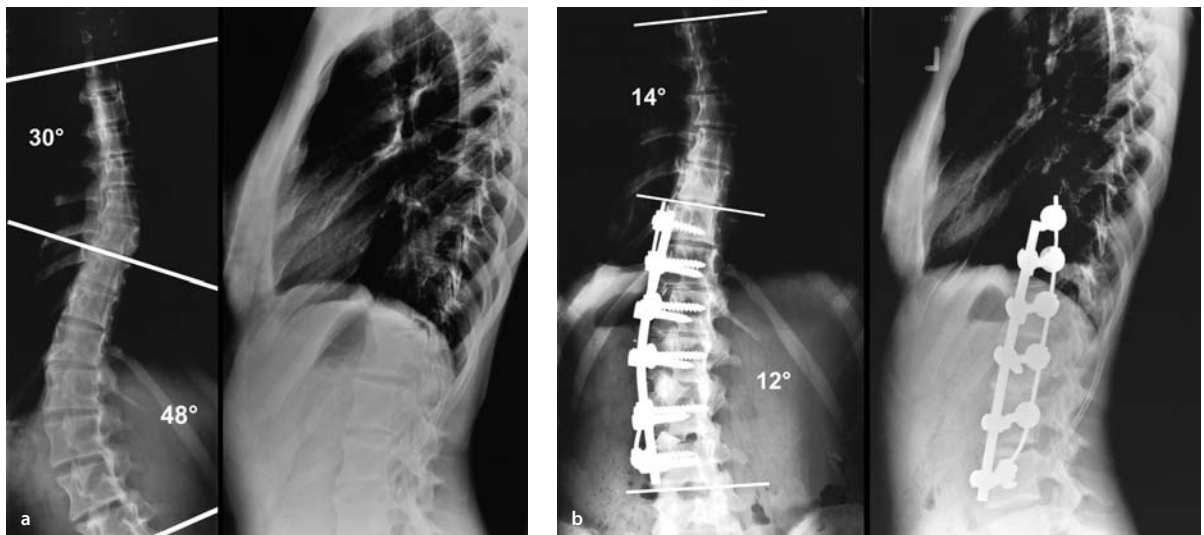


■ Fig. 3.50a, b. Schematic presentation of the operation with USS instrumentation. **a** The hooks and screws inserted on the concave side are secured to the rod, which is curved into the kyphosis but straight in the frontal plane. Distraction does not occur, or else only occurs indirectly. **b** Postoperative situation

■ Fig. 3.48 and 3.50). The centered vertebra is located on the x-rays, i.e. the vertebral body in the area of the thoracolumbar junction that is best centered over the middle of the sacrum. This segment does not usually correspond to the end vertebra of the thoracic scoliosis, but is usually slightly lower. This vertebra is optimal for the insertion of the caudal screw. Where possible, the cranial end of the spondylodesis should also be located in a centered vertebra. For a type 2 Lenke curve with two structural thoracic curves the instrumentation must be placed in a much more cranial location (often as far as T1). In the lower part of the spine we prefer screws, in the upper part hooks, but it does not seem to play a major role whether hooks or screws are used [96]. We regularly perform a *rib hump resection* at the same time as the scoliosis operation. In all cases, the procedure should be supplemented by careful fusion, with the opening of all intervertebral joints and the insertion of autologous cancellous bone. We perform an *anterior release* (usually by thoracoscopy) before the posterior straightening, both for very severe curvatures (to improve the conditions for correction) and in patients younger than 12 years (to prevent the crankshaft phenomenon; see below).

In cases of very severe and rigid scoliosis we perform a thoracoplasty on the concave side (i.e. resection of rib segments and fixation to the rod on the posterior side). This improves the correction options and the final cosmetic appearance. In young healthy patients with idiopathic scoliosis, the bilateral thoracotomy and destabilization of the ribs is surprisingly well tolerated.

Alternatively, a double-rod system can be inserted via an anterior approach for lordotic thoracic curves. The hypokyphosis can be corrected rather more efficiently thanks to the removal of the intervertebral disks.



■ Fig. 3.51a, b. 14-year old female patient with lumbar scoliosis (Lenke type 5C). **a** Preoperative AP and lateral x-rays of the spine. **b** X-rays 1 year postoperatively after ventral correction with a double rod (combination of Zielke rod and Spinefix rod)

### Purely anterior methods with two rods

Posterior methods must be instrumented down to the centered vertebra, i.e. usually at least 1 (or 2) segment(s) below the end vertebra. This greatly restricts the mobility of the spine in the lumbar area, and instrumentation down to L4 or even L5 is associated with a high risk of disk degeneration and lumbar back pain [15]. We therefore consider anterior methods to be more appropriate for type 5 lumbar and thoracolumbar scolioses, since these only require instrumentation down to the end vertebra. We use two rods (a flexible Zielke rod for derotation and a rigid USS rod for stabilization) (■ Fig. 3.46). Posterior fusion is additionally required in female patients who have not yet reached the menarche and boys under 13 years of age in order to avoid subsequent kyphosing.

### Combined anterior and posterior methods

If both the lumbar and thoracic curves have to be instrumented (types 3, 4 and 6), a two-stage approach is recommended:

1. Anterior straightening of the lumbar scoliosis (one rod is sufficient in this case).
2. Posterior straightening of the thoracic scoliosis with USS instrumentation (1 week later).

In this method instrumentation down to the end vertebra is sufficient and, for the posterior part of the procedure, the spinal fusion does not need to be extended caudally as would normally be the case with purely posterior interventions (■ Fig. 3.52).

### Early onset scolioses

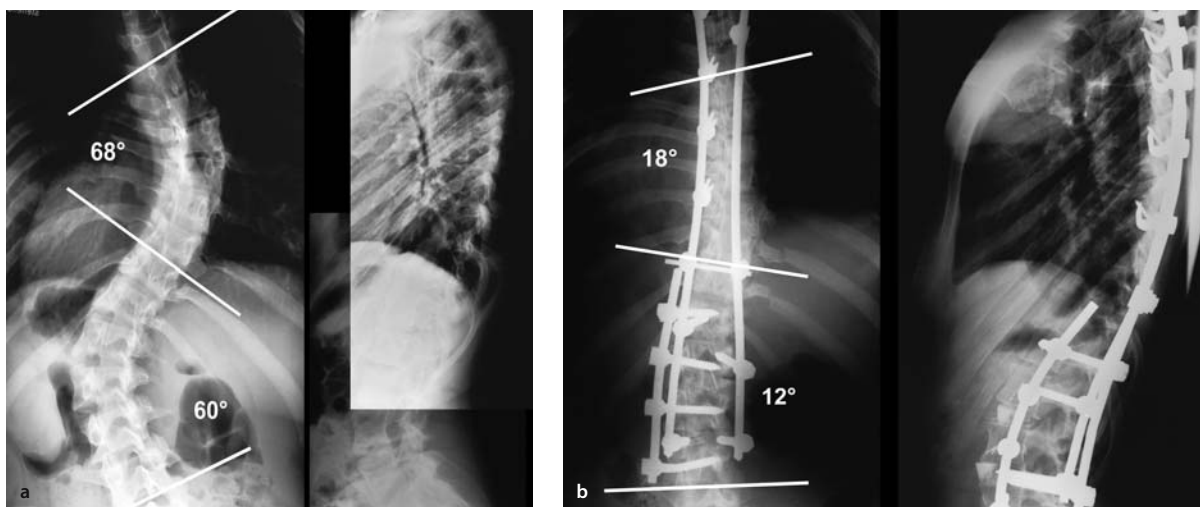
In these patients the doctor should try to keep the extent of the curvature within tolerable limits by conservative measures up until the age of 10. Brace treatment is usually

required. If the scoliosis remains stable for a prolonged period in children under 10 years, the brace can be dispensed with from time to time. If the scoliosis progresses rapidly from the very earliest years of life, we currently perform a correction procedure on the thorax with the VEPTR instrumentation (= »vertical expandable prosthetic titanium rib«). This system is described in greater detail in ► chapter 3.1.7. The advantages of this method are the enlarged thoracic space, the efficient slow straightening and the reduced risk of spontaneous stiffening (in respect of all implants secured directly to the spine itself). The risk of progression increases substantially with the onset of the pubertal growth spurt, and the stiffening operation is also generally required at this stage [40, 88]. This should always be performed simultaneously from the anterior and posterior sides regardless of the anatomical location.

### Non-fusion methods

If early onset scolioses progress rapidly before the age of 10 years, »growing rods« are commonly used. This technique goes back to Harrington, who initially did not fuse the instrumented segments [38]. The rod is either lengthened periodically, or it serves as a splint and allows growth of the spine (but, of course, the rod itself does not »grow«, it is only passively lengthened). A typical representative of this principle is the »Luquet trolley« [80]. Other single and dual rod systems are commonly used [1]. We have used »growing rods« for decades. In our experience spontaneous fusion of the spine inevitably occurs after 3 to 4 years because of the stiffening effect of any rod which is fixed to the spine. This is why we nowadays prefer the VEPTR, which allows movement between the rod (which is fixed to the ribs) and the spine.

A new non-fusion technique for adolescent scoliosis uses titanium staples at the anterior convex side of the



■ Fig. 3.52a, b. 15-year old female patient with triple curve (Lenke type 4C). a Preoperative AP and lateral x-rays of the spine. b 1 year after correction with a combined ventral and dorsal procedure)



scoliosis [5]. Though it primarily avoids fusion of the spine, the aim of preservation of mobility probably cannot be reached. The staples stiffen the spine and they hardly can ever be removed. They also produce a growth disturbance. In our view, a more promising method would be a temporary continuous growth stimulation at the concave side. With such a method preservation of mobility should be possible. We are working on such a development.

### Supplementary remarks

The sagittal plane and the extent of the rotation must be taken into account when selecting the procedure and the extent of the spinal fusion.

In children under 12 or for very severe scolioses (over 60°), we always perform an anterior (thoracoscopic) disk removal procedure before a posterior correction. We supplement the (lumbar) ventral derotation spondylosis (because of the risk of additional kyphosing) with posterior tension-band wiring.

➤ In almost all idiopathic thoracic scolioses we routinely resect the *rib hump* at the same time as the straightening procedure.

### Results of surgical scoliosis treatment

#### Frontal plane (correction of the Cobb angle)

Corrections of between 28% [42] and 55% [67] have been achieved with the *Harrington* instrumentation, while corrections of 55% have been calculated for the use of the *Harrington-Luque* technique [67]. The correction results reported for the *Cotrel-Dubousset* instrumentation are between 48% and 69% [89, 109]. On average, the result is comparable with that for the *Harrington-Luque* method. Figures as high as 87% are reported for the anterior *Zielke procedure* [108]. Average corrections of over 70% are also possible with the modern anterior double-rod systems [36, 37]. As a rule, anterior techniques with disk removal produce better correction results than posterior operations.

#### Sagittal plane (kyphosing/lordosing)

While *Harrington* rods exacerbate the pre-existing pathological lordosis of the thoracic spine observed in most cases, the combination of *Harrington* rods with segmental wiring according to *Luque* and the *Cotrel-Dubousset* procedure produce a (moderate) kyphosing effect. The anterior procedures (*Zielke*, *Dwyer*) tend to produce kyphosing of the spine as a result of the removal of the disks. This is not desirable in the lumbar or thoracolumbar region (where these operations are predominantly sited) and the kyphosing can be avoided, or at least reduced, by the insertion of substantial allogeneic bone chips or synthetic implants.

#### Horizontal plane (derotation)

Not much information is available in the literature on the effect of *Harrington* rods on the rotation of the vertebral

bodies. In our own investigation we observed that the *Harrington* instrumentation increases the rotation, particularly if compression rods are used. Even more marked is the rotating effect if the two rods are connected with each other. Since segmental *Luque* wires pull on the vertebral arches on the concave side, they produce a derotating effect, though this is only slight. Nevertheless, derotation of 5°–10° is still possible [67, 101]. Much has been written about the derotating effect of the *Cotrel-Dubousset* system. During the operation the rod is turned through approx. 90°, which gives the impression that substantial derotation is taking place. In fact this only amounts to approx. 0°–10° [55].

The most efficient method in terms of derotation is *ventral derotation spondylosis (Zielke procedure)*. Since the anterior longitudinal ligament and the intervertebral disks are removed during this procedure, the strongest forces of resistance to the derotation are eliminated. The mobility of the spine is also greater in the lumbar compared to the thoracic area. As we ourselves have discovered, average derotations of 20° to 30° are perfectly possible.

### Long-term results

Genuine long-term results are available, particularly after *Harrington* procedures: Several studies have reported on follow-up periods of over 20 years [17, 18, 42, 75, 94]. A correction loss of 5–10° occurs over time [17, 42]. However, the functional result is generally very good and the frequency of back pain is no greater than in a healthy comparison population [75]. Scoliosis surgery does not adversely affect partnerships or pregnancy [18]. Only patients with a badly distorted sagittal profile experience increased episodes of back pain [94].

### Complications of scoliosis surgery

The following possible complications are associated with scoliosis surgery:

#### Early complications

■ *Neurological lesion*: Although the risk is relatively low in idiopathic scoliosis, it is highly relevant as the occurrence of such a lesion can be devastating for the patient and his family. Good preoperative briefing is important in this respect. Differing figures have been stated for the risk in the more recent literature. A meta-analysis by the Scoliosis Research Society found an incidence of 0.3% of partial lesions, but no complete lesions, in 2031 operations [107]. Another larger-scale study reported four cases of partial lesions in 1090 operated patients (0.4%) [7]. Three of the four lesions were of vascular origin. The combined anterior and posterior procedure and severe kyphoses were found to be risk factors. Much higher percentages, however, were cited in another study (17 lesions in 667 operated patients = 2.5%) [69].



### 3.1.4 · Idiopathic scolioses

To minimize the risk it is important, particularly for atypical scolioses, to rule out any other cause by MRI (e.g. a syrinx or other intraspinal anomaly). An MRI scan is recommended, though not absolutely essential, in cases of typical scoliosis. Intraoperative monitoring is also important. Recording *somatosensory and motor potentials* during the operation is the most reliable way of detecting lesions at an early stage and eliminating the triggering mechanical factor [64]. The chances of a recovery are much better with an immediate reaction than one produced several hours later. Motor potentials are considerably influenced by anesthetic drugs and their reliable recording is only possible in close consultation with the anesthetists. In our own hospital, therefore, the intraoperative potentials are monitored by the anesthetists themselves. If this method is not available, an intraoperative wake-up test should be performed.

- *Infections* are, statistically speaking, the complications most likely to occur. Recent careful studies have reported infection rates for idiopathic scolioses ranging from 1.7% (i.e. 22 out of 1250 patients [12]) to 4.7% (i.e. 23 out of 489 patients [85]). In most cases these were superficial infections. More problematic are the deep infections, which must be treated with antibiotics until the spondylodesis has consolidated and any metal implants can be removed. Such infections often occur only at a late stage (i.e. after several months). An increased risk exists for patients with severe acne [34]. We treat any patients diagnosed with acne with antibiotics for a month before the operation (in consultation with the dermatologist).
- After anterior procedures or rib hump resections, a *pneumothorax* or *pleural effusion* are very common complications. Prolonged drainage is sometimes required, although an uncomplicated recovery invariably occurs in young healthy patients.
- A *chylothorax* can occur after anterior procedures [19]. In our own experience, spontaneous reabsorption occurred in the two relevant cases.

#### Late complications

- A particular problem is posed by postoperative *decompensation* of the spine. This is observed more frequently in cases involving the use of the Cotrel-Dubouset instrumentation for King type II scolioses (corresponding to Lenke 2C or 3C) [57, 89, 110]. This complication is thought to be attributable to excessive rotation of the lumbar countercurve during the maneuver for derotation of the thoracic spine. This problem occurs much less frequently with the USS system which, like segmental wiring, works by applying transverse correction forces.
- Another serious problem is the *loss of lumbar lordosis*. This risk particularly applies with anterior procedures [61, 71], but can be avoided if a correct technique is

employed. Surgeons must also be careful to ensure the preservation of lumbar lordosis in surgical correction from a posterior approach.

- Another possible complication is *correction loss*, although nowadays this occurs much less frequently with modern instrumentations than when the Harrington operation prevailed. The correction loss always occurs during the first three years, after which time the patient's condition stabilizes. It is particularly pronounced if a *pseudarthrosis* establishes itself, which can frequently lead to *rod fracture*. If the metal has to be removed for any reason, correction loss can subsequently recur as a result of instrumentation-related osteoporosis. Not least for this reason, the metal should not be removed if possible [75].
- A special problem is encountered after surgery on young patients requiring a fusion before the appearance of the iliac crest apophysis (Risser I). Correction loss and increasing rotation occurs in these patients as a result of the continuing growth of the vertebral bodies [25, 40, 92]. This is termed the *«crankshaft phenomenon»*, and is also observed after a purely anterior correction. Consequently, an anterior spondylodesis must always be combined with a posterior fusion in young patients (Risser 0).
- Long-term observations have shown that patients can remain symptom-free for decades after correctly-performed scoliosis operations [17, 18, 42, 75, 94]. It is important that the spine should remain in alignment. Fusions down to L3 or lower can be problematic. Increased pain can be expected at a later stage particularly after stiffening procedures down to L4 or L5 [30]. The end vertebra of lumbar scolioses is usually L3. Fusion should not continue beyond the end vertebra in the ventral derotation spondylodesis, in contrast with all posterior procedures. We therefore always instrument and fuse idiopathic lumbar scolioses from the anterior side and no lower than L3. Patients with lumbar scolioses are associated with an increased risk of later back pain even without surgery. Particularly serious problems can be expected in connection with decompensation of the spine. For lumbar scolioses, there is an especially great need for a treatment method that avoids stiffening of the affected section of the spine. In our hospital we are currently developing a method that allows correction of the scoliosis via an externally extendable implant. Whether this method will one day be able to help avoid stiffening of the scoliotic section of the spine in children and adolescents remains to be seen.

#### Our therapeutic strategy for idiopathic adolescent scolioses

The therapeutic strategy for idiopathic adolescent scolioses in our hospital is shown in ■ Table 3.2.

**Table 3.2. Treatment concept for idiopathic scoliosis**

Growth phase	
Scoliosis angle less than 20°	No treatment
Scoliosis angle 20°–30°	If progression is conformed and more than a year of the pubertal growth spurt remains (in girls up to a year after the menarche or Risser III) prescribe brace, SpineCor treatment or Schroth therapy
Scoliosis angle 30°–40° (thoracic) and 30°–50° lumbar	If more than a year of the pubertal growth spurt remains: brace treatment
Scoliosis angle from 40° (thoracic) and from 50° lumbar	Surgery usually indicated. In case of decompensation (spine out of alignment) in thoracolumbar or lumbar scolioses sometimes also with smaller angles
After completion of growth	
Scoliosis angle less than 40° (thoracic) or 60° lumbar	No treatment
Scoliosis angle 40°–60°	Thoracic: surgery if the patient wants cosmetic improvement Lumbar: surgery recommended only if decompensation is present
Scoliosis angle greater than 60°	Surgery recommended since further progression is likely during adulthood

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### 3.1.5 Scheuermann's disease

» *Shy young lad, not yet a man, can't stretch his back because of Scheuermann.* «

#### Definition

Growth disorder of the spine with narrowing of the intervertebral disks, wedge vertebrae formation, collapse of endplates and kyphosis in the affected area. The disease can occur at the thoracic, thoracolumbar or lumbar level.

— *Synonyms:* Adolescent kyphosis, kyphosis juvenilis, roundback deformity.

#### Historical background

The disease was first described in 1921 by H. W. Scheuermann [15]. In 1936 the same author coined the term »kyphosis juvenilis«. In 1930 C. G. Schmorl discovered the collapsed intervertebral disks that were typical of the condition [16].

#### Frequency

Figures on the frequency of this condition vary considerably since it is assessed according to widely differing criteria. Frequencies ranging from 1% [1] to as high as 15% [12] have been calculated depending on the threshold value specified for the kyphosis. Reports on the predominance of either sex are also contradictory [1, 12]. In our own clinical experience cases of Scheuermann disease that need to be treated are much rarer than scolioses requiring treatment.

#### Etiology

##### — Mechanical factors:

Tall adolescents, competitive athletes (track-and-field athletes, javelin throwers, rowers, racing cyclists). In 62 excessively tall girls (over 5 ft. 11 in.), Scheuermann was found in 30% of cases [17].

##### — Endogenous factors

Although there is no clear evidence to indicate that the disease is inherited, it does occur more frequently in individual families [9]. In a pair of monozygotic twins the typical changes were found at the same level of the spine, which suggests that a genetic component is probably involved [6].

##### — Posture

In contrast with scoliosis, posture does play a significant role in the development of Scheuermann disease. A long-term hyperkyphotic posture results in increased pressure in the anterior sections of the spine, thus promoting the onset of Scheuermann disease.

##### — Psychological factors

Although the scientific data are scant, it is nevertheless clear that psychological factors play an important role in the development of Scheuermann disease. The

affected individuals are often adolescents with a very dominant parent who adopt a subservient posture and dare not contradict that parent or those with a weak, undeveloped personality. In many cases, these patients have an unconscious desire to conceal something, e.g. a funnel or keeled chest.

##### — Osteoporosis

Investigations (e.g. based on density measurements on CT scans) have shown that osteoporosis is also present in patients with a severe case of Scheuermann's disease.

#### Pathogenesis

While Scheuermann himself assumed that kyphosis juvenilis involved aseptic bone necrosis, similar to that encountered in Perthes disease [15], subsequent studies [2] revealed that the condition actually involves a *weakening of the cartilaginous ring apophyses of the lower and upper endplates of vertebral bodies*. These ring apophyses are the actual growth zones of the vertebral bodies. The mechanical strength of this cartilage can be weakened during the pubertal growth spurt, as occurs in proximal femoral epiphyseal separation. The increased internal pressure in the nucleus pulposus of the disks can cause *disk tissue to protrude through the cartilaginous apophysis* into the vertebral body, thereby displacing the bone trabeculae locally. The trabeculae react by forming an area of sclerosis around the disk tissue, producing a »Schmorl node« or »endplate collapse« or – if the process takes place at the anterior edge of the vertebral body – apophyseal ring herniation. Since the protruding tissue reduces the volume of the nucleus pulposus, the disk appears *narrowed* on the x-ray. The growth of the cartilaginous ring apophysis is impaired overall as a result of the constant anterior pressure, leading to atrophy, i.e. a reduction in the height of the vertebral body anteriorly, and *wedge vertebra formation*. This anterior pressure increases in line with the steadily increasing kyphosis, thereby inhibiting growth. This vicious circle leads to progression of the kyphosis. However, the condition can also develop in a primarily lordotic area of the spine. In this case it is not so much the constant pressure that is important, but rather the constitutional weakness of the cartilaginous apophysis. The diminished growth on the anterior side will eventually lead to kyphosis at the lumbar level as well.

#### Clinical features

The clinical manifestations appear during puberty and are greatly dependent on the *site* of the disease. Thoracic kyphoses hardly cause any symptoms at all, but do produce a visible deformity. On the other hand, patients with a thoracolumbar or lumbar case of Scheuermann disease are often strikingly straight with a flat back. Such patients can experience symptoms at an early stage, i.e. even during puberty.



**!** When adolescents present with severe lumbar back pain with no history of trauma, one should always consider the possibility of lumbar or thoracolumbar Scheuermann's disease. Pigmentation over the spinous processes is often an outwardly visible sign of local kyphosis in the lumbar area.

During the examination it is important to note the fixation of the kyphosis. A flexible kyphosis is not indicative of Scheuermann disease. Only if the kyphosis cannot be corrected during the examination should the possibility of Scheuermann's disease be considered.

One striking feature is the increased finger-floor distance that is almost invariably measured in Scheuermann patients as a result of contraction of the hamstrings. This applies in all cases, regardless of the location of the disease. While the cause of this muscle contracture remains unclear, it may be an expression of a generally contracted posture in Scheuermann patients. Contraction of the pectoral muscles is always present in the thoracic form.

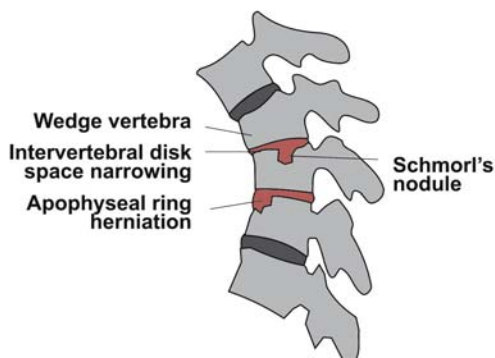
**Radiographic findings**

The typical radiographic changes are shown in Fig. 3.53 and 3.54.

**On the lateral x-ray of the thoracic or lumbar spine we observe:**

- Schmorl nodes
- Apophyseal ring herniation
- Wedge vertebrae
- Intervertebral disk narrowing

These findings may be located purely at the thoracic (Fig. 3.54), thoracolumbar or lumbar (Fig. 3.55) levels. At the thoracic level they are associated with hyperkyphosis, whereas a lumbar finding may initially only be accompanied by slight flattening of the lordosis. Only in



**Fig. 3.53.** Schematic presentation of radiographic changes in Scheuermann's disease

severe cases does actual lumbar kyphosis occur. Fig. 3.56 shows the measurement of the overall kyphotic angle and the angle on wedge vertebrae.

**Differential diagnosis**

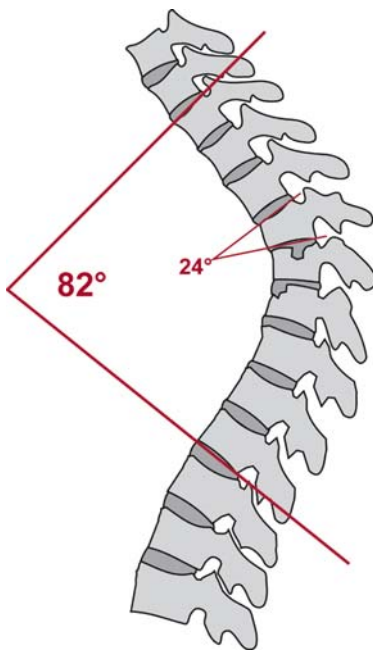
The distinction from normal findings can occasionally prove difficult. Are irregular endplates sufficient for diagnosing a case of Scheuermann's disease? Does one Schmorl node count as Scheuermann's disease? How many wedge vertebrae with a particular angle are needed for confirming the diagnosis?



**Fig. 3.54.** Radiographic changes in the thoracic spine of a 13-year old boy with Scheuermann disease, including apophyseal ring herniation, intervertebral disk narrowing and wedge vertebrae (arrows)



**Fig. 3.55.** Lumbar Scheuermann's disease in a 14-year old girl. Note the apophyseal ring herniation (arrows) on vertebral bodies L1 and L2 and the kyphosing in this area



■ **Fig. 3.56.** Measurement of the wedge angle and overall kyphotic angle in Scheuermann disease: Straight lines are drawn through the endplates: through the inferior and superior endplates of the same vertebra for measuring the wedge shape, and through the endplates of the two vertebrae that are most severely tilted towards each other for the overall kyphotic angle. The angle formed by these two lines is the overall kyphotic angle

The statements in the literature are very contradictory. We apply the following rules:

- In the *thoracic area* the overall kyphotic angle and the clinical findings are the deciding factors. Thoracic Scheuermann's disease is diagnosed, regardless of the radiographic changes, if the overall kyphotic angle exceeds  $50^\circ$  and the kyphosis is clinically fixed. If the x-ray shows two or more wedge vertebrae of  $>5^\circ$  or Schmorl nodes / apophyseal ring herniations, the condition can be diagnosed even if the overall kyphotic angle is less than  $50^\circ$ . Disk narrowing and endplate irregularities are not sufficient in themselves for confirming the diagnosis. Since the height of the disk on the x-ray depends greatly on the projection this is difficult to assess.
- In the *thoracolumbar and lumbar area* the diagnosis can even be made with a wedge vertebra of  $>5^\circ$  or one Schmorl node / apophyseal ring herniation. While the measured kyphotic angle is not relevant for diagnostic purposes, it is useful for assessing the severity and prognosis of the condition.

It can sometimes prove difficult to distinguish between wedge vertebrae caused by Scheuermann disease and those resulting from *compression fractures*. The following findings on the lateral x-ray suggest a compression fracture:

- irregularities at the anterior edge of the vertebral bodies, possibly a pre-existing fragment,
- absence of disk narrowing above the wedge vertebra,
- smooth boundary of the upper endplate of the affected vertebral body.

The patient's history obviously plays an important role, although it should be borne in mind that trauma details reported by adolescents can be misleading in both positive and negative senses. Not every adolescent considers a fall from a moped to be »trauma«. On the other hand, patients may be keen to associate back pain with a particular event that was certainly not capable of causing injury.

### Associated diseases

#### ■ *Scoliosis*

Over 50% of patients with Scheuermann disease also suffer from *scoliosis* to a greater or lesser extent [10]. This is directly connected to the Scheuermann's disease and has nothing to do with idiopathic scoliosis. It occurs when wedge vertebrae develop asymmetrically, i.e. a lateral wedge is formed. This mechanism contrasts strongly with the process involved in the development of idiopathic scoliosis, where the vertebral bodies grow faster anteriorly than posteriorly and create extra space by rotation (► Chapter 3.1.4). Since diminished growth occurs on the anterior side in Scheuermann disease, the associated scoliosis involves less rotation compared to the idiopathic form, and it is obviously not lordotic (as in the idiopathic form) but rather kyphotic. The prognosis for Scheuermann scoliosis is relatively good and severe lateral curvatures rarely develop.

#### ■ *Spondylolysis*

Adolescents with Scheuermann's disease are also associated with an increased incidence of *spondylolysis* [10]. This particularly applies with the thoracic form, which is compensated for by hyperlordosis, thereby increasing the pressure on the interarticular portion of L5. The spondylolysis is known to be caused predominantly by mechanical factors (► Chapter 3.1.6).

### Course, prognosis

- *Fixed, thoracic kyphoses of less than  $50^\circ$*  do not represent a problem in adulthood, and back pain is no more common or stronger than in normal individuals. However, the outwardly visible deformity can be a psychological burden (particularly for women).
- In cases of *fixed, thoracic kyphoses of more than  $50^\circ$* , back pain is no more frequent, but is likely to be more intense than in normal individuals. Such patients tend to choose physically less demanding occupations, while lung function is only impaired in very severe kyphoses [11].
- *Kyphosis of more than  $70^\circ$*  can also be progressive in adulthood [5].

— Thoracolumbar and lumbar forms of Scheuermann disease are often painful in adolescence, and the prognosis in terms of pain during adulthood is poor because of the flat back or lumbar kyphosis [3]. Lumbar kyphoses shift the center of gravity anteriorly, which has to be compensated for by increased postural work by the paravertebral muscles.

### Treatment

#### Available options:

- Physiotherapy
- Brace treatment
- Surgery

### Physiotherapy

Physiotherapy is only indicated if fixed kyphosis and/or radiological evidence of Scheuermann disease are present during the pubertal growth phase. While the kyphosis remains flexible and no radiographic changes are apparent, the patient is merely suffering from a postural abnormality rather than Scheuermann's disease. *It is more effective to manage postural abnormalities by encouraging the patient to practice some sporting activity than by expensive physiotherapy.* Adolescents rarely enjoy physiotherapy and are almost never able to perform regular exercises on a daily basis. It is more useful to persuade adolescents to practice a sport that they really enjoy. The specific sport involved is of secondary importance. Active, corrective physiotherapy is indicated, however, in a case of fixed kyphosis. The only inappropriate sports are rowing, cycling with drop handlebars (Fig. 3.57) and weight-lifting.

### Brace treatment

Brace treatment should be considered for a thoracic kyphosis of more than 50° in a patient who is still

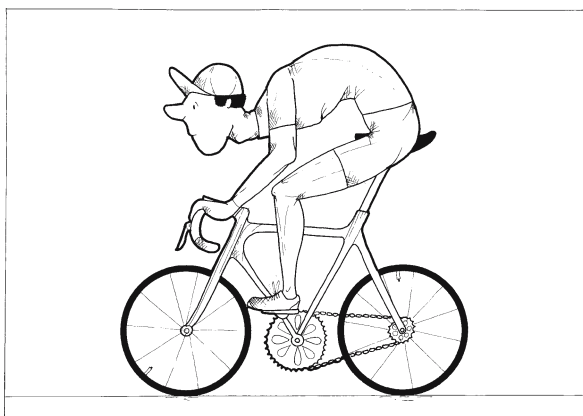


Fig. 3.57. Inappropriate sports for patients with Scheuermann disease include cycling in a racing cyclist's position

growing. In contrast with scoliosis, a kyphosis can not only be stabilized by brace treatment but also corrected in a patient with sufficient growth potential [4, 8, 14]. The wedge vertebrae are straightened by the compensatory growth of the anterior sections [7] (Fig. 3.58). Of course, a precondition for a successful outcome is good compliance, i.e. the brace must actually be worn. Experience has shown that compliance is rarely adequate. Possible braces for thoracic Scheuermann disease are straightening braces with a three-point action (e.g. the Gschwend brace) or extending braces with a neck ring (e.g. the Milwaukee brace). However, we generally use the smaller Becker brace (Fig. 3.59) fitted with a recl-

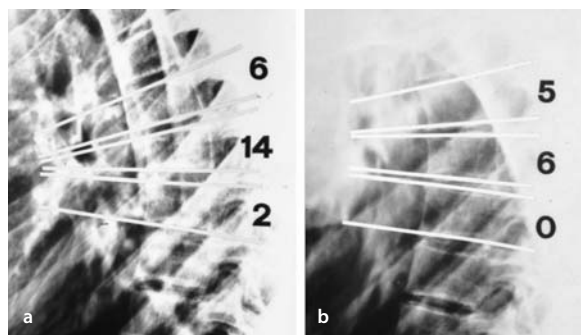


Fig. 3.58a, b. If growth potential is still present, wedge-shaped vertebral bodies can still be straightened out with brace treatment. a Wedge vertebrae in Scheuermann disease in a 14-year old girls. b Restraightened vertebral bodies two years later, after 18 months of brace treatment (figures refer to wedge angle in degrees)

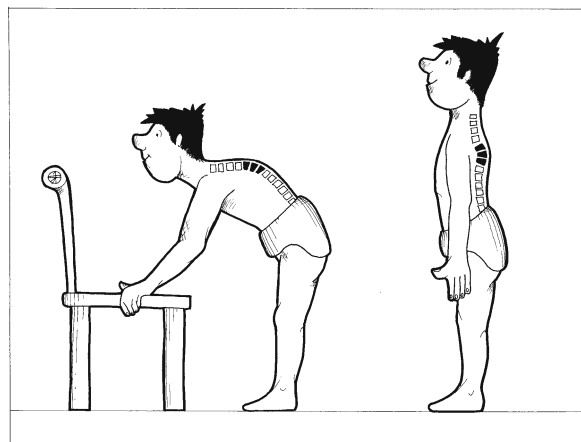
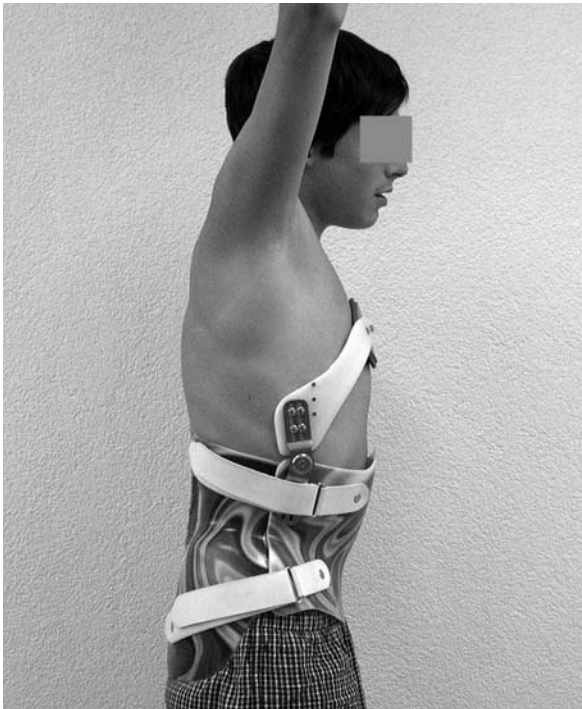


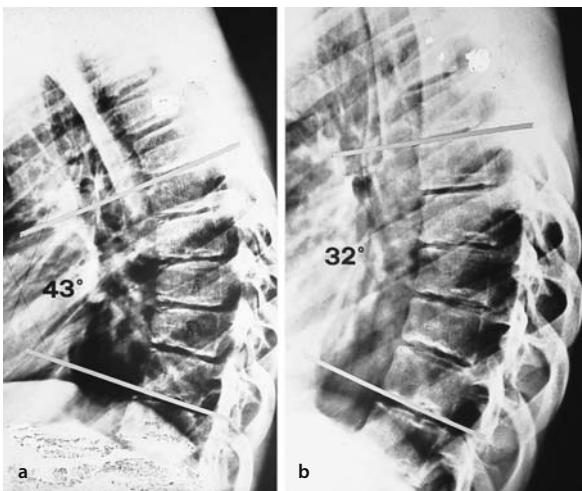
Fig. 3.59. Principle of Becker brace preparation for the treatment of thoracic Scheuermann disease. Only when the brace kyphoses the lumbar spine to a substantial extent is the patient forced to straighten his thoracic spine otherwise he will fall forwards. For the preparation of the cast (whether as a case for a plastic brace or a definitive plaster brace), the patient must support himself by placing his hands on a chair to ensure adequate kyphosing of the lumbar spine. The brace should not extend up as far as the apex of the kyphosis, but should end roughly at the level of the lower end vertebra of the kyphosis so that the patient is able to straighten up

### 3.1.5 · Scheuermann's disease

nation bracket. The principle of this Becker brace relies on its being fitted while the patient's lumbar spine is in maximum kyphosis. At the back the brace extends only to just *below* the start of the kyphosis. While the brace is worn, the center of gravity is shifted forward by the kyphosing of the lumbar spine, forcing the patient actively to straighten his thoracic spine to prevent himself from toppling forward. Passive straightening can



■ Fig. 3.60. Brace for thoracic Scheuermann's disease with an adjustable reclination bracket



■ Fig. 3.61a, b. 12-year old male patient with thoracic Scheuermann disease: a before brace treatment, b after 1 year of brace treatment. The kyphosis has returned to normal

also be achieved with the use of the reclination bracket (■ Fig. 3.60).

Results for brace treatment with good compliance: 2/3 improved (■ Fig. 3.61), 1/3 unchanged or deteriorated [7, 14] (■ Fig. 3.62). However, a certain amount of criticism is also now being aimed at brace treatment, calling its effectiveness into question [17], primarily because of the unknown factor of compliance. Authors rightly complain that the (few) existing studies are inadequately controlled. Since the kyphotic posture often represents a protest against the parents, the intrinsic motivation to correct it is sometimes completely lacking. A brace is not effective from kyphoses of 70° or more [14]. If optimal compliance is desired, a plaster cast must be prepared in a similar manner.

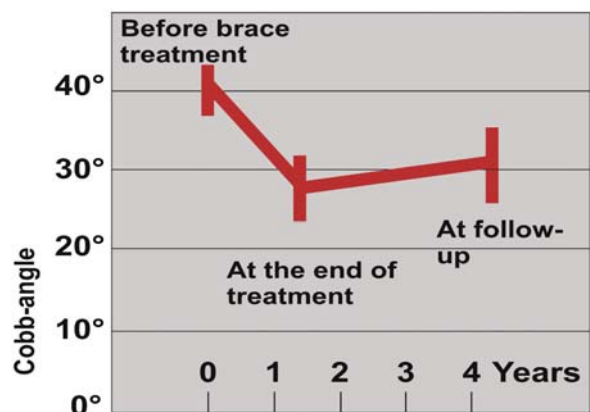
A lordosing 3-point brace can be used for thoracolumbar and lumbar Scheuermann disease. Since the prognosis in this form of the disease is poor in relation to later back pain, we tend to use a *cast brace*, prepared while the patient is in a position of ventral suspension. This will enable the lumbar kyphosis to be corrected back to lordosis while the patient is still growing (■ Fig. 3.63).

#### — Brace wear period

In contrast with idiopathic scoliosis, a brace used for the treatment of kyphosis need only be worn until the kyphosis is corrected and not until the completion of growth. Subject to good compliance, this usually takes 1 year.

#### — Clinical and radiographic follow-up

AP and lateral x-rays of the complete spine are needed to decide whether brace treatment is indicated. When the brace is ready, its effect must be checked radiologically by lateral views. Clinical checks are then arranged every 3 months, and x-rays should be recorded every 6 months (lateral only) until the patient is weaned off the brace.



■ Fig. 3.62. Results of brace treatment in Scheuermann disease: In contrast with scoliosis, a genuine correction that persists even after completion of treatment can be achieved with the brace





■ Fig. 3.63. Example of the correction of a lumbar kyphosis in Scheuermann disease after correction with a lordosing cast prepared while the patient is in a position of ventral suspension (► Chapter 3.1.12). a Before treatment, b after 6 months in a cast brace

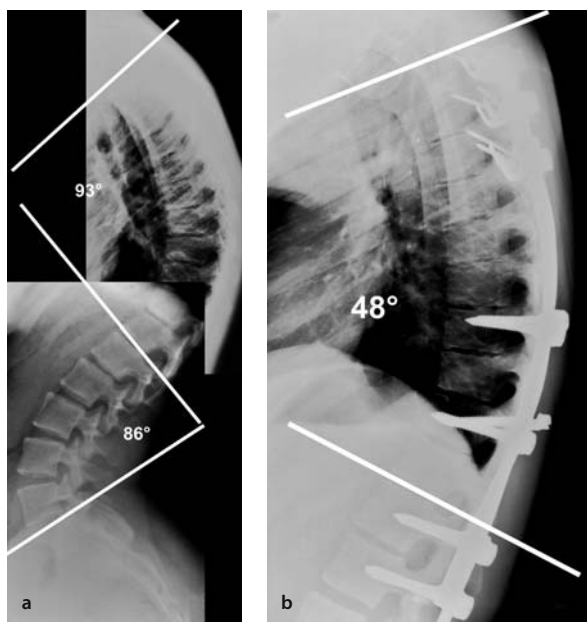
### Surgical treatment

This is only rarely required in Scheuermann's disease, since surgery is appropriate only if a thoracic kyphosis exceeds  $70^\circ$  and is generally indicated for cosmetic reasons. The prognosis in respect of symptoms is relatively good. For lumbar kyphoses on the other hand, an operation tends to be indicated for medical reasons since persistent and significant symptoms are usually present in cases of severe lumbar kyphoses. Surgical procedures can involve posterior or anterior approaches. While our practice in the past has involved the combination of anterior and posterior approaches, we now generally employ a purely posterior approach with wedge osteotomies and thereby create space for the posterior compression. This correction is generally sufficient [13] (■ Fig. 3.64).

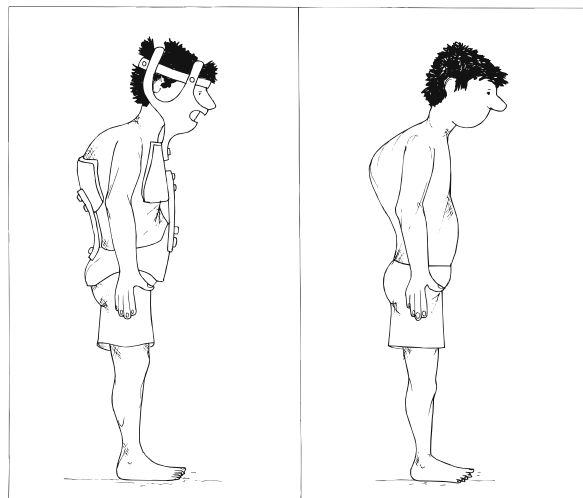
The possible complications of surgical treatment are similar to those for scoliosis surgery (► Chapter 3.1.4). In very severe kyphoses, the force of gravity works against all therapeutic efforts, and hyperkyphosis can occur in the non-instrumented area after correction of a kyphosis. For this reason, the instrumentation should, if possible, not only be used in the kyphotic area, but should extend to the start of the lordosis. No statistically evaluable data are available on the risk of neurological lesions, although the risk is probably similar to that for scoliosis surgery. Operations for kyphosis are more likely to be associated with the risk of vascular damage. On the other hand they involve compression rather than distraction. Other complications include infections, pseudarthroses, rod fractures, correction loss.

### Summary of treatment recommendations

The treatment recommendations are summarized in ■ Table 3.3.



■ Fig. 3.64a, b. Radiographs before (a) and 1 year after (b) surgical correction of a kyphosis in a 14-year old girl undergoing a dorsal tension-band wiring procedure (USS instrumentation). Osteotomies were performed from a posterior approach at the apex of the kyphosis at several levels



The curved back is a protest against pressure from the parents. Even a brace is often unable to straighten the spine in such cases...



**Table 3.3.** Overview of therapeutic recommendations for Scheuermann's disease

Overall kyphotic angle	Therapeutic recommendation
<50°	Physiotherapy (only for a <i>fixed</i> kyphosis, otherwise no treatment)
50°–80°	Brace treatment and physiotherapy
>80°	Possibly operation
Lumbar and thoracolumbar Scheuermann's disease	Possibly cast brace, indication for surgery for pronounced, fixed lumbar kyphosis

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## 3.1.6 Spondylolysis and spondylolisthesis

» If leaning back triggers a painful crisis, this may be a sign of lumbar spondylolysis «

### Definition

- **Spondylolysis:** Defect in the pars interarticularis on the vertebral arch
- **Spondylolisthesis:** Vertebral slippage, i.e. forward displacement of a vertebral body on the adjacent vertebra, possibly with additional tilting in the direction of a kyphosis
- **Linguistic derivation:** spondylos = vertebra, olisthesis = slipping

### Classification

Spondylolysis and spondylolisthesis in the area of the lumbosacral junction are the price we pay for our upright gait, and these conditions are not found in any other mammal. The lumbar spine is kyphosed in the quadruped vertebrate, and the lordosis of the lumbar spine in humans appears to be the precondition for the development of this clinical picture. Spondylolysis can, but does not necessarily, lead to vertebral slippage or spondylolisthesis. On the other hand, slippage is not always caused by a spondylolysis. *Spondylolisthesis* is traditionally classified according to the following causes [27]:

- isthmic (i.e. caused by a spondylolysis),
- dysplastic,
- degenerative,
- congenital,
- traumatic.

This classification is of very limited value. In isthmic spondylolisthesis dysplasia of the vertebral arch is also frequently present [17], indicating the absence of any clear-cut dividing line between the isthmic and dysplastic types. On the other hand, trauma plays an important role particularly in isthmic spondylolisthesis, making it almost impossible to differentiate between a single episode of trauma or repetitive events.

### Etiology

The following *etiological factors* are involved:

- mechanical,
- genetic.

The *mechanical* relationships in the area of the pars interarticularis are such that during *hyperextension* the lower joint facet of L4 comes into contact with the pars interarticularis of L5. Forced hyperextension causes pressure to be applied by the joint facet on the pars interarticularis, where a growth zone is located [12, 20]. Repeated trauma, for example caused by certain sports involving lumbar hy-

perextension, leads to fracturing of the pars interarticularis and thus to spondylolysis. The local morphology of the joint facets also plays a crucial role [6]. The more sagittal the facet is oriented, the greater the risk of a lesion [12].

A spondylolysis can also develop on a mechanical basis at the caudal end of a long fused section of spine. *Scheuermann's disease* is associated with an increased incidence of spondylolysis since lumbar hyperlordosis is usually also present in this condition because of the hyperkyphosis of the thoracic spine.

In addition to mechanical factors, *genetic factors* also play an important role. In the dysplastic type, in particular, up to 30% of first-degree relatives also have a pars defect [1]. An increased incidence is also found in certain races (e.g. among the Inuit [29]).

There are also isolated reports of a single traumatic event, rather than repeated trauma, causing the spondylolysis [22], although this generally occurs at a higher level than L5.

### Frequency

➤ The prevalence in the white population is 6.4%, compared to 1.1% in the black population [30]. A frequency of over 50% has been calculated for the Inuit [25, 29].

Among Caucasians, there is an increased incidence of 15% – 30% for practitioners of the following sports [11, 18, 19, 26, 28] (■ Fig. 3.65):

- gymnastics,
- ballet,
- javelin throwing,
- weight-lifting,
- football,
- swimming.

The incidence is also increased, at 21%, in patients with a cerebral palsy (i.e. spastic tetraparesis with flexion contrac-



■ Fig. 3.65. Spondylolysis occurs more commonly in sports involving pronounced reclinability of the back...

ture of the hip), because the flexion contracture is compensated by a lumbar hyperlordosis and because increased spinal movements are needed to control the trunk [9].

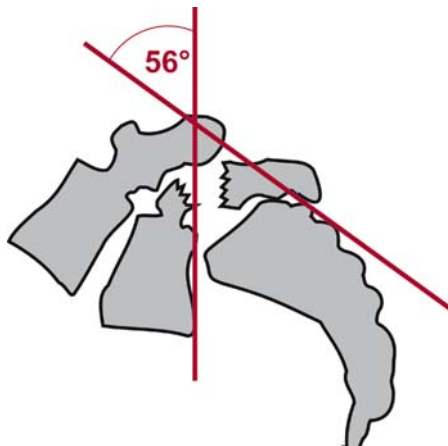
### Diagnosis, clinical features

Only a small proportion of patients with spondylolysis are actually symptomatic. Any symptoms that do occur generally fall into the category of lumbar pain. The pain is present during the day, occurs after prolonged sitting or standing and is typically movement-related. Reclining movements in particular are painful. On *clinical examination* we usually find that pain is elicited when the affected spinous process, i.e. generally L5, is pressed or vibrated. In the event of spondylolisthesis, pain typically occurs in the area of the hamstrings. The increasing kyphosis between L5 and S1 causes the center of gravity to be shifted forward. In order to correct this, the pelvic tilt must be countered by tension exerted by the hamstrings. As a result, the constantly tensed muscles become contracted and painful.

Not all symptoms and pain are caused by the spondylolysis and the contracted hamstrings. The defect in the vertebral arch causes instability and consequent painful tensing of the lumbar paravertebral muscles. This abnormal mobility is difficult to demonstrate on functional x-rays in the conscious patient (because of the muscle tensing), hence the widely differing opinions expressed in the literature on the extent and significance of this instability [12, 14, 20].

The clinical examination also includes the provocation of pain while the patient is in maximum reclination. A typical finding is the absence of symptoms while the patient bends forward, followed by sudden shooting pains at the lumbosacral junction on straightening up.

In *severe spondylolisthesis* or even *spondyloptosis* kyphosing occurs between the sacrum and the vertebral body L5, i.e. the lumbosacral kyphotic angle is less than 90° (■ Fig. 3.66). This leads to anterior displacement of

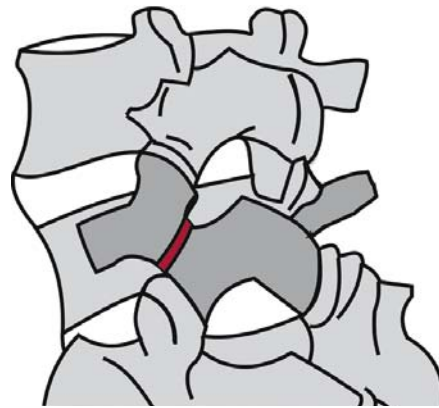


■ Fig. 3.66. Measurement of the lumbosacral kyphotic angle (see text)

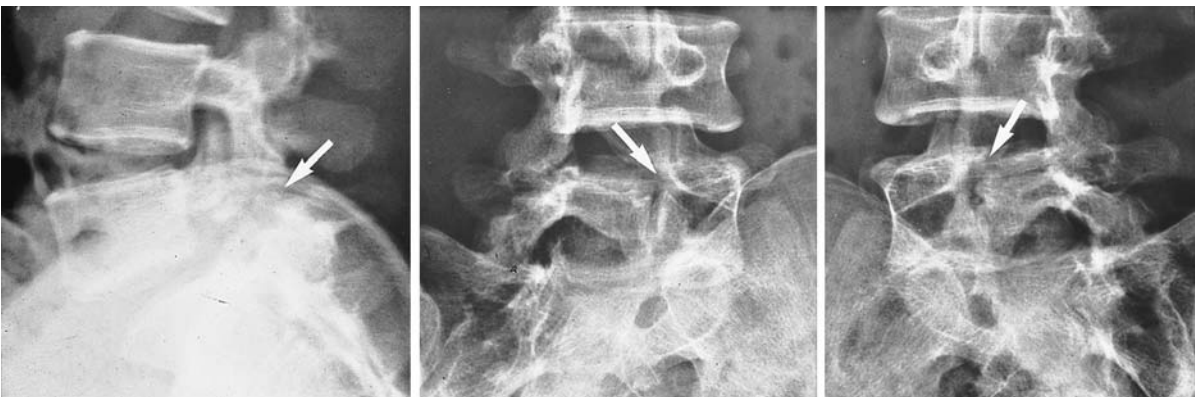
## 3.1.6 · Spondylolysis and spondylolisthesis



■ **Fig. 3.67.** Patient with spondylolysis. Note the straightening of the pelvis, the lumbosacral kyphosis and the compensatory lordosing of the thoracic spine



■ **Fig. 3.69.** Schematic presentation of the bone configuration on the oblique x-ray in spondylolysis. Darker section: The »dog«. The spondylolysis represents the dog's collar



■ **Fig. 3.68.** 16-year old male patient with bilateral spondylolysis of L5. Lateral view and oblique views from both sides

the center of gravity. The patient tries to compensate for this mechanically undesirable position, on the one hand by straightening the pelvis, which leads to tensing of the hamstrings, and on the other by lordosing the rest of the spine in order to return the center of gravity to a more dorsal position. This process results in painful contracture of the lumbar paravertebral muscles (■ Fig. 3.67).

### Radiographic diagnosis

In its initial, developing stages, spondylolysis is often not discernible on conventional x-rays. A *bone scan*, however, will show increased uptake before the bony defect is visible on the x-ray. If the area of lysis is sufficiently wide it can usually be easily detected on a *lateral x-ray*, although the radiographer should ensure that the x-ray is centered on the lumbosacral junction. On an AP view, the spondylolysis can only be seen if the pelvic tilt is eliminated. The spondylolysis is best viewed on *oblique x-rays* (■ Fig. 3.68 and 3.69): On these x-rays, recorded at an angle of 45°, the vertebral arch resembles a dog, with the transverse process forming the nose, the upper joint process the ear, the pars interarticularis the neck and the lower joint process the front

legs. The spondylolysis then appears as an oblique »collar« around the pars interarticularis (the »neck«) (■ Fig. 3.69). The lysis is also clearly visible on a *CT scan*. While the *MRI scan* is preferred nowadays because of the radiation exposure associated with CT scans, interpretation of the findings is slightly more difficult with these images.

### Site

In over 95% of cases, isthmic spondylolysis affects segment L5, and vertebral slippage may have occurred between L5 and S1. Only a small proportion of spondylolyses in adolescents or young adults are known to occur at different levels (L1–L4), and these are often traumatic in origin [22]. The degenerative spondylolisthesis that occurs in later adulthood, by contrast, tends to affect the segment L4/L5.

If spondylolisthesis occurs, we can subdivide the severity of the slippage into stages, and the classification described by Meyerding in 1932 [15] is still commonly used today (■ Fig. 3.70).

In addition to the slip angle, the extent of the kyphosis is also relevant (■ Fig. 3.66). This is measured as the angle

between the tangent to the back of the sacrum and the line passing through the lower edge of the 5th lumbar vertebra [31]. A kyphotic angle of less than 90° is considered pathological. Since these measurements are not very accurate, a definitive statement can only be made if distinct deviation is present [3].

The Meyerding classification of spondylolisthesis is shown in [Table 3.4](#) and [Fig. 3.70](#).

### Natural history

Most cases of spondylolysis remain asymptomatic throughout life. The incidence of spondylolysis was investigated in a study involving 500 schoolchildren and covering the years 1954–1957 [2]. The rates were 4.4% for 6-year olds, 5.2% for 12-year olds and 5.6% for 14-year olds. Additional spondylolisthesis was observed in 10% of cases. The same

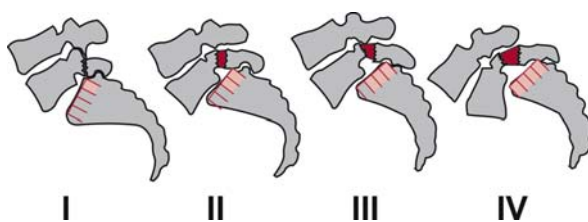
individuals were reviewed 45 years later. Spondylolysis was found in 6.0% of cases and additional spondylolisthesis in 28%. None of the subjects was symptomatic.

In our own hospital we diagnosed »spondylolysis« or »spondylolisthesis« in 31 patients between 1955 and 1974. The same patients were followed up in 1993, i.e. after an average of 28 years [7]. The average age at the time of first diagnosis was 13 years, and 12 patients complained of symptoms at that time. At the 28-year follow-up, three patients were unfit for work for reasons unrelated to the spondylolysis, while all of the rest were 100% fit for work. Eight patients complained of occasional slight load-related lower back pain, although this was not sufficient to cause them to consult a doctor or take any medication. Two patients complained of daily lower back pain for which they were receiving medical treatment. Both patients eventually underwent spinal fusion. One was subsequently symptom-free, while the other was fully fit for work despite the continued presence of symptoms. Thus, of the 31 patients, 21 (68%) were symptom-free after 28 years, 8 (26%) reported minor symptoms and 2 (6%) had required surgery.

Not infrequently a *scoliosis* (usually lumbar) also develops as a consequence of the spondylolysis, irrespective of whether the spondylolysis is unilateral or bilateral. The cause is possibly a muscle spasm. Spontaneous neurological lesions have also been reported in association with spondylolisthesis in isolated cases [13].

**Table 3.4.** Classification of spondylolistheses according to Meyerding

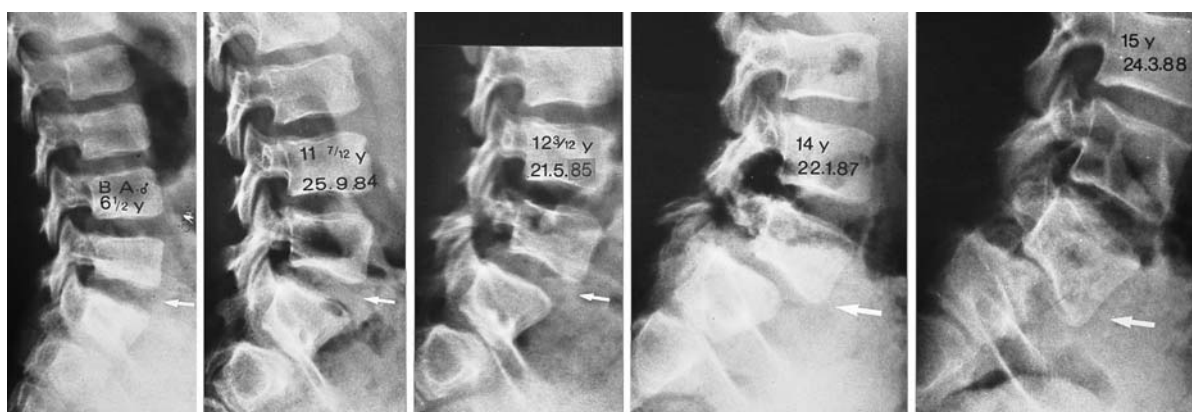
Grade	Displacement
Grade I:	<25%
Grade II:	25–50%
Grade III:	51–75%
Grade IV:	>75% slippage



**Fig. 3.70.** Staging (I–IV) according to Meyerding (see text)

### Progression of the spondylolisthesis

In the above-mentioned study of 500 schoolchildren, spondylolisthesis was found in 10% of the 6-year olds and in 28% of the individuals as adults. Since the literature does not include a single case of a spondylolytic spondylolisthesis in infancy, the vertebral slippage is almost always acquired during life in association with upright walking. Congenital cases of spondylolisthesis are, however, seen in neonates ([Chapter 3.1.7](#)). [Fig. 3.71](#) shows the course



**Fig. 3.71.** Spontaneous progression of an isthmic spondylolisthesis. At the age of 6 1/2 and 11 1/2 years the pars interarticularis is still intact. Spondylolysis is clearly visible by the age of 12 1/4, with an olithesis of

less than 25%. By the age of 14, the olithesis exceeds 30%. By the age of 15 the vertebral body L5 shows slippage of more than 50%



### 3.1.6 · Spondylolysis and spondylolisthesis

of a progressing spondylolisthesis in a patient between the ages of 6 and 15.

#### Treatment

The following therapeutic options are available for symptomatic patients:

- physical therapy
- brace or plaster cast
- surgery

#### Physiotherapy

Adolescents who develop symptoms caused by spondylolysis should be prescribed physical therapy, with the aim of strengthening and relaxing the tensed lumbar muscles. Providing advice on sporting activity is also important. Exercises involving reclinatio

#### Brace treatment

A brace treatment or the use of a support corset can be useful, since the resulting immobilization provides pain relief. The purpose of the brace treatment is to stabilize the condition and manage the pain. The brace should be fitted so as to provide a slight kyphosing action, such as that provided by a »Becker brace« (► Chapter 3.1.5). However, the kyphosing effect should not be as pronounced as that produced in the treatment of thoracic Scheuermann's disease.

#### Surgical treatment

The following surgical treatment options are available:

##### Spondylolysis

- direct screw fixation,
- posterolateral fusion,
- anterior fusion.

##### Spondylolisthesis

- fusion in situ,
- straightening and fusion with posterior instrumentation.

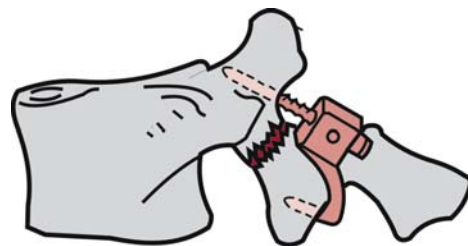
#### Spondylolysis and spondylolisthesis – grades I and II

**!** Since spondylolysis is a common condition and usually asymptomatic, the indication for surgery should be decided cautiously.

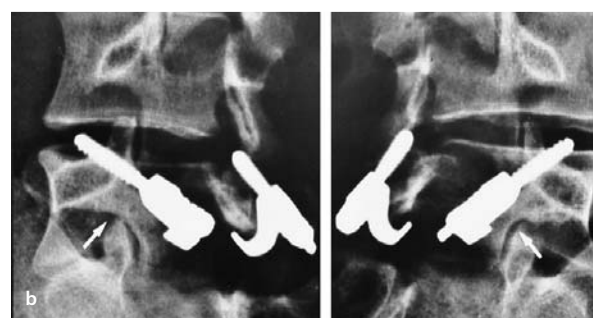
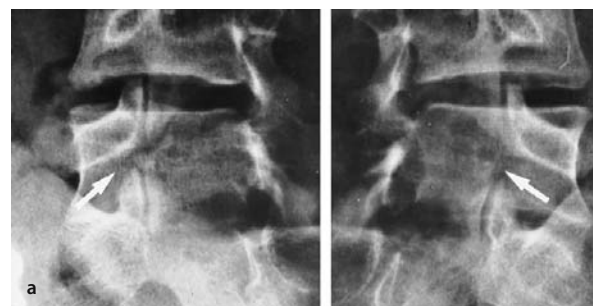
If the symptoms persist despite conservative management, surgery is occasionally indicated. A simple case of spondylolysis without major spondylolisthesis (maximum of 1 cm vertebral slippage) can be managed by direct screw fixation of the spondylolysis with a hook screw.

The principle of this operation is shown in ■ Fig. 3.72. The operation does *not* lead to stiffening of a segment since it actually constitutes a fracture or pseudarthrosis treatment. The spondylolysis is packed with cancellous bone and then a screw with a hook is inserted in such a way that the spondylolysis is compressed and the screw does not hamper bony contact at the narrow point of the spondylolysis. This method was described by Morscher in 1984 [16].

■ Fig. 3.73 shows a clinical example. We have performed this operation 73 times to date. The results for the first 33 patients were evaluated in 1992 after an average period of 3 years [8]. Half of the patients were under 20 years old at that time, while the rest were over 20. The results for those under 20 were good in almost 90% of cases, whereas this only applied to 65% of those over 20 years



■ Fig. 3.72. Hook screw according to Morscher [8, 18]. The screw is inserted over the spondylolysis. Autologous cancellous bone is packed into the actual spondylolysis. Tightening of the nut over the hook produces compression in the pars interarticularis defect



■ Fig. 3.73a, b. 16-year old female patient. Oblique x-rays bilaterally before (a) and 1 year after spondylolysis screw fixation (b). The bony consolidation of the spondylolyses is clearly visible



old. This difference is explained by the presence of disk degeneration in most of the over 20-year olds and the fact that the pain was caused not by the spondylolysis, but by the disk degeneration. A certain reduction of the spondylolisthesis can be achieved in isolated cases (■ Fig. 3.74). In many cases, screw fixation of the spondylolysis can probably prevent premature degeneration of the disk. Other studies have also reached similar conclusions [10, 21].

Since spondylolysis screw fixation constitutes a fracture treatment and can accomplish complete restoration of the situation we consider that the procedure is indicated in all cases in which conservative treatment has failed to produce any improvement after 3 months, the spondylolisthesis has progressed or proven instability is present. This is particularly to be expected with a dome-shaped deformity of the upper edge of the sacrum. In individual cases this dome-shaped change has returned to normal after screw fixation of the spondylolysis. In doubtful cases, an MRI scan should show whether disk degeneration is already present [23].

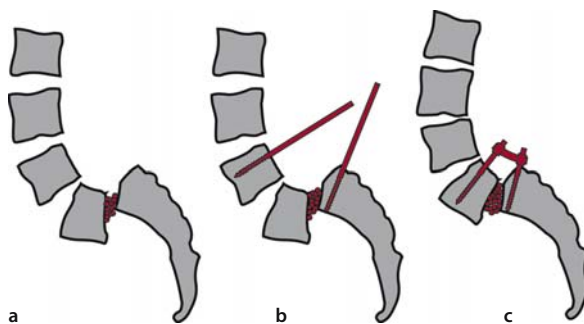
#### Spondylolisthesis (Grades III–IV) or spondyloptosis

If a spondylolisthesis of more than 50% is present (Meyering III or IV), surgical stabilization is likewise indicated. Provided no kyphosis is present (lumbar index over 90°), fusion in situ will be sufficient, either in the form of a posterolateral fusion or an anterior spondylodesis. We personally prefer the anterior fusion, since further progression cannot be prevented with a purely posterolateral fusion [24].

If kyphosis is present, i.e. a lumbar index below 90°, or if spondyloptosis has even occurred, correction is required. This involves a 2-stage procedure. In the first stage, the disk must be removed from the anterior side and the space filled with cancellous bone. In the second stage, the sacrum and the vertebral body L5 are grasped

from the dorsal side with transpedicular instruments. L5 is pulled back and, at the same time, lordosed. We use the »Universal Spinal System« (USS) for this operation. If the vertebral body L5 is so low that the screws cannot be inserted into its pedicles from the dorsal side, we fix the screws in the vertebral body L4 and perform the reduction maneuver accordingly. After successful reduction, we then transfer the screws to the vertebral body L5 and stabilize L5/S1 monosegmentally (■ Fig. 3.75 and 3.76).

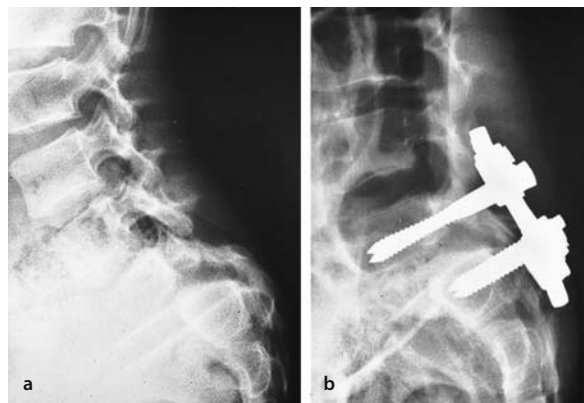
This procedure was developed at our hospital by Dick [4], and we have treated 41 patients to date by this method. The restoration of lordosis is a particularly important aspect of the reduction. The translation backwards should not be overdone, and a reduction of 50% is perfectly adequate. The risk of a nerve root lesion at L5 or S1 primarily arises from the posterior displacement of the vertebral body rather



■ Fig. 3.75a–c. Schematic presentation of the procedure for correcting a spondyloptosis (after Dick [3]). a Removal of the disk from a posterior approach and packing with cancellous bone. b Insertion of Schanz screws into vertebral bodies L4 and S1 (the pedicles of L5 cannot usually be reached initially). Reduction from the posterior side: particularly lordosing, the translation is only corrected by up to 50% (because of the risk of root compression). c Relocation of the Schanz screws from L4 to L5, fixation with vertical struts



■ Fig. 3.74a, b. 15-year old male patient. Lateral x-rays (a) before the operation. Grade I spondylolisthesis is present. (b) 1 year after spondylolysis screw fixation. The olisthesis was partially reduced by the operation

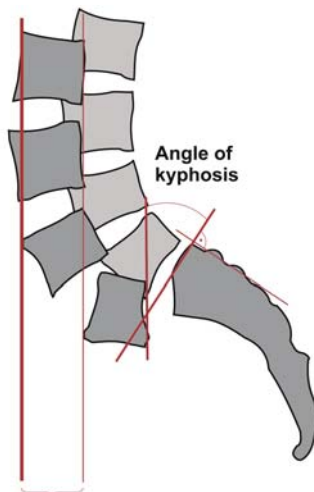


■ Fig. 3.76a, b. 18-year old female patient with grade IV spondylolisthesis. Lateral x-rays: a preoperatively; b 1 year postoperatively after concurrent correction from the ventral and dorsal sides

### 3.1.6 · Spondylolysis and spondylolisthesis

than the correction of the kyphosis. On the other hand, the kyphosis correction has a much greater effect than the posterior displacement on the forward shifting of the center of gravity of the upper body. For functional purposes therefore, the angular correction is more important than the rectification of the slippage (■ Fig. 3.77).

Follow-up examination of the first 18 patients confirmed that all patients were pain-free after reduction



Anterior displacement of the center of gravity

■ Fig. 3.77. Schematic presentation of the *shift in the center of gravity* in severe spondylolisthesis (grade IV) with kyphosis between L5 and the sacrum (*dark*). Normal spine: *light*

of the olisthesis from 67% to 37% and after correction of the kyphosis by 19°. While the preoperative neurological symptoms (in five patients) had disappeared, two patients (with no preoperative neurological problems) had a persistent foot levator muscle weakness after the procedure [5].

#### **Our therapeutic strategy for spondylolysis and spondylolisthesis**

The therapeutic strategy for spondylolysis and spondylolisthesis in our hospital is shown in ■ Table 3.5.

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■ Table 3.5. Therapeutic strategy for spondylolysis and spondylolisthesis

Growth age	
Spondylolysis with or without spondylolisthesis grade 0–II, no symptoms	No treatment
Spondylolysis with or without spondylolisthesis grade 0–II, typical pain	Physiotherapy, avoid lordosing exercises; if persists for more than 6 months, poss. brace, if it continues to persist poss. spondylolysis screw fixation
Spondylolysis, progressive spondylolisthesis grade 0–II	Spondylolysis screw fixation
Spondylolisthesis grade III without kyphosis	Fusion in situ
Spondylolisthesis grade III–IV with kyphosis or spondyloptosis	Reduction and ventral and dorsal spondylodesis
After completion of growth	
Spondylolysis with or without spondylolisthesis grade 0–II, no symptoms	No treatment
Spondylolysis with or without spondylolisthesis grade 0–II, typical pain, under 20-years old	Proceed as for growth age
Spondylolysis, spondylolisthesis grade 0–IV, over 20-years old with symptoms	If symptoms fail to respond to conservative treatment, poss. fusion in situ

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### 3.1.7 Congenital malformations of the spine

» *Crooked back, boy still young, poses problems for the lung.* «

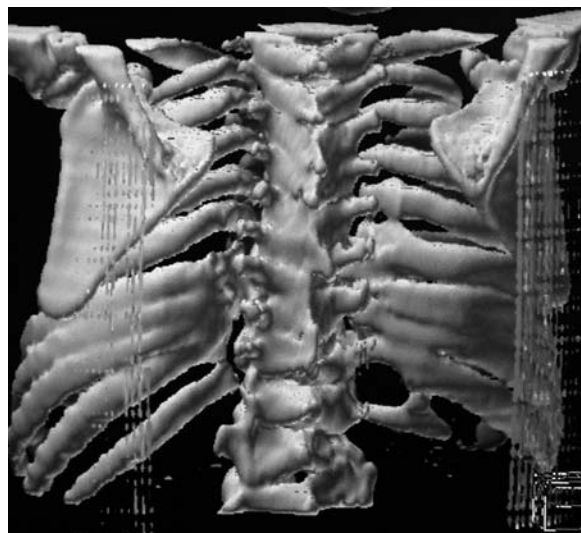
#### Definition

Congenital deformity of the axial skeleton at one or more levels leading to axial deviations in the sagittal (congenital kyphoses) and frontal (congenital scolioses) planes, possibly combined with rotation.

#### Etiology

Most congenital malformations of the spine are acquired during pregnancy. A hereditary or familial factor is involved in only around 1% of cases [7, 15]. Hereditary forms are usually associated with multiple anomalies. However, an increased incidence of idiopathic scoliosis has been observed in families of patients with congenital scoliosis [14].

For multiple congenital anomalies of the vertebral bodies (excluding meningocele), a risk of 5%–10% exists for subsequent siblings. One hereditary form is spondyl thoracic dysplasia described by Jarcho and Levin [9] with multiple bilateral segmentation defects, fused ribs and segmental aplasia (■ Fig. 3.78). This condition exhibits autosomal-dominant inheritance. Inherited multiple deformities likewise occur in Vacterl syndrome: in addition to vertebral anomalies, this syndrome is characterized by anal atresia, tracheoesophageal fistulas, esophageal atresia, renal malformations and dysplasia of the radius [10].



■ Fig. 3.78. CT (3D reconstruction) of the thorax and spinal column in Jarcho-Levin syndrome. Note the fusion of the ribs on both sides

### 3.1.7 · Congenital malformations of the spine

In the majority of congenital malformations, the causative factor is assumed to be toxic damage during the pregnancy. The damage must occur before the stage of ossification, i.e. in the initial stages of cartilage formation or earlier, and must therefore have taken place before the 10th week of pregnancy. The more extensive the malformation, the earlier the noxious factor has produced its effect. Most anomalies are probably formed during the 5th and 6th week of pregnancy [12]. Folate deficiency appears to play a role in the development of myelomeningocele [3].

#### Associated deformities

A range of deformities frequently occur in association with malformations of the spine:

##### ■ Fusion of the ribs

The ribs are often fused together to form a bony mass in the region of the malformation. This is typically observed on the side of an unsegmented bar (see below). The resulting scoliosis shows a strong tendency to progress.

##### ■ Spinal dysraphism

Around 20–30% of patients with congenital malformations show intraspinal anomalies [12, 16], most commonly intraspinal deformities with a unilateral unsegmented bar and a hemivertebra on the opposite side in the area of the thoracolumbar junction (52% diastematomyelias). The following *intraspinale deformities* can occur:

- Diastematomyelia, i.e. intraspinal bar formation,
- Cysts,
- Teratoma,
- Lipoma.

It is very important for such intraspinal deformities to be diagnosed prior to surgical procedures, since the risk of neurological lesions during operation is higher than usual when these anomalies are present [1].

Additional *associated deformities* also exist in around a third of patients with malformations of the spine. Heart defects have been observed in 7% of these patients, Sprengel deformity in 6%, cleft lip and palate in 4%, shortening of the extremities in 4%, clubfoot in 13% and a talus verticalis in 1% of cases. To these can be added hypoplasia of the mandible, renal aplasia or horseshoe kidney, uterine agenesis etc. Since anomalies of the genitourinary tract appear to be particularly common (up to 40%), an ultrasound scan of the abdomen and kidneys is indicated in every case of congenital scoliosis. Anomalies of the spine (particularly of the cervical spine) are more common in *certain syndromes*, for example in neurofibromatosis, Larsen syndrome, diastrophic dwarfism, mucopolysaccharidosis and also in spondyloepiphyseal dysplasia.

By definition, *myelomeningocele* is always also associated with a congenital malformation of the spine. In

most cases the anomaly only affects the open arches, although segmentation and formation defects in the form of hemivertebrae or wedge vertebrae of varying severity also tend to be present.

##### ■ Pulmonary dysfunction

In most cases of anomalies in the thoracic area, the spine is shortened, causing the thoracic space to be reduced as well. This especially applies in the presence of fused ribs, and the condition is known as thoracic insufficiency syndrome« [4, 6]. If excessive rotation of the vertebral bodies also occurs, the thorax becomes deformed, in turn producing an adverse effect on lung function. The stiffening of the affected section of the spine likewise contributes to a reduction in thoracic excursion and thus pulmonary function.

#### Occurrence

Almost no information is available about the epidemiology of congenital malformations of the spine, primarily because the anomalies in the rarest cases are hereditary and cannot usually be detected at birth. In a study involving the screening of 1,800 schoolchildren in the UK, two congenital scolioses were discovered [8], producing a prevalence of just over one in a thousand. While this figure is probably too high, malformations of the spine are very common, although many of them are harmless and do not produce any symptoms (and are then subsequently detected only by chance).

#### Classification

##### Classification based on the general shape

- Congenital scolioses
- Congenital kyphoses
- Combined malformations

##### Special types

- Arnold-Chiari malformation, meningocele
- Congenital spondylolisthesis.

##### Classification based on the type of malformation

- Structural defect
- Segmentation defect
- Combined malformations

The term »*Klippel-Feil syndrome*« says nothing about the type of malformation, but simply describes the location, i.e. the cervical spine. The term is very unspecific and includes all the bony deformities affecting the neck.

##### Classification based on the site of the lesion

- Occipitocervical (occiput down to C1)
- Cervical (C2–C6)



- Cervicothoracic (C7–T1)
- Thoracic (T2–T11)
- Thoracolumbar (T12–L1)
- Lumbar (L2–L4)
- Lumbosacral (L5 and S1)
- Sacral.

The most important distinction is between *formation defects* and *segmentation defects*.

**Formation defects**

Fig. 3.79 shows the various types of formation defect, each of which involves the incomplete formation of a vertebral body. A vertebral body that is dysplastic on one side is known as a wedge vertebra. If one side is completely absent, a hemivertebra is said to be present. The defect on the vertebral body can occur on the lateral, dorsal or ventral aspects. These malformations are therefore described as lateral, ventral or dorsal hemivertebrae or wedge vertebrae. A vertebral body lacking a middle section is known as a butterfly vertebra. The remaining part of the vertebral

body may show normal growth plates, but may also be fused with the adjacent segment, in which case one talks of an incarcerated hemivertebra or wedge vertebra.

**Segmentation defects**

In segmentation defects, the intervertebral space is not formed properly, which means that the growth plates are missing at the corresponding locations. If the complete intervertebral space is missing then a block vertebra is said to be present (Fig. 3.80). If the segmentation is only absent in a specific area of the vertebral bodies, this is described as an unsegmented bar. This bar can be located laterally, ventrally or dorsally. Since the bar is usually in an anterolateral position, the growth disorder caused by the unsegmented bar results in rotation of the affected vertebral bodies.

**Combined malformations**

Not infrequently segmentation and formation defects occur in combination. An anterolateral unsegmented bar combined with a hemivertebra on the opposite side is

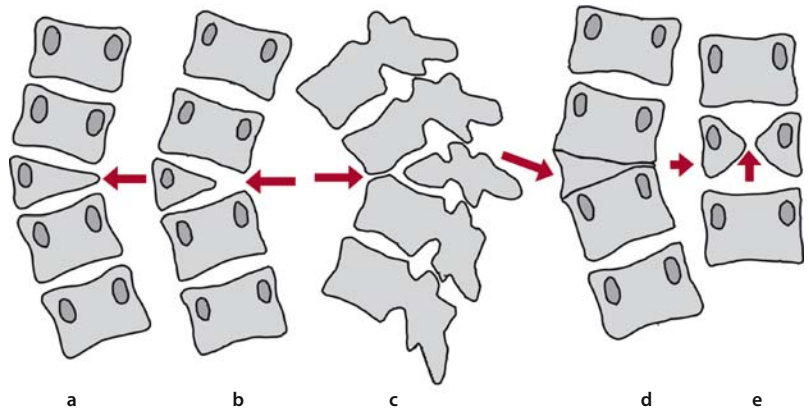


Fig. 3.79a–e. Formation defects: a wedge vertebra; b hemivertebra; c dorsal hemivertebra; d incarcerated hemivertebra; e butterfly vertebra

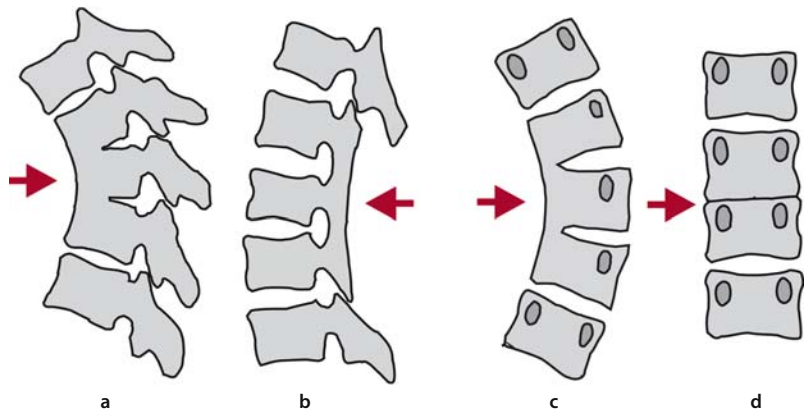


Fig. 3.80a–d. Segmentation defects: a ventral bar; b dorsal bar; c lateral bar (unilateral unsegmented bar); d block vertebra



## 3.1.7 · Congenital malformations of the spine

especially common. The prognosis for this combination is very poor. At cervical level also, combined malformations with dorsal bar formations and ventral formation defects commonly occur as part of a Klippel-Feil syndrome. However, these malformations can also affect any part of the spine.

**Natural history**

The following average annual progression rates were observed in a follow-up study of 242 patients with congenital scolioses [11]:

- Wedge vertebra: increase of 2.5° per year
- 1 hemivertebra: increase of 1.9° per year
- 2 hemivertebrae: increase of 2–3° per year (slightly higher in the lower thoracic spine area)
- Unilateral unsegmented bar: up to the age of ten 2° per year and subsequently 4° per year, in the mid-thoracic spine 5° per year, in the thoracolumbar junction area 6° per year
- Unilateral unsegmented bar and contralateral hemivertebra: increase of 10° per year
- Block vertebra: not a progressive deformity, but the relevant spinal section is shortened.

■ Table 3.6 summarizes the prognosis for the individual conditions. ■ Fig. 3.81 and 3.82 illustrate how varied the progression of the scoliosis can be depending on the underlying malformation.

**Diagnosis**

The malformation is primarily diagnosed during infancy, often as a chance diagnosis based on a chest or abdominal check x-ray. An outwardly visible deformity

is apparent only if rotation is present. Rotations are particularly likely to occur with an anterolateral unsegmented bar. An x-ray of the cervical spine should always be recorded in cases of a thoracic or lumbar anomaly, since associated malformations frequently occur in this area (up to 25%).

Close *radiographic monitoring* is important during early childhood, and annual x-rays are indicated until the pattern of progression is fairly clear.

Careful *neurological examination* is also essential. If a neurological lesion is suspected, an MRI scan, usually under general anesthesia, should be recorded. If diastematomyelia or a tethered cord syndrome is present (■ Fig. 3.83), neurological observation is especially important. As soon as a progressive neurological lesion is detected, the patient must be investigated with respect to possible surgical removal of the spinal anomaly.

An important and frequently neglected aspect is the *evaluation of lung function*. Since it is almost impossible to measure vital capacity in small children, the thumb deflection test is useful for estimating the extent of thoracic excursion (■ Fig. 3.84). Children with severe congenital scolioses are often short of breath, do not romp around appropriate to their age and show failure to thrive.

One particular form of congenital malformation of the spine is a *myelomeningocele* (► Chapter 3.1.10).

**Treatment**

The surgical treatment of congenital scolioses has undergone revolutionary changes in recent years with the introduction of the technique of *thoracostomy and straightening with the titanium rib according to Campbell (vertical expandable prosthetic titanium rib; VEPTR)* [4], which has

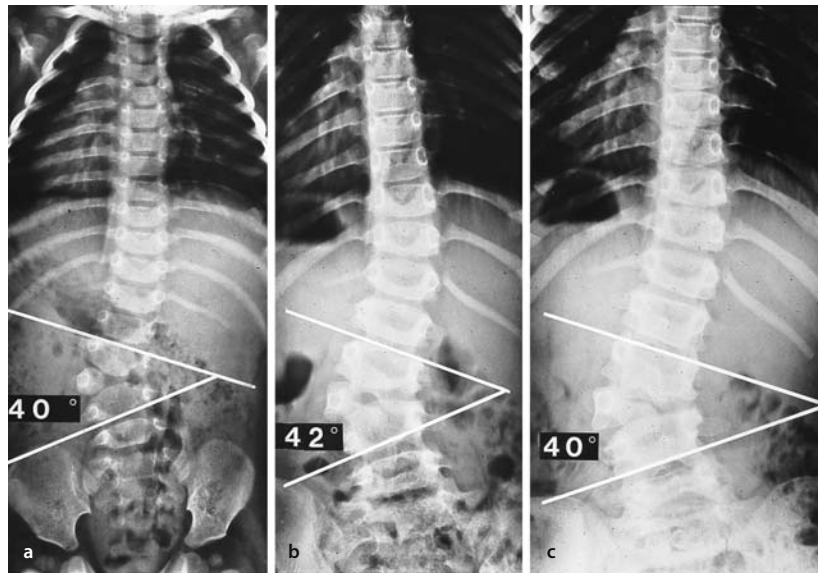
■ **Table 3.6.** Risk of progression for various types of spinal deformities (after McMaster and Ohtsuka 1982 [11])

Localization	Type of deformity					
	Block vertebra	Wedge vertebra	Hemi-vertebra, single	Hemi-vertebra, double	Unilateral unsegmented bar	Unilateral unsegmented bar and contralateral hemivertebra
	Progression in grade/year					
Upper thoracic spine	<1°	up to 2°	up to 2°	up to 2.5°	up to 4.5°	5°–6°
Lower thoracic spine	<1°	up to 2°	up to 2.5°	up to 3°	5°–6.5°	6°–7°
Thoracolumbar	<1°	up to 2°	up to 2°	up to 2.5°	6°–9°	>10°
Lumbar	<1°	<1°	1°	(–)	>5°	(–)
Lumbosacral	(–)	(–)	up to 1.5°	(–)	(–)	(–)

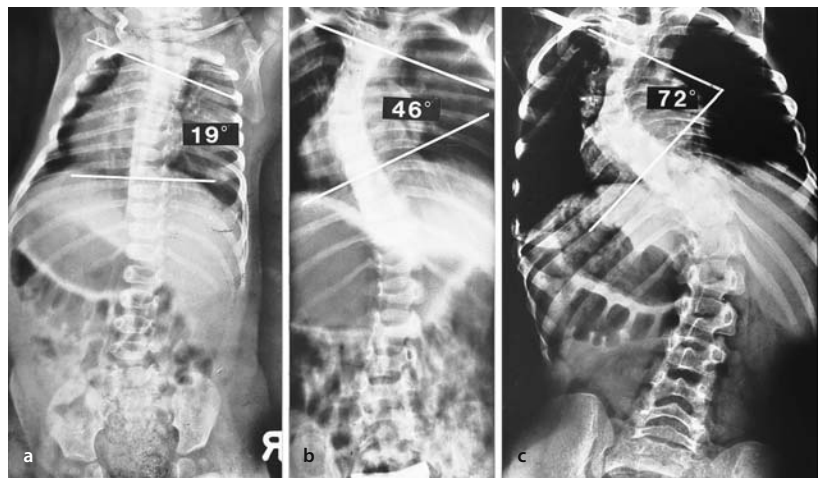
■ surgical treatment occasionally required

■ surgical treatment almost always required

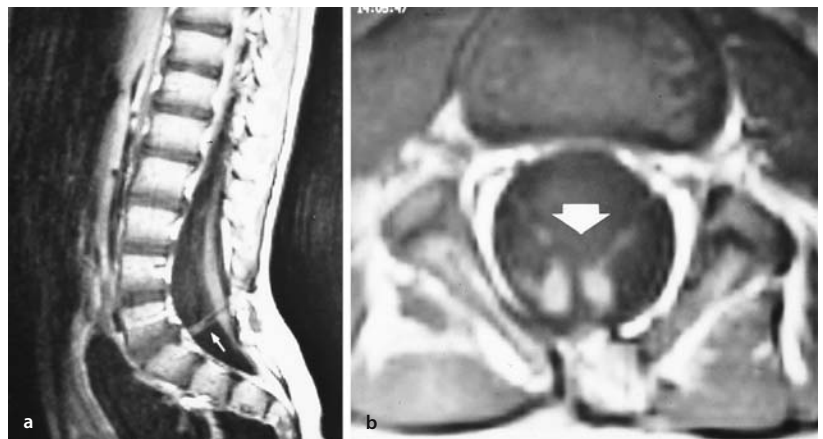
(–) too few cases to state figures



■ Fig. 3.81a–c. X-rays of a lumbar hemi-vertebra *without progression*: a at the ages of 1 year, b 5 years, c 10 years

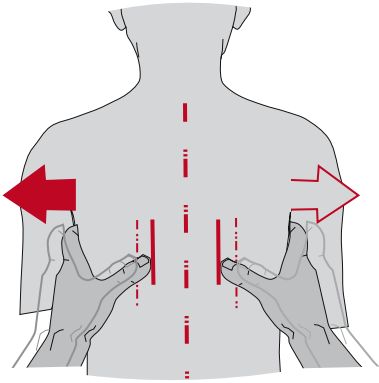


■ Fig. 3.82a–c. X-rays of a thoracic unilateral unsegmented bar *with pronounced progression*: a at the ages of 10 months, b 3 years, c 5 years



■ Fig. 3.83a, b. *Intraspinal malformation* in a 6-year old boy (MRI scans). This dysraphism (*arrow*) leads to a tethered cord syndrome, i.e. traction on the spinal cord with increasing growth

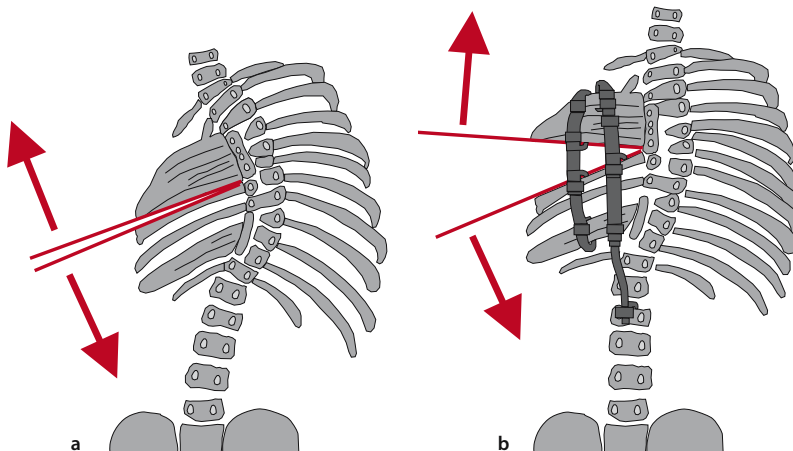
## 3.1.7 · Congenital malformations of the spine



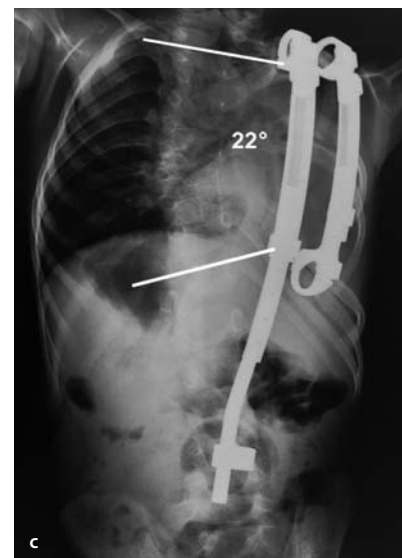
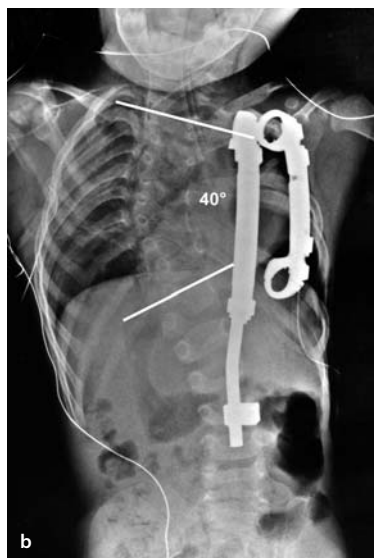
■ **Fig. 3.84.** *Thumb deflection test* [3]. The thorax is grasped from behind so that the thumbs are at the same distance from the spine on both sides. The patient is then asked to take a deep breath. The lateral movement of the thumbs is graded: 0= no excursion, 1=0.5 cm, 2=1 cm, 3 >1 cm

made previous treatments obsolete. The technique took 14 years to develop, and around 1500 children in the USA have since undergone this surgery at selected hospitals. In Basel we were the first hospital to introduce this technique, in 2002, under the direction of Dr. Campbell, and we have since acquired experience with approx. 40 patients (■ Fig. 3.85 to 3.87).

The primary objective in developing the instrumentation was to achieve separation of the fused ribs and subsequent distraction of the ribs. It gradually became apparent that this technique was also extremely effective in correcting other congenital scolioses. In the operation a special instrument is inserted between the ribs, which are then distracted. Depending on the individual situation, a rod can also be fixed to the lumbar spine, although the spine itself is not stretched, but rather the ribs. If the ribs have fused together on both sides, the instrument



■ **Fig. 3.85a, b.** Principle of the correction of congenital scolioses with the VEPTR instrumentation. **a** Preoperative situation with unilateral bar and rib fusions on the concave side of the scoliosis. *Red:* Planned division of the fused ribs. **b** Postoperative situation after osteotomy between the fused ribs and insertion of the VEPTR instrumentation followed by distraction



■ **Fig. 3.86a–c.** 20-month old child with rib fusions on the right, agenesis of the right lung and severe scoliosis. **a** AP x-ray. **b** Situation after

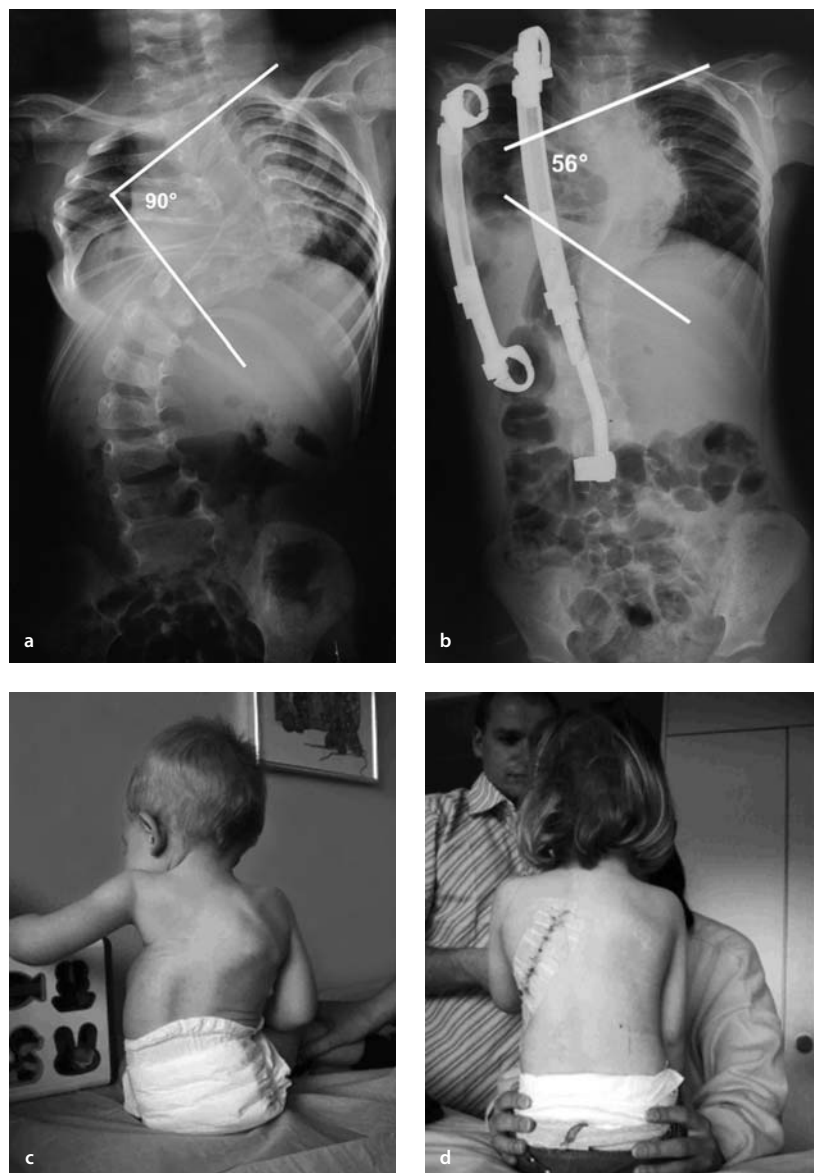
correction with the VEPTR instrumentation. **c** View 1 year after the first operation and distraction on two occasions

can also be used bilaterally. After a few months, the instrument must be expanded through a small opening to improve the correction. This procedure must be repeated several times.

This technique has several major advantages over conventional operations performed on the spine itself, which generally involve a fusion of the affected spinal sections. The stiff stabilization at the spine itself provokes a spontaneous fusion after about 3 years, even if no active fusion has been performed. The operation, and resulting expansion, are not performed on the spine, which reduces the risk of a neurological lesion. Nor is the spine stiffened. On the contrary, the shortened side of the spine is stimulated to grow. This not only makes the spine straighter but also longer. The lung volume is also increased (in some

cases by a substantial amount) [6]. The mobility of the spine is preserved because each breath produces a movement between the ribs (and the implant) and the spine. This does not happen when spinal rods are inserted, even without fusion. A particularly welcome feature is the stimulation of spinal growth, including at sites where the growth zones are lacking (with the unilateral bar). One of our colleagues, working together with Dr. Campbell, has confirmed the existence of this growth [5]. The distraction results in growth stimulation, as we have also learned from leg lengthening procedures.

The drawbacks of this technique are the need to repeat the lengthening and the high costs of the implant. The use of the instrumentation has its limitations in very kyphotic malformations.



**Fig. 3.87a–d.** Back of a 2-year old child with severe congenital scoliosis with fused ribs on the left. Preoperative x-ray and photograph (a, c). Clinical situation after correction with the VEPTR instrumentation and distraction on two occasions (b, d)



### 3.1.7 · Congenital malformations of the spine

The requirements for performing operations with the VEPTR instrumentation are very demanding. The following are required

- a pediatric spinal surgeon,
- a pediatric surgeon,
- a pediatric chest physician,
- a pediatric anesthetist,
- a pediatric intensive care unit,
- facilities for intraoperative motor and sensory spinal cord monitoring in very small children.

The latter facilities pose particular difficulties. The monitoring only works if there is excellent coordination with the anesthetist as most anesthetics affect the signals.

The thoracostomy procedure has rendered almost all other surgical treatments for congenital abnormalities of the spine obsolete. We believe that there are no longer any indications for stiffening or growth-retarding procedures. The purpose of these operations was always to keep the spine as straight as possible, while the problem of the small thoracic volume was ignored, and even deteriorated, in many cases.

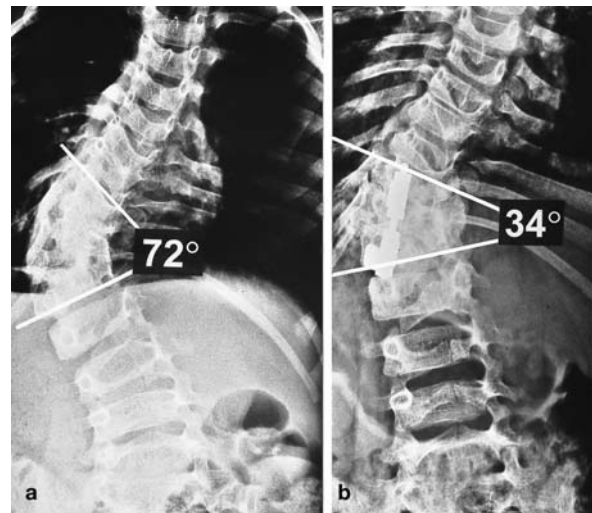
Hemivertebrectomy is indicated for hemivertebrae that cause decompensation of the spine or that are located posteriorly. The operation can be performed either exclusively from the posterior side or simultaneously from the anterior and posterior sides. The correction can then be performed from the posterior side using compression instrumentation. The surgeon should be careful to avoid constricting the nerve roots on the side to be compressed (■ Fig. 3.88). A hemivertebrectomy in the area of the cervical spine is particularly hazardous as the presence of the vertebral artery constitutes an additional complication (■ Fig. 3.89). Congenital spondylolysis/spondylolysis can also pose a problem in this procedure (■ Fig. 3.90).

**! Before carrying out a hemivertebrectomy in the lower thoracic or upper lumbar area, the surgeon must be aware of the course of the Adamkiewicz artery (a pre-operative angiogram is therefore essential in every case). Injury to this vessel can induce a vascular-related paraplegia or paraparesis.**

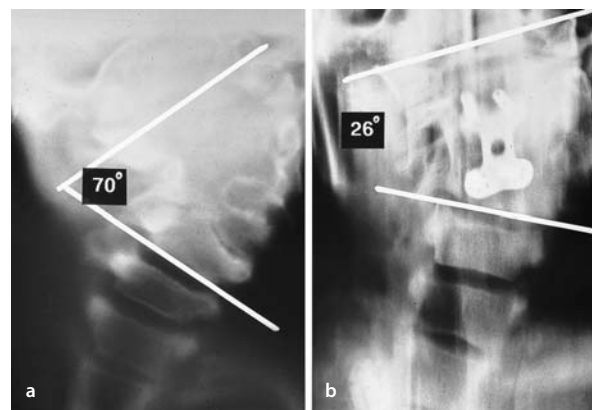
The principles for treating spinal deformities associated with *myelomeningocele* are described in ► chapter 3.1.10.

#### Complication risks

The most feared complication of surgical treatment is *paraplegia*, although this is almost never caused by direct injury to the spinal cord. The most hazardous maneuver is direct *distraction* (which we no longer use since the introduction of the VEPTR instrumentation). The existence of an intraspinal anomaly (which occurs in approx. 16% of cases [1, 2]) can lead to tension being exerted on



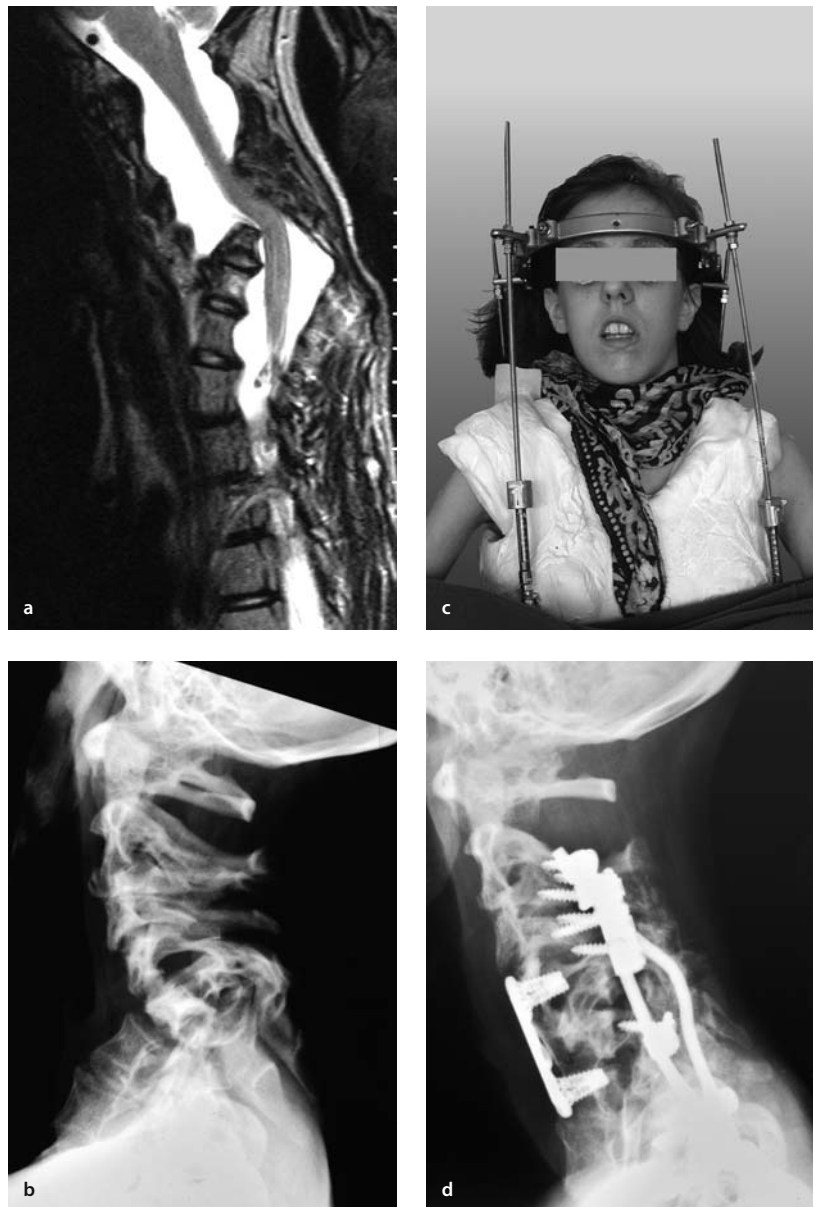
■ Fig. 3.88a, b. 6-year old boy with left-sided hemivertebra at T1 level and contralateral unilateral unsegmented bar and pronounced progression of the scoliosis. a Postoperatively; b 1 year after *hemivertebrectomy* from a ventral and dorsal approach and insertion of a compression rod on the convex side



■ Fig. 3.89a, b. Conventional tomograms of the cervical spine of an 11-year old girl with *Klippel-Feil syndrome* with multiple deformities and pronounced tilting of the head. a Postoperatively; b 2 years after *hemivertebrectomy* from a ventral and dorsal approach and insertion of a ventral titanium plate

the cord and thus to paresis or paralysis. During the early stages of surgical treatment at the end of the 1960's, we ourselves observed two cases of paraplegia after an operation with the Harrington distraction rod. We have since operated on around 200 patients with congenital scoliosis (out of approx. 600 observed cases), and no further cases of irreversible injury have occurred. One transient paraparesis occurred as a result of compression of the *Adamkiewicz artery*. However, intraoperative monitoring indicated the presence of this lesion, and removal of the compression rod produced a partial recovery even during the operation. Full remission was subsequently achieved after the operation.





■ Fig. 3.90a–d. Congenital spondylolysis. a Preoperative MRI. b Preoperative x-ray. c Photo with halo external fixator. d Post-operative x-ray

Vertebrectomies at the *cervical spine* level are particularly hazardous because the surgeon must be careful to avoid injury not only to the cord but also to the vertebral artery.

Another complication associated with posterior spondylodeses in very young patients is what is known as the *crankshaft phenomenon* [17], which involves the progression of the scoliosis, including rotation, as a result of the continuing growth of the vertebral bodies anteriorly. For this reason, a posterior spondylodesis should never be performed on its own in young patients, but the fusion should always involve a combined anterior and posterior approach, even if only one segment is affected.

### Summary recommendations for the surgical treatment of congenital scolioses

Our recommendations for the surgical treatment of congenital scolioses are summarized in ■ Table 3.7.

- These principles are highly simplified, and numerous factors must be taken into account in each individual case, including the extent of the curvature, progression, sagittal profile, rotation, the extent of the countercurve, compensation options, alignment, etc. Deciding on the appropriate treatment and selecting the best time for the operation requires considerable experience.

**Table 3.7. Recommendations for the surgical treatment of congenital scolioses**

Anomaly	Treatment
Wedge vertebra, block vertebra, butterfly vertebra	Generally no treatment
Single <i>lateral</i> hemivertebra of the mid and lower thoracic or lumbar spine	Generally no treatment
Single <i>dorsal</i> hemivertebra of the mid and lower thoracic or lumbar spine	Hemivertebrectomy from a posterior approach
Single hemivertebra of the cervical, upper thoracic or lumbosacral spine	Hemivertebrectomy from an anterior and posterior approach
Double hemivertebra, whole spine	Hemivertebrectomy from an anterior and posterior approach
Unilateral bar	Instrumentation with VEPTR
Unilateral bar and contralateral hemivertebra	Instrumentation with VEPTR, possibly hemivertebrectomy in addition
Intraspinal deformity	Neurosurgical resection

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## 3.1.8 Congenital muscular torticollis

### Definition

Congenital, unilateral contraction of the sternocleidomastoid muscle with inclination of the head towards the side of the shortened muscle, rotation towards the opposite side and facial asymmetry.

### Etiology, pathogenesis

For a long time it was assumed that congenital muscular torticollis was caused by birth trauma during delivery from a breech presentation. However, this theory does not explain why a pulled muscle should result in a permanent contraction of that muscle. Normally, a traumatic muscle lesion heals up more or less completely without any complications. Moreover, hardly any infants are delivered from a breech presentation nowadays, since a cesarean section is generally performed for this intrauterine position. But congenital muscular torticollis has by no means disappeared from the scene. Nor has the microscopic examination of biopsy preparations taken during surgical treatment revealed any form of hemosiderin deposits [9] such as would be expected after a pulled muscle. While congenital muscular torticollis is indeed often associated with a breech presentation, it has probably nothing to do with the birth process.

Muscular torticollis is also associated with other congenital anomalies, e.g. congenital hip dysplasia and club-foot. Microscopic examination reveals a fibrosis of the muscles that is sometimes seen after necrosis [9]. An abnormal intrauterine posture may be a contributory factor and result in a kind of compartment syndrome [4]. The occurrence of torticollis in families has been observed [5]. Ocular causes are not infrequently involved [11].

### Occurrence

Congenital muscular torticollis is relatively common, although corresponding epidemiological figures are not available. In a study in Japan involving 7,000 infants, the combination of head rotation, adducted hip and scoliosis was observed in 40 cases [8]. This would correspond to an incidence of approx. 0.5%, which also seems realistic in this part of the world.

### Clinical features, diagnosis

Congenital muscular torticollis can be diagnosed on the basis of purely clinical criteria. On palpation of the contracted sternocleidomastoid muscle, the doctor can frequently feel a lump or a kind of tumor, generally in the distal part of this muscle. This can affect both the sternal and clavicular parts of the muscle (■ Fig. 3.91). The infant's head is inclined towards the side of the contracted muscle, turned towards the opposite side and almost invariably shows asymmetry of varying degree, otherwise known as plagiocephaly. Head rotation towards the side

of the shortened muscle and head inclination towards the other side is usually significantly restricted. In older children, the sternocleidomastoid muscle is palpable as a tough cord, and it is usually easy to detect whether the clavicular part, the sternal part or both parts are shortened. A clicking sound is also occasionally elicited by a stretching manipulation [2]. Imaging is not usually necessary for the diagnosis and is only indicated if the head is in an oblique position and there is no clear contraction of the sternocleidomastoid muscle. X-rays of the cervical spine are often difficult to interpret in patients with muscular torticollis since the bony structures are distorted and the vertebral bodies are not shown in the standard projection. This factor should be taken into account in the evaluation of the x-ray. Secondary asymmetry of the atlas is also a possibility (■ Fig. 3.92).

! The facial asymmetry is not just present as a primary sign, but can also develop secondarily or become exacerbated if the torticollis persists for a prolonged period. Furthermore, the patient's brain becomes accustomed to the oblique position, which is eventually sensed as »straight« by the child itself. In such cases, the corrected, objectively straight, position is experienced as oblique.

### Differential diagnosis

The most important differential diagnosis is the *Klippel-Feil syndrome*. This condition covers all bony anomalies



■ Fig. 3.91a, b. Contracture of the sternocleidomastoid muscle in an 8-year old girl (a), particularly affecting the clavicular part. This tenses especially during rotation to the opposite side (b)



■ **Fig. 3.92.** Secondary asymmetry of the atlas in congenital muscular torticollis in a 6-year old boy. An os odontoideum is also present as a secondary finding

of the cervical spine, which can be relatively discreet, as found for example in the form of a unilateral dysplasia of the joint surfaces of the axis. However, the asymmetry of the joint surfaces can also occur secondarily after the prolonged presence of muscular torticollis. Another diagnosis to be differentiated is *paroxysmal torticollis* (also known as *Grisel syndrome*) [6, 10]. This rare condition particularly affects small children and manifests itself in the form of a severe torticollis that usually occurs after a peripharyngeal infection. Examination of the atlantoaxial joint in cadavers has shown the presence of a system of lymphovenous anastomoses in the epidural sinus which is responsible for the drainage of septic exudates in peripharyngeal inflammation. The hyperemia in this region explains the atlantoaxial subluxation. Since no lymph node stations are present, exudates spread out directly in the C1/C2 joint. Grisel syndrome usually resolves spontaneously, although immobilization is occasionally required, while atlantoaxial arthrodesis is indicated in very rare cases. Another important differential diagnosis is a tumor in the cervical spine (particularly an *osteoblastoma*) or a soft tissue tumor. The possibility of an *ocular cause* for the torticollis should also be considered. Examination by an ophthalmologist is indicated particularly if no restriction in passive mobility or shortening of the sternocleidomastoid muscle is found [11]. Additionally, a *unilateral hearing difficulty* can lead to a habitual oblique positioning of the head.

The differential diagnosis should also include the possibility of *secondary muscular torticollis*, which occurs

particularly in older children or adolescents, and also of course in adults, usually after a forced rotational movement or forced posture maintained over a prolonged period. In this case, the torticollis is attributable to a myogelosis, i.e. to a hardening of a band of muscle, although the hardening is always palpable in the area of the neck muscles (usually the trapezius) and not ventrally, i.e. not in the sternocleidomastoid. Pseudolocking of the intervertebral joints may play a role. Therapeutic measures include chiropractic manipulations, heat treatments, muscle relaxants and physiotherapy.

### Associated conditions

As mentioned above, congenital muscular torticollis is associated with a hip dysplasia or clubfoot in almost a third of cases. Congenital muscular torticollis is also part of the *prune belly syndrome* [7], which is characterized by a deficient abdominal wall, cryptorchism, renal malformations, congenital torticollis and frequently associated with hip dysplasia, clubfoot or vertical talus.

### Treatment, prognosis

The following options are available for the treatment of congenital muscular torticollis:

- physical therapy,
- orthoses (cervical collar),
- plaster fixation,
- surgical distal and/or proximal tenotomy.

### Conservative treatment

Conservative treatment for the neonate consists of physiotherapy, the aim of which is to stretch the shortened sternocleidomastoid muscle. This is not possible without »harassing« the infant to a certain extent. In a substantial proportion of cases it is possible to rectify the problem even during the first year of life. We no longer continue our former practice of administering cortisone injections. Since we assume that the fibrosis is due to a muscle necrosis arising from a compartment syndrome, cortisone treatment is not particularly appropriate as it achieves the opposite in actually promoting the necrosis. Nor do we primarily employ immobilization with a cervical collar or plaster fixation as stretching of the muscle might only occur in the extreme position, which is unacceptable for small children.

Since the prognosis for successful conservative treatment is not so good after the first year, the orthopaedist must then decide whether surgery is indicated.

### Surgical treatment

The surgical treatment involves a distal tenotomy of the clavicular and/or sternal part of the sternocleidomastoid muscle, depending on the section that is actually shortened. If possible, a non-shortened section should be left intact since the sternocleidomastoid muscle is very



important for the neck outline. If the section is removed completely, the shape will be altered. If both sections of the muscle are completely fibrotic, one section must be lengthened in a Z-shape. We no longer perform a proximal tenotomy in the area of the mastoid process. Since the facial nerve and its branches pass directly through the area of the attachment of the sternocleidomastoid, the risk of injury is relatively high. The argument cited in favor of a proximal tenotomy is that the scar is less visible on the hairline. However, we do not believe that this argument is particularly strong since scars in the anterior part of the neck almost always look very good because the skin is not very taut in this area.

The treatment is by no means completed with this (relatively minor and safe) operation. Intensive physical therapy is required postoperatively. The sternocleidomastoid muscle must be stretched for several months until scar formation has concluded. The scar in the muscle has a strong tendency to retract, and this trend must be counteracted very consistently. We support the physical therapy with a cervical collar that inclines the head towards the opposite side. We do not believe that plaster fixation is very useful postoperatively since the muscle is not adequately stretched when in a position of slight lateral inclination, thus ruling out intensive physiotherapy.

Older children's »world view« will also need to be straightened out. They should be encouraged to look in the mirror each day for a prolonged period and concentrate on a straight posture. This is another important task for the physical therapist. The prognosis for torticollis after surgical correction is good [3, 12]. If tumorous thickening is also present in addition to the muscle shortening, however, the prognosis is not so good [2]. In older children with pronounced facial scoliosis this will often persist and no longer correct itself spontaneously. For this reason, the surgeon should not wait too long before operating.

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### 3.1.9 Thoracic deformities

#### Definition

Symmetrical or asymmetrical protuberances or depressions in the area of the sternum, and possibly other thoracic sites. We distinguish between funnel chest as a hollow over the sternum and keeled chest, which involves forward projection of the sternum. Apart from these conditions, certain atypical thoracic deformities also exist.

#### 3.1.9.1 Funnel chest

#### Definition

Funnel chest involves a symmetrical or asymmetrical inward displacement of the sternum and adjacent ribs. It usually affects the central or caudal section of the sternum.

— **Synonyms:** Pectus excavatum, Pectus infundibuliforme

#### Etiology, pathogenesis

Although not an actual hereditary condition, there is a definite genetic predisposition and several genes appear to be involved. Funnel chest usually develops spontaneously. Secondary forms after thoracotomies, for example after cardiac surgery or tumor resections, rarely occur.

#### Occurrence

Funnel chest is relatively common, although epidemiological data are not available. In our own office we see around 10 new patients each year.

#### Clinical features, diagnosis

Funnel chest is diagnosed primarily on the basis of the clinical features. Retraction of the sternum several cen-



### 3.1.9 · Thoracic deformities

timeters behind the thoracic plane is observed. In most cases, either the proximal or the distal section is affected. Most deformities are asymmetrical (sternum 80%, rib cage approx. 50% [4]). Retraction in the distal part of the thorax is also often observed. In the majority of cases, the diagnosis can be made even during the first year of life. A simple parameter for monitoring the progress of funnel chest is the depression below thoracic level in centimeters. The condition can be better assessed by stating the relationship between thoracic breadth, thoracic depth and indentation. An objective picture can be obtained by raster stereography (▶ Chapter 3.1.4) or other surface measuring techniques.

*Cardiac and pulmonary function* are of crucial importance in assessing severe forms of funnel chest and will need to be investigated if the condition is very pronounced. An ECG, exercise ECG, echocardiography must be recorded and the vital capacity measured in such cases. However, a reduction in the performance of the heart and lung is detected in only the rarest of cases, i.e. usually in those patients in whom the thoracic diameter is reduced by more than half.

Psychological distress is much more frequently encountered in funnel chest than the functional restriction, although very few adolescents will openly admit that they feel uneasy about their condition. They are embarrassed by their funnel chest and tend unconsciously to conceal it by pulling the shoulders forward. This kyphoses the spine and involves the risk of developing *Scheuermann's disease*. In fact, we very frequently observe radiological and clinical signs of Scheuermann's disease in connection with funnel chest. In my experience, over half of patients with funnel chest also suffer from thoracic, and occasionally also thoracolumbar or lumbar, Scheuermann disease (■ Fig. 3.93).

Funnel chest is also relatively frequently associated with a *scoliosis* (15%–20% [5, 8]). Funnel chest is also typically seen in the *prune belly syndrome* [1]. Out of 40 patients a thoracic deformity was observed in 11 cases.

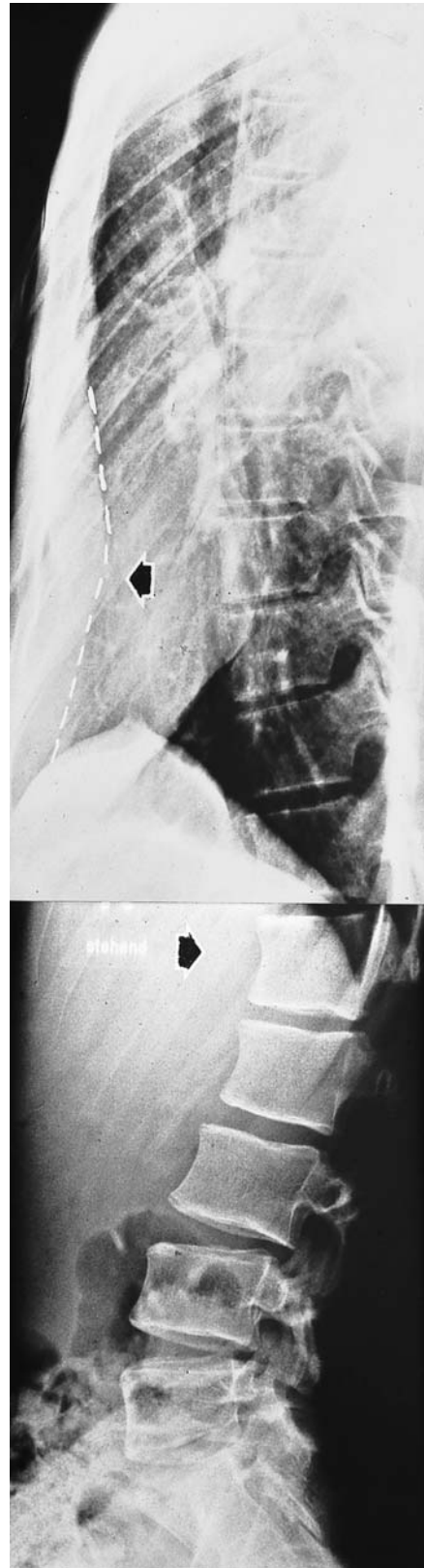
#### Radiographic findings

A lateral x-ray of the rib cage will clearly show the depression of the sternum (■ Fig. 3.93). The depth of the depression can also be correlated with the depth of the thorax. Additional imaging procedures (CT) are only required preparatory to surgery.

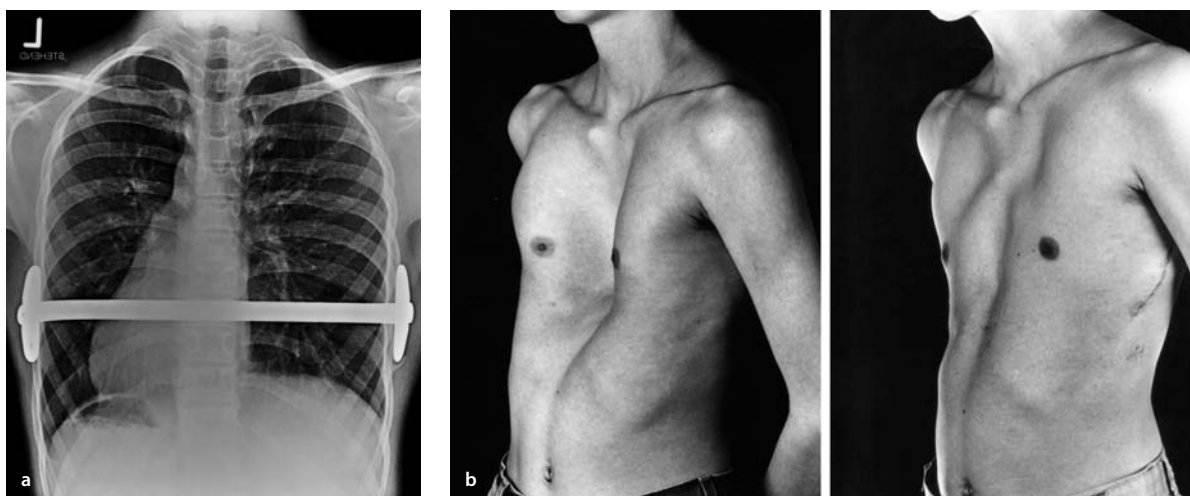
#### Treatment

Conservative treatments to date have proved ineffective. Recently we have been evaluating a treatment with a kind of *suction bell* applied to the chest daily for 30–60 min.

A clear *indication for surgery* only exists if cardiac or pulmonary function are restricted as a result of the reduction in thoracic volume. Such is very rarely the case



■ Fig. 3.93. 17-year old male patient with *funnel chest* (top arrow), extremely flat thorax and lumbar Scheuermann disease with corresponding lumbar kyphosis (bottom arrow). Bottom: lateral x-ray of the spine



■ Fig. 3.94a, b. Funnel chest correction by the Nuss method. a Thoracic x-ray with insertion of a curved bar in a 16-year old boy. b Clinical picture before (left) and after (right) funnel chest operation

however. If cardiovascular function is not diminished, the psychological distress can constitute a relative indication for surgery. Now that a satisfactory surgical technique is available in the form of the Nuss technique (see below), we also accept this indication in such cases. It is important, however, that the corresponding decision is taken not by the parents or the doctor, but solely by the patient, and only after, or towards the end of, puberty. With the exception of very severe deformities with restricted cardiovascular function, we never perform this operation before this point.

Previously employed surgical techniques involving correction performed directly on the deformity itself often showed unsatisfactory results, leaving a – frequently unsightly – residual scar directly over the sternum. Silicone implantation was also associated with certain complications (e.g. migration of the implant), and the acceptance level was not very good (probably as a result of problems with breast implants that were extensively aired in the media).

The Nuss method [7] significantly improved the situation (■ Fig. 3.94). In this thoracoscopic technique, two small lateral incisions are made to allow insertion of a curved bar that exerts outward pressure on the sternum. It can thus be described as a kind of internal splinting. The bar is left in situ for 2–3 years, after which time it can then be removed. We perform this operation jointly with pediatric (thoracic) surgeons because of the possible risk of injury to the pericardium. Nuss himself has reported on over 148 patients. Three patients required revision surgery, in one case because of a pneumothorax, and in the other two because of displacement of the bar [7]. Other authors have also reported low complication rates [6]. We

ourselves have operated on 30 patients to date and have not experienced any serious complications.

### 3.1.9.2 Keeled chest

#### ► Definition

Deformity of the thorax with keel-shaped protrusion of the sternum.

— Synonyms: Chicken breast, pectus carinatum

#### Etiology, pathogenesis

Keeled chest, like funnel chest, occurs as a result of a predisposition and is not an actual hereditary condition. Another possible cause is rickets, although this is very rare nowadays.

Secondary forms occur in

- scolioses with pronounced rib prominence,
- mucopolysaccharidoses (Morquio syndrome, etc.),
- collagen disorders such as Marfan syndrome and in
- barrel chest.

#### Occurrence

Keeled chest is rarer than funnel chest, although precise epidemiological data are not available.

#### Clinical features

Keeled chest involves a symmetrical or asymmetrical protrusion of the sternum (■ Fig. 3.95), either proximally or distally. The attachment point of the ribs is frequently asymmetrical, causing the whole sternum to be slightly tilted. A depression of the thorax is also occasionally present, generally in the distal section. This indentation then accentuates the appearance of the keeled chest.



■ Fig. 3.95. 14-year old boy with keeled chest



The psychological effects of thoracic deformities should not be underestimated. Terms involving comparisons with animals («Chicken breast») can prove especially hurtful

Keeled chest is *never* associated with a *functional restriction* as the heart and lungs are always able to develop sufficiently. The condition is therefore a purely *cosmetic problem*. Just as with funnel chest, however, keeled chest is frequently associated with *Scheuermann's disease*. Here, too, this association is probably due to the patient's unconscious attempts to conceal the deformity by pulling the shoulders forward and adopting a kyphosed spinal posture. This fact underlines the importance of the role played by posture in the development of Scheuermann's disease. Whether the mechanical deformation of the thorax is also responsible for the development of Scheuermann's disease remains doubtful. Although we know from our experience with vertebral fractures that a fracture of the sternum on the same side results in considerable instability and exacerbation of the kyphosis, keeled chest does not involve any significant reduction in strength, which means that this factor plays a subordinate role at most.

❗ **Try to avoid using expressions such as »pigeon chest« or »chicken breast« when talking to patients and parents. Like almost all expressions taken from the animal world and applied to humans, these terms have very negative connotations.**

### Findings of imaging techniques, radiographic findings

The extent of the keeled chest can also be measured objectively by raster stereography (► Chapter 3.1.4) or another surface measuring technique.

### Treatment

The following *treatment options* are available:

- conservative treatment with a brace,
- surgical correction of the thoracic wall.

The *conservative treatment* with a brace is much more promising for keeled chest than for funnel chest since, in the former condition, pressure can be exerted from outside to produce a genuine corrective effect. Good results have been reported in the literature [2] and we have been able to confirm this in our own experience. However, the brace is not always accepted by the patient and the treatment should be implemented primarily in younger children, as it will no longer have much effect by the time of adolescence. The brace only needs to be worn at night. I try to persuade parents of children with a pronounced keeled chest of the advantages of brace treatment as, in my view, surgical treatment is hardly

ever appropriate. Silicone implantation is not possible in keeled chest since it is already a case of »too much« rather than »too little«.

### 3.1.9.3 Atypical thoracic deformities

In addition to the common condition of funnel chest and the much rarer keeled chest, a series of other thoracic deformities also exists (the following percentages are based on a sample size of 1,410 chest wall corrections [3]):

- split sternum (0.5%),
- sternal synostosis (2.3%),
- condition after reconstruction of a diaphragmatic hernia (0.5%),
- extremely flat thorax (17%).

A split sternum and premature sternal synostosis always require treatment, and this should be left to the specialist as such deformities are extremely rare. *Flat thorax* is a very specific problem in which the thorax should always be considered in connection with the spine. We see flat thorax primarily in association with *idiopathic adolescent scoliosis*, but also in connection with *lumbar Scheuermann's disease* (■ Fig. 3.93). If treatment is indicated, this should also involve the spinal deformity. Correction of the thoracic deformity on its own is hardly ever indicated. The primary aim in correcting the spinal deformity is to restore the thoracic kyphosis. This can be achieved by a relatively sophisticated procedure involving combined anterior and posterior approaches. The problem of congenitally fused ribs is discussed in ► chapter 3.1.7.

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## 3.1.10 Neuromuscular spinal deformities

F. Hefti and R. Brunner

### Definition

Scolioses and kyphoses in neurological conditions (primarily cerebral palsy, myelomeningocele, poliomyelitis) or primary muscle disorders (muscular dystrophy).

### 3.1.10.1 Predominantly spastic paralyses

### Definition

Deformation of the spine associated with neurogenic disorders, usually cerebrally induced and involving altered muscle tone (particularly in cerebral palsy, but also in spinocerebellar degeneration). The clinical pictures are discussed in detail in ► chapters 4.7.1 and 4.7.2.

### Occurrence

While the reports in the literature concerning frequency show substantial differences, one thing is certain: both the prevalence and the severity of scoliosis are largely proportional to the severity of the neurological disorder. One study conducted in homes caring for patients with cerebral palsy observed an incidence of 67% of scolioses over 60° in those with severe tetraparesis [17].

### Clinical features

Patients with severe spastic tetraparesis lack trunk control and therefore find it difficult, or even impossible, to maintain an upright posture. While the extremities often show severe spasticity, the trunk may be hypotonic. In an upright position, the patients tilt to one side or adopt a kyphotic posture, often resulting in long C-shaped deformities. If the patients develop a hip flexion contracture, straightening of the legs during lying or standing causes the pelvis to be tilted forward and the lumbar spine to adopt a lordotic posture, which can likewise assume extreme proportions. Patients who are able to control their head and trunk to a certain extent try their best to hold their head as upright as possible, which can result in a compensatory countercurve in the proximal part of the spine (compensatory bending towards the opposite side in scoliosis or cervical lordosis in a kyphotic deformity). Any combination of these deformities is possible depending on the posture of the patients and the externally acting forces.

While the deformity in *younger children* can appear very pronounced in an upright position with no external guidance on both clinical and radiological examination, the mobility of the spine is largely preserved as a rule. In the *second decade of life* such spinal deformities become increasingly structurally fixed and can cause severe chronic pain. The *pain* is predominantly triggered by the ribs coming into contact with the iliac crest. Although



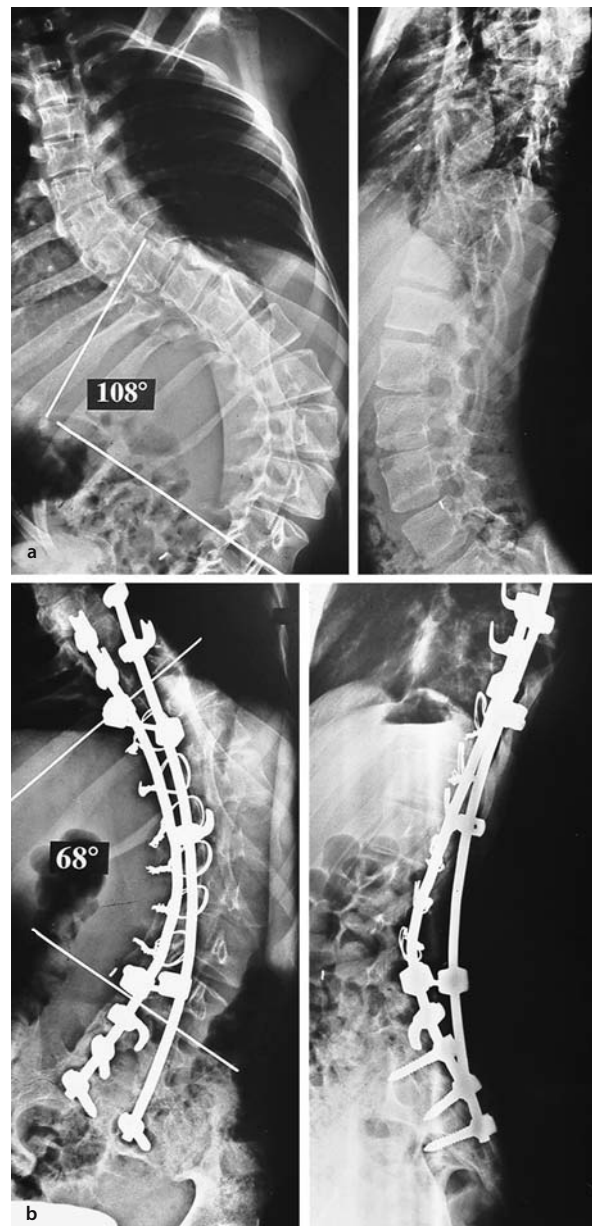
### 3.1.10 · Neuromuscular spinal deformities

progression of the scoliosis ceases once the thorax comes into contact with the ilium, the pain grows stronger and, with the passage of time, is even present on lying as the contracted muscles cause the ribs to rub against the iliac crest. While children with very severe spastic cerebral palsies are unable to complain about the pain verbally, this does not imply its absence. However, those who look after such patients generally notice when the children do experience pain.

#### Radiographic findings

Compared to an idiopathic scoliosis, a neurogenic scoliosis associated with cerebral palsy shows the following features:

- The scoliosis is in the form of a broad C-shaped arch: In patients with severely impaired balance and body control, the characteristic *countercurves* observed in idiopathic scolioses are *absent* (■ Fig. 3.96). This correlates directly with the patient's mental and neurological status. This lack of countercurves is most marked in patients who are unable to either sit or stand independently, whereas cerebral palsy patients who are capable of walking always have a countercurve of varying degree on both sides of the main curve, although they are often unable to straighten themselves out as well as patients with idiopathic scolioses.
- *Pelvic obliquity* is very characteristic of neuromuscular scolioses and is particularly pronounced in severe cases of cerebral palsy. Pelvic obliquity and hip dislocation can mutually influence each other. The hip on the higher side of the pelvis is particularly at risk since it is adducted. There is no statistical correlation, however, between the side of the hip dislocation and the direction of the pelvic obliquity [7].
- The *apex* of the scoliosis in cerebral palsies is usually at *thoracolumbar* level, occasionally at the low thoracic or lumbar level, but hardly ever at the level of T6–T9, as commonly found in idiopathic scolioses.
- The *rotation* of the vertebral bodies at the apex is *less pronounced* in neurogenic scolioses with a small to medium Cobb angle than in idiopathic scolioses, but *more pronounced* with Cobb angles over 50° [6]. While the correlation between rotation and the Cobb angle is linear in idiopathic scolioses, this is not the case with neurogenic scolioses, which show only slight rotation with a small Cobb angle, but disproportionately high rotation with a large Cobb angle.
- In contrast with idiopathic scolioses, neurogenic scolioses are frequently *associated* with a kyphosis. The kyphoses are usually thoracic and severe hyperlordosis is often present at the lumbar level. In certain patients the kyphosis is the dominating factor, overriding the lateral curvature in terms of severity. A lumbar kyphosis is also present in rare cases, which can pose particular problems for surgical treatment.



■ Fig. 3.96a, b. 19-year old patient with severe cerebral palsy and long-curved thoracolumbar scoliosis with minimal countercurves. a Preoperative AP and lateral views. b 1 year postoperatively after correction and stabilization with the Spinefix instrumentation and segmental wiring

#### Treatment

##### Therapeutic objectives

Most patients are so severely disabled that they are confined to a *wheelchair*. The seat of the wheelchair must take into account the problems associated with the sitting position and the spinal deformity and be adapted accordingly.

The treatment of the spinal deformity pursues the following *objectives* in these patients:



- Stabilize the spine, and thus the trunk, so that the patient can assume an upright position. Stabilization of the trunk usually also improves the head control, in some cases giving the patient some head control for the first time. When the patient is upright, the unstable trunk tilts to one side as a result of weak muscle tone. Gravity pulls on the trunk, exacerbating the deformity, which becomes increasingly fixed, particularly during growth.
- Correct the shape.
- Prevent exacerbation of the deformity.

### Conservative treatment

*Brace treatment* is possible provided the spine can be straightened sufficiently to allow the axial pressure to be deflected so that it is over the spine in the upright position. This goal can best be achieved if the *plaster cast* is prepared in a position of *hypercorrection*, because the patient will tend to spring back to his abnormal shape while wearing the brace. Brace treatment is indicated if the Cobb angle is between around 30° and 70°. No precise limit can be stated, since other factors unrelated to the severity of the scoliosis are also important, for example obesity, tolerability of the brace and the material, respiratory impediments, disorders of the airways and acceptance by the parents and caregivers. The brace should not be fitted too tightly at the thorax because of the need to allow chest movements for breathing. If a brace is indicated it must be worn whenever the patient is in an upright position, because it replaces the postural function of the trunk muscles and must counter the deforming force of gravity. If the scoliotic curvature is the predominant factor and the kyphotic tendency is minimal, we currently use a flexible brace that permits trunk movement but provides sufficient lateral stabilization (■ Fig. 3.97). In our experience, the use of such a brace rarely produces pressure points.

In order to improve the *head control* while sitting or standing, the *upper body* can be *inclined backwards* while the patient is in the wheelchair, producing a deckchair-like position. The head lies on a *headrest*, providing better head control. On the other hand, the patient's gaze is directed upwards, making contact with his environment more difficult. Stabilization of the trunk also improves head control, enabling an upright position to be adopted. Other options for improving head control are the wearing of a *cervical collar* or a *Glisson sling*. While these aids are accepted in cases of extremely abnormal postures, parents and caregivers tend to disapprove of them.

In *severe deformities*, the deformation of the spine causes posture-retaining force to be transferred to the brace at the apices when the trunk is in an upright position (sitting or standing), regularly resulting in pressure points. If the brace is widened at these points, the patient sinks further down, producing new pressure points or



■ Fig. 3.97. Dynamic brace in spastic cerebral palsy. This allows flexion movements yet still provides adequate lateral support

transferring the pressure onto the skeleton as the spine collapses completely and the thorax comes to rest on the pelvis.

At this point, the *limits of conservative treatment options* have been reached. The patient is no longer able to sit up, and the increasing asymmetry means that the patient can only lie on one side, considerably aggravating their nursing care.

### Surgical treatment

When deciding on the indication, extension of stiffening and the surgical procedure, we must make a basic distinction between *two situations* for patients with a cerebral palsy:

- patients who are able to walk
- patients who are unable to walk

Since the mental faculties parallel, to some extent, the motor skills in these patients, the requirements for mobility of the spine differ very widely, depending in each case on whether they can or cannot walk. A patient who is able to walk requires the ability to rotate the trunk for this purpose. In neurological diseases, spinal movement often increases to compensate for the stiff, spastic posture of the legs. In addition, the patient must be able to bend

and turn. The *therapeutic objectives* are accordingly very wide-ranging.

➤ The following *objectives* apply to *patients who are able to walk*:

- Correct the curvature (in order to improve balance),
- Prevent progression,
- Prevent decompensation.

The objectives here are similar, therefore, to those for the treatment of idiopathic scoliosis, except that the cosmetic aspects are not so important in this situation. Surgical treatment is indicated from a Cobb angle of approx. 50° if corresponding growth potential is still present, since a brace is no longer able to halt progression beyond this level of severity. Decompensation is another important factor in deciding whether surgery is indicated.

❗ **Evaluating the correct level of instrumentation for a walking patient with neurogenic scoliosis and selecting the right surgical technique are two of the most difficult tasks in scoliosis surgery.**

The evaluation of motor skills and the options for spontaneous compensation are critical. A surgeon conducting an assessment on the basis of the same criteria employed for an idiopathic scoliosis runs the risk of misinterpreting the situation with the consequent possibility of decompensation, since the spontaneous correction tendency is lacking or incomplete. An important principle to bear in mind is that the lowest instrumented vertebral body must be centered and, if possible, horizontal after the operation. For the aforementioned reasons, however, instrumentation down to the sacrum should be avoided [18]. If the decompensation cannot otherwise be eliminated then fixation to the pelvis is unavoidable. In some cases the patient loses the ability to walk or else walking is restricted [20], and the patient and caregivers must be informed of this risk. Particularly rigid scolioses must be approached both from the anterior and posterior sides.

➤ The basic situation is different for patients with cerebral palsy who are *unable to walk*, for whom the following *surgical objectives* apply:

- Preserve the ability to sit,
- Improve decompensation,
- Improve the nursing care situation,
- Prevent pain,
- Stabilize the spine,
- Avoid the need for a brace.

In these patients it should be possible to achieve good correction and adequate balance for sitting by straightening a long section of the spine, with instrumentation including the sacrum. In very severe, rigid curvatures, the scoliosis must be approached from both the ventral and dorsal sides, while the dorsal approach is sufficient for less pronounced curves. In the two-stage procedure, the

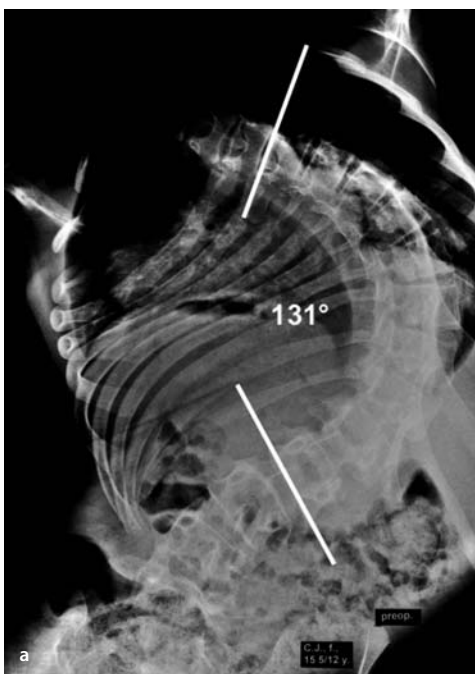
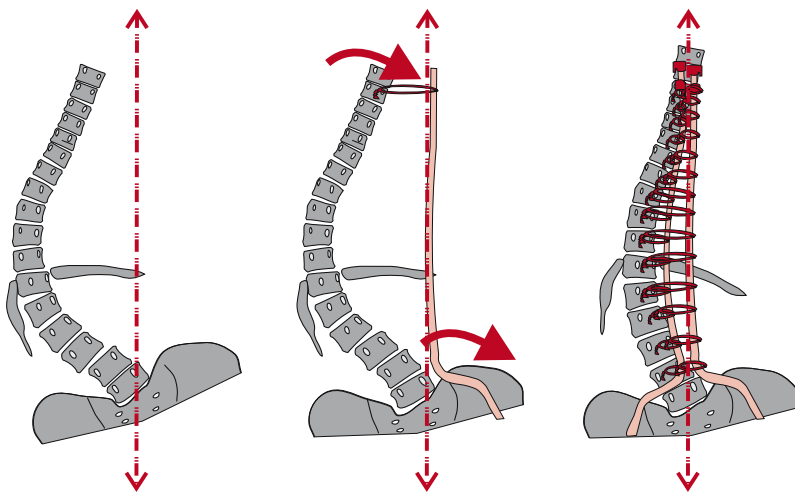
intervertebral disks are removed from the anterior side in the first operation, followed by instrumentation from the posterior side in the second operation. For patients who are unable to walk, we tend to fix the instrumentation to the pelvis and use the *Luque-Galveston technique* [10]. In this method, a rod is first anchored in the pelvis in the planned correct position, and the spine is then pulled onto the rod using segmental wires. A second rod on the convex side of the scoliosis and likewise fitted with segmental Luque wires provides added stability (■ Fig. 3.98, 3.99).

We do not treat patients with severe scolioses preoperatively with a halo (= a ring secured to the head). Halo extension for several weeks is unpleasant for the patients. Rigid scolioses can be corrected just as effectively with ventral disk removal.

A particular problem is posed by severe scoliosis in small children under 10 years of age. If substantial progression has already taken place prematurely, the operation must be carried out before adolescence. However, spondylodesis should be avoided at this young age. For patients in this age group we use a distraction rod that is fixed at the cranial and caudal ends with one hook in each case and pushed through the muscles. The rest of the spine is not exposed during this procedure. This rod has purely extending and stabilizing effects. After about a year, the patient must be reoperated and the distraction increased. An excessively long rod is always selected initially to allow for subsequent distraction, although the rod may need to be replaced. However, this procedure cannot be continued for more than three years, as the spine stiffens up, even without spondylodesis, as a result of the immobilization induced by the rod. Consequently, the definitive correction and spondylodesis must take place, at the latest, after three years, otherwise the spine will spontaneously stiffen in a less favorable position. Whether the *Campbell procedure with the titanium rib (VEPTR)* described ▶ chapter 3.1.7 is also suitable for scolioses caused by cerebral palsy remains to be seen. We have already employed the technique successfully several times in patients with flaccid paralysis (MMC).

The treatment of *neurogenic kyphoses* is another difficult task. Surgery is generally indicated if the overall kyphotic angle exceeds 80°, since no external orthosis is capable of halting the progression of the kyphosis beyond this angle. Nor can the patient raise his head sufficiently in such cases. Very pronounced kyphoses lead to an impairment of lung function and thus to a deterioration in quality of life and a reduced life expectancy. Since the forces acting on the kyphosis are usually substantial, dorsal tension-band wiring on its own is not generally sufficient for halting its progression. A combined procedure involving an initial ventral disk removal and straightening with allogeneic bone grafts inserted into the intervertebral disk spaces is usually required. In a second stage, the sur-

**Fig. 3.98.** Principle of the Luque-Galveston technique in severe scoliosis and spastic tetraparesis. On the concave side of the curve a rod is anchored in the pelvis in an orthograde position as possible. The spine is then segmentally pulled onto the rod, thereby correcting not only the scoliosis but also the pelvic obliquity. A second rod anchored in the pelvis on the convex side and fitted with segmental wires ensures adequate stability

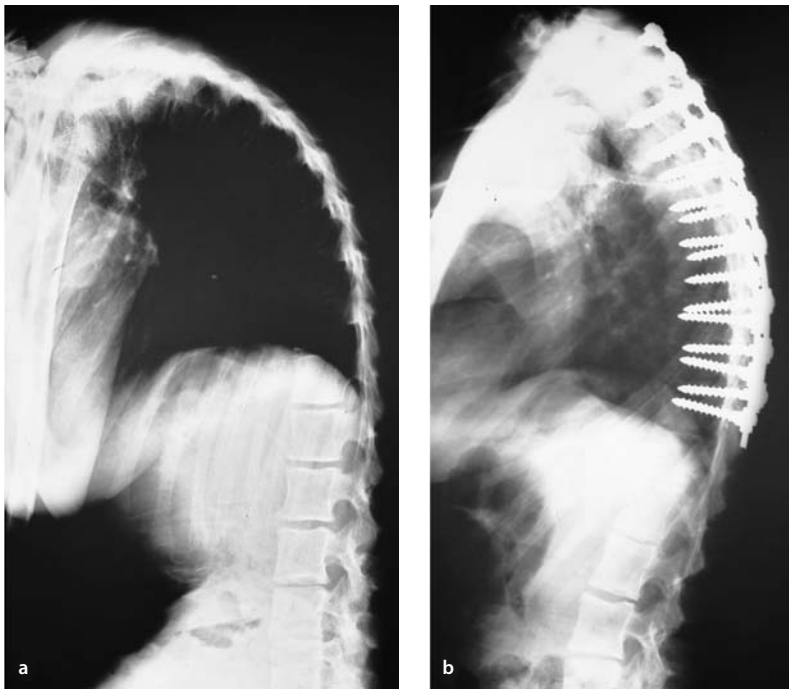


**Fig. 3.99a, b.** Luque-Galveston technique. Case history. **a** Severe neurogenic scoliosis in cerebral paresis. **b** Situation after correction with rods anchored in the pelvis and segmental wiring (after [10])

geon performs dorsal cuneiform osteotomies at several levels and tension-band wiring with a system consisting of transpedicular screws and a compression rod. In our case we use the titanium pediatric USS instrumentation, as the screws and rods are rather smaller than usual and thus more suitable, because the instrumentation generally has to be continued up to a very high thoracic level (Fig. 3.100). It is important that the kyphosis should be corrected over a prolonged section. At the cranial end, in particular, the instrumentation must continue up as close

as possible to the start of the cervical lordosis, irrespective of the location of the apex of the kyphosis. If the kyphosis is only instrumented in the low thoracic area, a new kyphosis will develop above the instrumentation as a result of kyphosing forces acting at this point.

**! Kyphoses must always be instrumented at the cranial and caudal ends right up to the turning point of the lordosis, otherwise the kyphosis may increase in the non-instrumented section.**



■ **Fig. 3.100.** 15-year old girl with severe cerebral palsy and massive kyphosis. **a** Lateral x-ray; **b** 1 year after ventral and dorsal correction and stabilization

### Results, risks and complications of surgical treatment

The greatest risk is posed by complications arising from the perioperative anesthesia. It is important to have a good anesthetic team with sufficient experience in anesthetizing these severely disabled children and adolescents. Particularly in patients who are unable to walk, the heart is poorly trained and has a diminished capacity to adapt to the perioperative stresses. Thus, for example, an intraoperative cardiac arrest can occur if substantial blood loss occurs, and we have sadly experienced a fatality in these circumstances. Given a certain risk of postoperative pneumonia, a lateral position may be appropriate as supine patients aspirate their saliva. Patients with severe epilepsy are also particularly at risk. Another problem is posed by the fact that patients with severe cerebral palsy are generally very thin and have very little muscle and subcutaneous fatty tissue over the sacrum. As a result, the rod ends at this point lie right under the skin, increasing the risk of decubitus ulcers. Overall, however, the complication rates are very low for a well-established team.

Since 1989 we in Basel have operated on 116 patients with neuromuscular spinal deformities, including 103 scolioses and 13 kyphoses. This figure only corresponds to around 3% of our patients with neuromuscular disorders. Nine patients underwent correction in stages (primarily uninstrumented) and a combined ventral and dorsal approach was employed in 12 cases. We have suffered 2 perioperative fatalities. 17 patients (14.6%) required revision surgery (twice in 2 patients). The reasons were infections,

with or without decubitus ulcer (9), hook pull-outs (particularly at the cranial end; 5) and decompensation (3). Experienced centers report similarly acceptable complication rates [8, 19].

### 3.1.10.2 Predominantly flaccid paralyses

#### ► Definition

Spinal lesions resulting in a predominantly flaccid paralysis and causing a neurogenic scoliosis (particularly poliomyelitis, traumatic paraplegia and spinal muscular atrophies).

#### Occurrence

*Poliomyelitis* used to be a very common illness, and the era of instrumented surgical scoliosis treatment started primarily with corresponding procedures for poliomyelitic scoliosis. In our own hospital, also, many scoliosis operations were performed for polio at the end of the 1960's. Today such scolioses have almost disappeared in the industrial nations, and are even very rare in the developing world. Our hospital supports a development aid project in Africa that is mainly concerned with scoliosis operations. In Africa we see more idiopathic scolioses and spinal deformities associated with systemic disorders than with poliomyelitis.

Posttraumatic scolioses associated with *paraplegia* occur typically in the growing child and rarely in full-grown patients. Fractures arising from torsion movements and

causing asymmetrical growth problems are especially problematic in the growing child. The asymmetrical paralysis of the muscles results in severe scoliosis. *Spinal muscular atrophy* is an autosomal recessive hereditary disorder of the anterior horn cells. We distinguish between a fairly severe form according to Werdnig-Hoffmann and a milder form according to Kugelberg-Welander (► Chapter 4.7.3).

### Clinical features

We can differentiate between *two curve types* that occur in flaccid paralyses:

- Scoliosis due to asymmetrical muscle activity.
- A collapsing spine due to excessive (symmetrical) flaccidity.

Most scolioses occur at the lumbar or thoracolumbar level, and purely thoracic scolioses are rare. The scolioses in poliomyelitis (as with those in predominantly spastic paralyses) also tend to show relatively little rotation with less pronounced curves with a Cobb angle of less than 40°, but are particularly badly distorted at higher curve angles [6].

### Treatment

If the scoliosis is associated with an asymmetrical paralysis, a brace treatment should be employed from a scoliosis angle of 20°. The brace can halt the progression of the scoliosis, at least to a certain extent. Surgery should be considered from a scoliosis angle of 40°. The operation consists of a posterior spondylodesis with straightening by two vertical struts and segmental wires. Since polio patients are usually able to walk, the sacrum should not be instrumented. Here, too, establishing the cranial and caudal ends of the instrumentation is not always easy. The selection of an inappropriate height may promote decompensation. A combined anterior and posterior procedure should be employed from a scoliosis angle of approx. 70°, i.e. removal of the disks ventrally and posterior instrumentation in a second operation. This can usually be achieved without difficulty in poliomyelitis. The perioperative risk associated with spinal muscular atrophy, however, is greater, and immobilization must be avoided at all costs. The patient should therefore undergo surgery before the scoliosis becomes so bad that an anterior and posterior fusion are required. If this situation cannot be avoided, not only are the disks removed during the primary anterior operation, but the spine is also instrumented with a ventral derotation spondylodesis. We then mobilize the patients and perform the second operation only after several weeks when the patient is fully rehabilitated. In the second operation with a posterior approach, the spine is instrumented with two vertical struts and segmental wiring. An alternative is the single-stage anterior procedure with a double rod (► Chapter 3.1.4).

If a *collapsing spine* is present, initial brace treatment is likewise possible. This helps defer the operation until the child has reached an age when the spine no longer has much growth potential left, if possible until the age of 10. The surgical procedure is similar to that for an asymmetrical paralysis. If the scoliosis angle is not too high, posterior straightening and segmental wiring may prove sufficient. In very severe scolioses and kyphoses, a combined anterior and posterior approach is necessary. A relatively high complication rate can be expected in patients with spinal muscular atrophy [1, 3].

### 3.1.10.3 Myelomeningocele

#### Definition

This condition involves a combination of a neurogenic scoliosis due to a predominantly flaccid paralysis with elements of a congenital scoliosis. Moreover, a pronounced (muscle-related or muscle-promoted) kyphosis can occur as a result of the anatomical anterior displacement of the dorsal muscles. The clinical picture is discussed in detail in ► chapter 4.7.2.

#### Occurrence

The prevalence of scoliosis in patients with myelomeningocele was found to be 69% in Sweden [14]. While the incidence is not age-dependent, it is connected with the level of the paralysis (at thoracic level the incidence was 94%). Kyphoses are much rarer and observed in very few patients. Still rarer are severe lordoses, which can occur particularly in an iatrogenic context following the use of a thecoperitoneal shunt.

#### Clinical features

The development of a spinal deformity in myelomeningocele is influenced by *three factors*:

- by asymmetrical *paralysis* of the muscles,
- by an *altered anatomical configuration* of the muscles,
- by *deformities* and segmentation defects of the spine.

What has been said for poliomyelitis-induced scoliosis also applies to scoliosis caused by *flaccid paralysis*. The asymmetrical muscle tension or the instability can cause a typical paralytic scoliosis. The level of the paralysis, in particular, crucially determines the extent of the scoliosis. While patients with low lumbar myelomeningoceles rarely develop a severe scoliosis, this is not generally the case for patients with a paralysis at thoracic level. It should be noted that the paralysis level can change, particularly if a *tethered cord syndrome* is present. This can lead to an exacerbation of the paresis and the problem must be tackled surgically at an early stage. A change in the paralysis level can also occur if



the patient has a poorly functioning valve as a result of the increased pressure in the ventricular system. In such cases, the paralyses are often combined with a spastic component.

The *change in the anatomy of the muscles* occurs as a result of the forward-bending of the vertebral arches that are open at the back. As a result, the muscles that are normally located on the dorsal side are shifted ventrally. These then act in addition to the ventral muscles, which are normally present, to produce a kyphosing rather than a lordosing effect. The spine thus lacks the normal dorsal tensioning. Such patients can develop very severe kyphoses. In most cases the kyphosis is already present at birth and continues to progress during growth. Closing the skin over the kyphosis during closure of the myelomeningocele is usually problematic. Since the scarred skin lies directly over the kyphotically projecting bone, a decubitus ulcer can rapidly develop in patients when they lie on their back, thereby causing additional problems.

*Congenital deformities and segmentation defects* are present in all myelomeningoceles. In fact, the open arch is already, by definition, a deformity. Segmentation defects are also almost invariably present, although these are frequently symmetrical and do not pose too great a problem, except for the disrupted growth in this area. Occasionally, however, the segmentation defect is also unilateral, and in these cases a very progressive and extremely rigid scoliosis can develop. One also occasionally sees hemivertebrae and wedge vertebrae. The progression of a scoliosis resulting from such deformities is not

especially great, although the scoliosis can also cause additional problems.

### Treatment

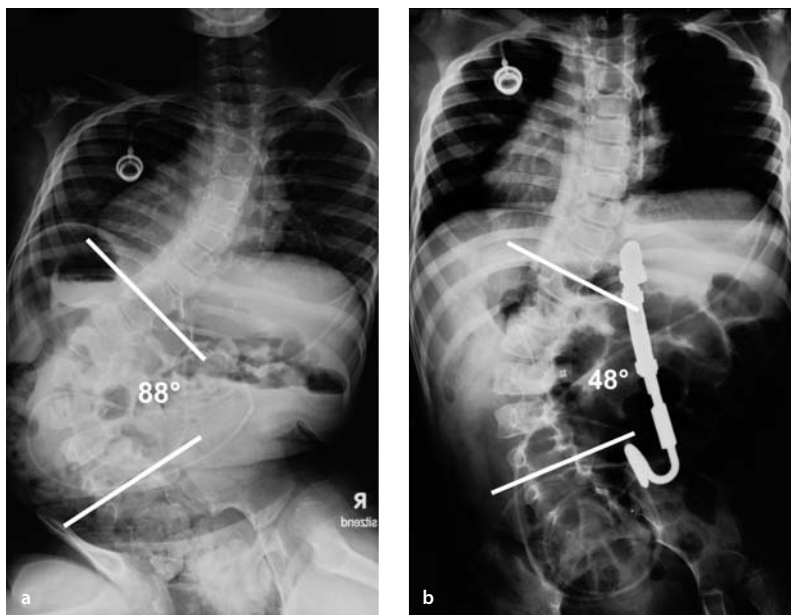
#### Conservative treatment

Scoliosis due to flaccid paralysis can – provided it is not too pronounced – be treated with braces. The brace treatment is particularly effective with Cobb angles between 20° and 40°, but is only indicated if definite progression is present. Here, too, we use the dynamic brace (■ Fig. 3.97), which gives wheelchair patients as much freedom of movement as possible. Brace treatment is ineffective in cases of kyphosis and scoliosis caused by congenital malformations.

#### Surgical treatment

If marked scoliosis is already present in childhood we currently use the *VEPTR instrumentation according to Campbell* (► Chapter 3.1.7), which is anchored to the ribs at the level of the upper end vertebra of the scoliosis and to the pelvis with a special hook (■ Fig. 3.101). This straightens the spine and preserves mobility without the need for fusion. However, the instrumentation must be extended every 6 months. Preserving mobility is particularly important in cases of myelomeningocele as such patients are only slightly restricted, if at all, as regards intelligence and remain highly active in their wheelchairs. They must be able to pick up objects from the floor, get up onto the wheelchair from the floor and transfer themselves onto a bed on their own.

If an adolescent patient presents with a scoliosis of >60° with decompensation, surgical treatment may be



■ Fig. 3.101a, b. 7-year old boy with myelomeningocele and severe scoliosis. a Postoperatively. b Situation after correction with the *VEPTR instrumentation according to Campbell*. The titanium rib is anchored to the ribs and, at the bottom, to the iliac crest on the concave side of the curve (for details of this procedure see

considered as an option. However, this decision requires much careful thought as the possible functional loss must be taken into account. In contrast with patients with normal sensory function, patients with a (high) myelomeningocele do not experience pain when the ribs come into contact with the pelvis. For these reasons we, together with patients and caregivers, tend increasingly to decide against an operation.

If surgical straightening and fusion are indicated, however, the surgeon should if possible combine an anterior and posterior approach in view of the prevailing forces and the invariable presence of osteoporosis. Fusion down to the sacrum should be avoided [21].

A special situation applies in severe lumbar kyphoses, as the scarred skin produced by the closure of the cele often results in decubitus ulcers that may subsequently require surgical restoration. In such cases correction will only be successful if the surgeon performs a kyphectomy with a wedge resection of several vertebral bodies (Fig. 3.102). In this procedure the spinal cord must be released and elevated from the kyphotic spine. Even if no neurological residual function is detected, the cord should not simply be ligated as the dural sac usually still possesses a certain drainage function and there is a risk of an increase in pressure. Serious complications can occur in connection with the often present Arnold-Chiari malformation. The kyphosis can be straightened by the wedge resection [9, 11], and the resulting configuration should ideally be fixed with screws and plates as there is hardly any space posteriorly for larger implants (rods). While a correction of the kyphosis of around 50%–60% is perfectly possible, the muscle tension cannot be changed and a subsequent correction loss cannot be ruled out. Plastic reconstruction may be required for the skin closure, e.g. with a gluteus medius flap.

Scolioses and kyphoses resulting from *congenital deformities* are treated according to the principles outlined in ▶ chapter 3.1.7. As mentioned above, the procedure involving VEPTR instrumentation is increasingly used nowadays.

### Complication risks

The surgical treatment of spinal deformities associated with myelomeningoceles involves technically difficult procedures. Since pulmonary function is not usually impaired, the perioperative anesthetic risks are not especially great. Since latex allergy is known to occur more frequently in such patients [4, 13], latex-free materials, particularly gloves, will need to be used. The skin complications associated with the scarred skin following closure of the myelomeningocele and the lack of sensory function should also always be considered. Special attention should also be paid to the possibility of tethered cord syndrome. If distraction is applied during the operation on a patient with tethered cord

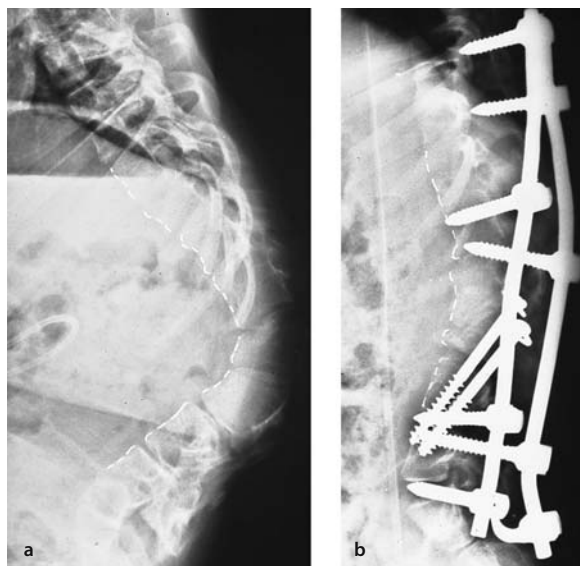


Fig. 3.102a, b. 10-year old girl with myelomeningocele and pronounced lumbar kyphosis. a Lateral x-ray of the lumbar spine; b 1 year after kyphectomy (wedge resection of vertebral bodies) and stabilization with the Spinefix instrumentation

syndrome, neurological function may be impaired [12]. Other intraspinal anomalies such as syringomyelia and Chiari malformation are also common in patients with a myelomeningocele. Pulmonary function tends to be improved by the scoliosis surgery [16].

### 3.1.10.4 Muscular dystrophies

#### Definition

Progressive muscular dystrophy refers to a group of hereditary disorders of varying severity with secondary scoliosis as a result of the muscle weakness. The various types of muscular dystrophy are described in detail in ▶ chapter 4.7.5.

#### Occurrence, etiology

Almost all patients with the more severe forms of muscular dystrophy (particularly Duchenne dystrophy, ▶ chapter 4.7.5) develop a scoliosis [2, 15].

#### Clinical features

The course of spinal problems in these clinical conditions is characterized by an initial regular development of the spine as the muscles largely possess normal strength. The ensuing muscle weakness, however, leads to hyperlordosis of the lumbar spine at an early stage. With increasing age (usually around 10 years in the commonest form, the Duchenne dystrophy), the patients *lose their capacity to*

walk and stand as a result of the loss of muscle power. This loss of power is also associated with instability in the trunk. While the spine largely retains adequate stability in the sagittal plane thanks to the ligamentous apparatus, such a corrective anatomical element is lacking in the frontal plane. If the spine deviates slightly to one side during standing or sitting, a *progressive scoliosis* can develop under the influence of gravity. In fact, the spine can collapse into a very severe scoliosis within two years. Consequently, patients with this underlying disorder should be examined regularly for spinal deformities after they have lost the ability to stand.

### Treatment

Brace treatments are not particularly effective since, on the one hand, they cannot stop the progression of the condition and, on the other, merely delay the generally inevitable operation. The surgeon is then confronted with the need for a complex operation because of a severe deformity, while the general condition of the patient has continued to deteriorate considerably.

**! In muscular dystrophies, early operations are indicated even with curves of 20°–30°. It is also important to stabilize the whole spine, from the upper thoracic level down to the pelvis, otherwise severe scolioses can occur at the end points of the fixation.**

Moreover, the patient will only be able to remain brace-free for a long time if the spine is surgically stabilized. If mobile lumbar segments remain, a support brace will continue to be needed to maintain the patient in an upright position.

In one study, 23 patients with a stabilized spine were compared with 32 patients who declined this operation. The ages and baseline curves were comparable in both groups. After five years, 75% of the operated patients were still alive, compared to just 15% of the non-operated patients [5]. This study impressively demonstrates that an early operation in these patients, who often do not live beyond the age of 20, not only improves their quality of life, but also prolongs their survival. As regards the surgical technique, the use of two vertical struts with segmental wiring has proved effective. We normally use the *Luque-Galveston procedure* in cases of muscular dystrophy (► Chapter 3.1.10.1). Some surgeons have also tried using telescopic rods and wiring without fusion, thus allowing the spine to continue growing. Early mobilization is important after the operation [1]. Patients with muscular dystrophy must start moving again within a few days postoperatively.

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### 3.1.11 Spinal deformities in systemic diseases

#### ➤ Definition

Scolioses and kyphoses associated with hereditary diseases, e.g. osteochondrodysplasias, generalized connective tissue disorders, metabolic bone disorders, chromosomal anomalies and dysostoses.

#### 3.1.11.1 Neurofibromatosis

#### ➤ Definition

Autosomal-dominant hereditary disorder characterized by café-au-lait spots and neurofibromas located almost anywhere on the body and frequently associated with scolioses. A distinction is made between four different types of scolioses, all of which are connected with the underlying condition. Radiographs show characteristic changes on the vertebral bodies and ribs. The disorder is described in detail ► chapter 4.6.8.6.

#### Occurrence

Neurofibromatosis is one of the commonest hereditary disorders, with a prevalence of 20.8 : 1,000,000 inhabitants [46]. The neurofibromatosis is associated with spinal changes in 15–20% of cases [2, 44].

#### Classification

Four types of spinal deformity can be distinguished (► Table 3.8 [6]). Types II–IV are usually termed »dystrophic« [2,6, 36].

#### Etiology

The occurrence of scoliosis in neurofibromatosis is explained by a variety of factors. While intraspinal neurofibromas are rare, paraspinal neurofibromas occur more frequently, although neurofibromas can often be completely absent in the spinal area, and not just in type I.

Congenital anomalies, particularly in the area of the cervical spine, are also often associated with the neurofibromatosis. Congenital spondylolistheses can also occur (illustration in ► chapter 3.1.7).

#### Clinical features

Scolioses in neurofibromatosis can lead to very pronounced, outwardly visible deformities. Since the scolioses are also frequently associated with strong rotation, the rib prominence is especially pronounced. The short-curved kyphosis seen in type IV can also be very disfiguring. Despite the rapid progression of the scoliosis, pain is rarely experienced. Neurological lesions are also extremely rare and can occur in connection with rib penetration into the spinal canal [20, 25] or with congenital olisthesis.

#### Radiographic findings

Except in type I, the radiological picture is highly characteristic. The dystrophic types II–IV are very short-curved. The diagnosis of neurofibromatosis can often be presumed just on the basis of the radiographic findings, even when its presence had not yet been suspected (as frequently happens).

In the dystrophic types II–IV, characteristic changes of the vertebral bodies and ribs are also observed:

- lateral wedge vertebrae,
- scalloping,
- penciling of the ribs.

► Fig. 3.103 shows the typical radiographic changes in schematic form [8]. The penciling of the ribs usually occurs on the convex side, but can also occur on the concave side. In most cases 2–5 ribs are affected. An MRI scan should always be recorded preoperatively to exclude any neurofibromas within the spinal canal.

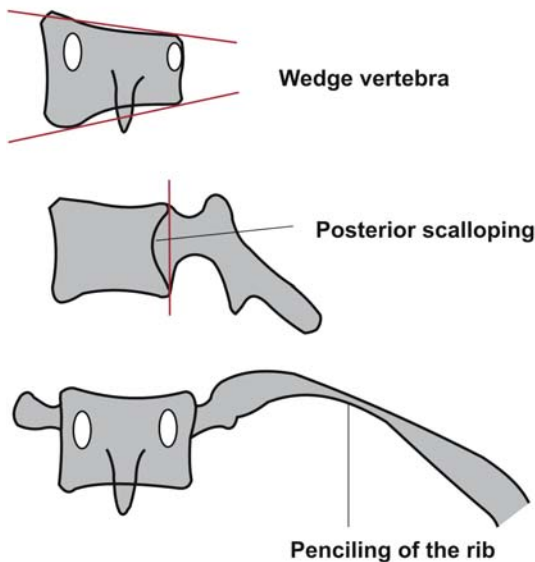
#### Treatment

Conservative treatment with a brace is not usually very effective and can only halt progression of the curve at

► Table 3.8. The 4 types of spinal deformities

Type	Features	Frequency	Severity of the scoliosis	Severity of the sagittal deformity
I	»Normal« scoliosis	+++	+	+
II	Short-curved scoliosis, thoracic lordosis	+	+	++
III	Short-curved scoliosis with harmonious kyphosis	++	+++	++
IV	Short-curved scoliosis with angular kyphosis	++	+++	+++





■ Fig. 3.103. Schematic presentation of typical changes on the x-ray in dystrophic neurofibromatosis scoliosis. (after [6])

best in type 1, but not in types II–IV. Surgical treatment is therefore required in most cases. As a rule, this must involve a combined anterior and posterior approach. On the ventral side, the intervertebral disks must be removed so that the very rigid curve can be straightened to some extent. From the dorsal side we use the («pediatric») USS instrumentation. The deformities in the sagittal plane must also be corrected as far as possible, although straightening these often very pronounced lordoses or kyphoses can prove difficult. Intraspinal neurofibromas should be ruled out preoperatively by means of an MRI scan as these must be removed before the scoliosis can be straightened.

### 3.1.11.2 Marfan syndrome

#### ► Definition

Autosomal-dominant hereditary disorder involving impaired collagen formation and generalized ligament laxity. In addition to ectopia lentis and aortic dilatation, scoliosis is the deformity most typically associated with the condition (described in detail in ► chapter 4.6.3.1).

#### Occurrence

One study found that scoliosis was present in 52 out of 82 not yet full-grown patients with Marfan syndrome (= 63%) [37]. Over half of patients with this syndrome therefore seem to develop a scoliosis, which often starts before the age of 10 [38].

#### Clinical features

Most scolioses in Marfan syndrome have their apex at the thoracic or thoracolumbar level and, like idiopathic scolioses, are almost always right-convex (50 out of 52 [37]). In contrast with idiopathic scolioses, however, they can be associated with very pronounced kyphoses, even if the lordosis is typical. The scolioses are large-curved and show a relatively strong tendency to become structural and also to be progressive. Progressive scolioses in such cases are usually relatively rigid. The frequency of spondylolysis and spondylolisthesis is above average in patients with Marfan syndrome [37].

#### Radiographic findings

Scolioses in Marfan syndrome resemble idiopathic scolioses, apart from the fact that they can also be associated with relatively pronounced kyphoses. There are no characteristic findings on x-rays of the spine. By contrast, the unusual relationship between the length and width of the metacarpals and phalanges can be observed on x-rays of the hand.

#### Treatment

Scolioses in Marfan syndrome are basically treated exactly like idiopathic scolioses. A brace treatment can be prescribed from a Cobb angle of between 20° and 40°. This often proves successful, although the tendency for the scoliosis to progress is greater, on average, in patients with Marfan syndrome than those with idiopathic scoliosis [39]. If there is a Cobb angle of >25° at Risser stage I, the chances of avoiding an operation are slim [39]. Surgical treatment must be considered from a Cobb angle of 40°. The operation is similar to that for idiopathic scoliosis, although secondary curves – unless they are also stiffened at this time – show a greater tendency to progress than in idiopathic scolioses [21]. Since cardiac and aortic malformations also commonly occur in Marfan syndrome, the cardiac situation should be carefully investigated before surgery. The surgical risk is accordingly somewhat higher than in idiopathic scoliosis.

### 3.1.11.3 Osteogenesis imperfecta

Osteogenesis imperfecta refers to a group of relatively common autosomal-dominant or -recessive disorders involving abnormal bone fragility, blue sclerae and hearing loss. The disorder is described in detail in ► chapter 4.6.3.2. Characteristic biconcave vertebral bodies can occur in the spine and can be associated both with scolioses and also with kyphoses [12]. Spinal deformities occur in 40–80% of patients with osteogenesis imperfecta [7, 15, 45].

The number of altered biconcave vertebral bodies is a prognostic criterion for the progression of the scoliosis [14]. As the scolioses are often very progressive, brace



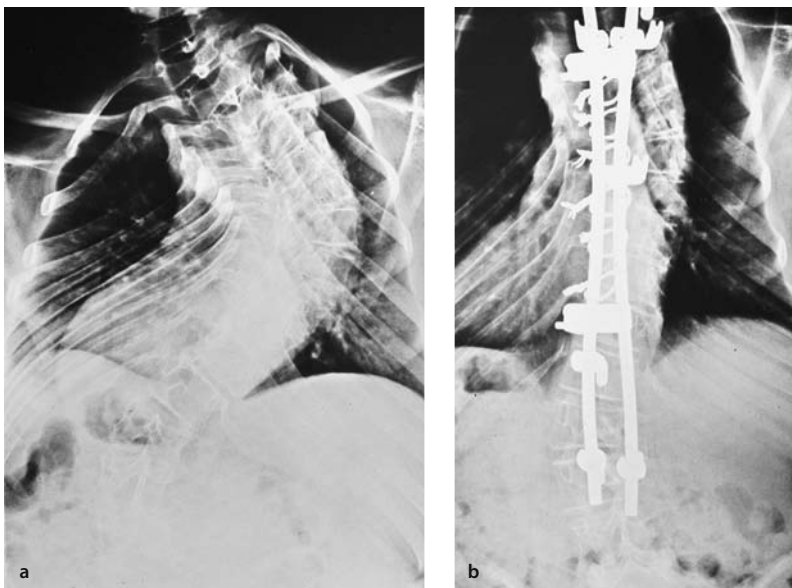
treatment usually proves ineffective. Since the curves are rigid and the stability of the vertebral bodies is not very good, a combined anterior and posterior approach should be selected at operation. As instrumentation we prefer a segmental procedure, as the correcting forces must be distributed in the most effective manner (■ Fig. 3.104). In spite of stable instrumentation the deformity can still be progressive [43]. If the pelvis is strongly tilted in a severe case of scoliosis, metal instrumentation should not be

used as it could penetrate the pelvis. In such cases we have used an extensive (tibial) allograft as a rod and secured this to the spine segmentally with nonabsorbable sutures (■ Fig. 3.105).

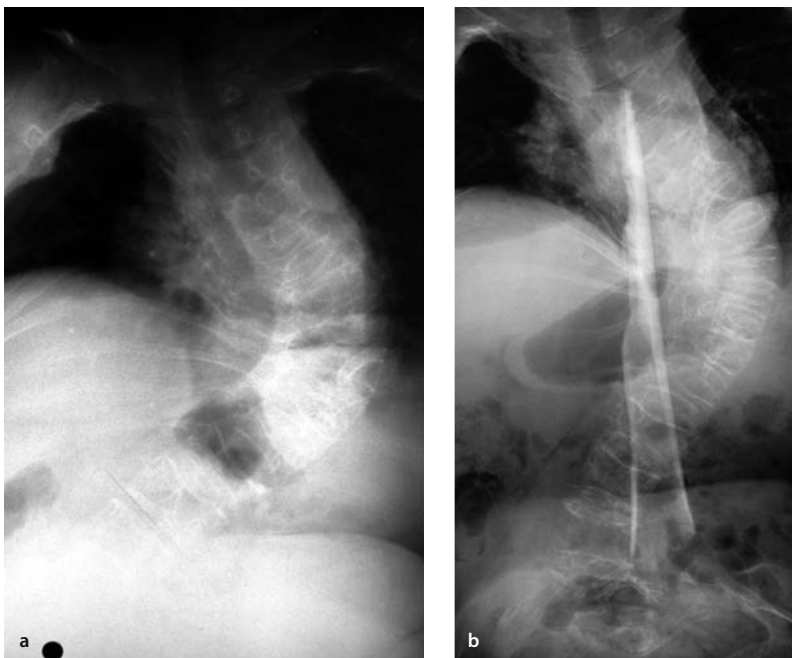
#### 3.1.11.4 Ehlers-Danlos syndrome

Ehlers-Danlos syndrome is a group of disorders involving impaired collagen formation and characterized by

■ Fig. 3.104a, b. 18-year old patient with *osteogenesis imperfecta* and severe thoracic scoliosis. a Postoperative AP x-ray. b 1 year after correction with Spinefix rod and segmental wiring



■ Fig. 3.105a, b. 14-year old girl with very severe *osteogenesis imperfecta* and decompensation of the spine. a AP x-rays of the spine. The pelvis was so thin that anchorage of a metal rod proved impossible. b The spine was straightened and stabilized with a substantial allograft from a tibia



### 3.1.11 · Spinal deformities in systemic diseases

excessive generalized ligament laxity, skin changes and fragility of blood vessels (described in detail in ► chapter 4.6.3.3). Although scolioses are not particularly common in patients with Ehlers-Danlos syndrome, when they do occur they can be very severe and also associated with pronounced kyphoses [22]. Such scolioses develop at a very young age. Brace treatment is prescribed with the aim of keeping the situation in check until the age of 10, when the indication for surgery can be assessed. The operation is performed in the conventional manner. Possibly there is an increased risk for vascular complications [3].

#### 3.1.11.5 Apert syndrome

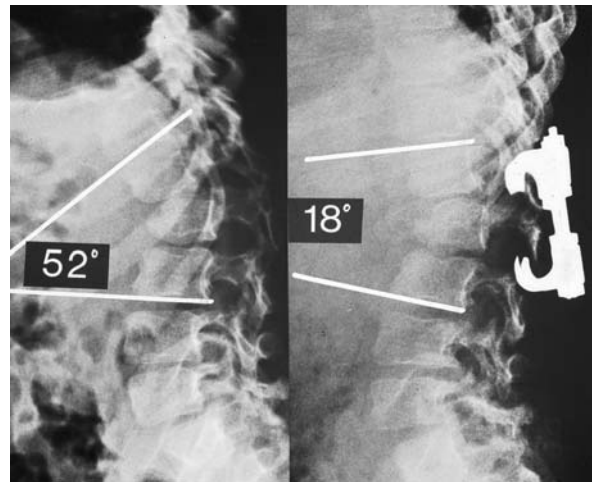
Apert syndrome is a congenital disease involving synostoses of the sutures of the skull, syndactyly and synostoses on the hands and feet (see detailed description in ► chapter 4.6.6.1). Highly typical findings in Apert syndrome include segmentation defects of the cervical spine, which occur in approximately 70% of cases. Half of these occur in isolation and half are multiple. The segmentation defects usually occur at the level of C5/C6 [42], but can also occur at C2/C3 level. Such cervical anomalies can restrict the mobility of the cervical spine, but rarely require therapeutic, or particularly surgical, intervention. An awareness of such anomalies is important, however, in respect of the induction of anesthesia for operations on the hands and feet.

#### 3.1.11.6 Fibrodysplasia ossificans progressiva

This is a rare autosomal-dominant hereditary disorder involving progressive calcification and ossification of the fasciae, aponeuroses, tendons and ligaments. This spreads in a cranial to caudal direction and from the center to the periphery (► Chapter 4.6.3.4). In a multicenter study in the UK, cervical anomalies were found on the x-rays of 5 out of 34 patients with this condition. The vertebral bodies were strikingly small, while the pedicles tended to be larger than normal. However, these anomalies did not have any clinical consequences [35].

#### 3.1.11.7 Mucopolysaccharidoses

Mucopolysaccharidoses are a group of disorders in which mucopolysaccharide metabolism is impaired, resulting in the storage of mucopolysaccharide components (► Chapter 4.6.2.4). Six types are distinguished depending on the enzyme defect in each case. Spinal changes are primarily found in type I (Pfaundler-Hurler) and type IV (Morquio syndrome). A very characteristic finding is platyspondylia, i.e. flattening of the vertebral bodies in which the central section projects forward like a tongue (► Fig. 3.106). This can lead to kyphosis



■ Fig. 3.106. Lateral x-ray of the thoracolumbar junction in a 9-year old boy with *mucopolysaccharidosis* (Morquio syndrome) and platyspondylia with subluxation of T12 over L1 (left). Right 1 year after dorsal tension-band wiring with compression rod

and vertebral slippage, particularly at the thoracolumbar transition, and the possible need for dorsal tension-band wiring. An increased incidence of atlantoaxial instabilities with a hypoplastic or even completely absent dens is also observed, particularly in Morquio syndrome [30, 40]. In one study cervical CT myelograms were recorded for 13 patients with Morquio syndrome. In all cases, the dens was hypoplastic, although most patients only showed slight atlantoaxial instability. In some of the patients the extradural soft tissues were thickened, which could have caused compression of the spinal cord. In these cases the threat to the cord was removed by occipitocervical fusion [30, 40]. Whereas the need for surgery to the cervical spine is relatively rare in our experience, dorsal tension-band wiring is often indicated for progressive thoracolumbar kyphosis in mucopolysaccharidoses.

#### 3.1.11.8 Achondroplasia

Achondroplasia is an autosomal-dominant hereditary disorder involving dwarfism and impaired enchondral ossification (► Chapter 4.6.4.1). Spinal examination reveals shortening of the pedicles with vertebral bodies of normal height, although the ossification centers are smaller than usual. Children show a long drawn-out kyphosis extending down to the lumbar spine, beneath which is an acute-angled lordosis. This kyphosis of the upper lumbar spine can be very problematic [1]. The shortening of the pedicular distances is the cause of subsequently developing spinal stenosis, which is highly

typical of achondroplasia and occurs sooner or later in most patients [28, 41]. Occipitalization of the atlas and narrowing of the foramen magnum have also been observed in cases of achondroplasia and can lead to myelopathy.

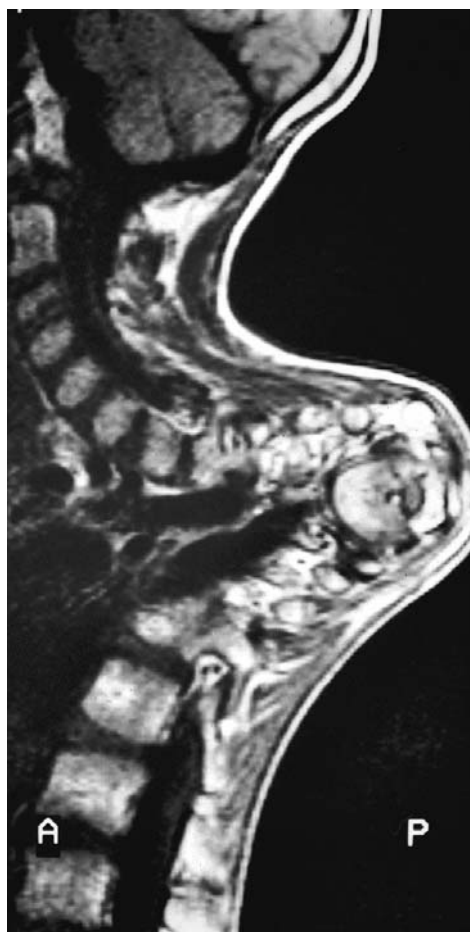
### 3.1.11.9 Diastrophic dwarfism

This rare autosomal-recessive hereditary disorder involves highly disproportionate dwarfism (► Chapter 4.6.4.4). The main spinal problem is the formation of an extremely pronounced kyphosis, which can become established at the cervical or upper thoracic level (■ Fig. 3.107). Roughly one third to one half of patients with diastrophic dwarfism show this kind of kyphosis [29, 33]. Some patients also develop neurological signs and symptoms at a very early stage as a result of incomplete arch closure at this level (■ Fig. 3.108). The effect is similar to that after a laminectomy in small children: The absence of dorsal tensioning leads to the formation of a severe kyphosis. It is very important to perform a dorsal fusion as soon as possible after the kyphosing is detected. This is performed without a metal implant but with external fixation with a halo or in a Minerva cast for 3 months. If this early treatment is neglected, later treatment will become very problematic because, once a severe kyphosis has become established, the dorsal spondylodesis will no longer be capable of straightening the kyphosis again. In such cases, anterior and posterior correction will be required, possibly even during childhood.

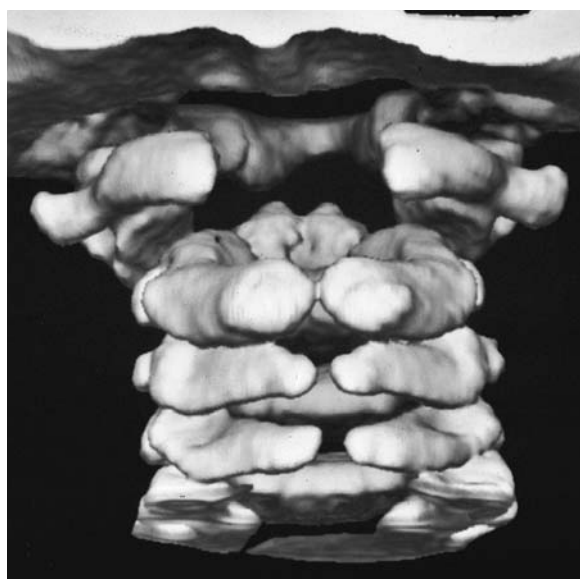
### 3.1.11.10 Spondyloepiphyseal dysplasia

This is a hereditary condition involving disproportionate dwarfism and occurring in both a more severe autosomal-dominant form and a milder X-chromosomal recessive form (► Chapter 4.6.4.9). The spinal changes vary somewhat, depending on the type of the illness. A typical finding is platyspondylia with subchondral irregularities and biconvex vertebral bodies (■ Fig. 3.109). As with mucopolysaccharidoses, this can lead to a thoracolumbar kyphosis with slight vertebral slippage [24]. These kyphoses may require posterior tension-band wiring.

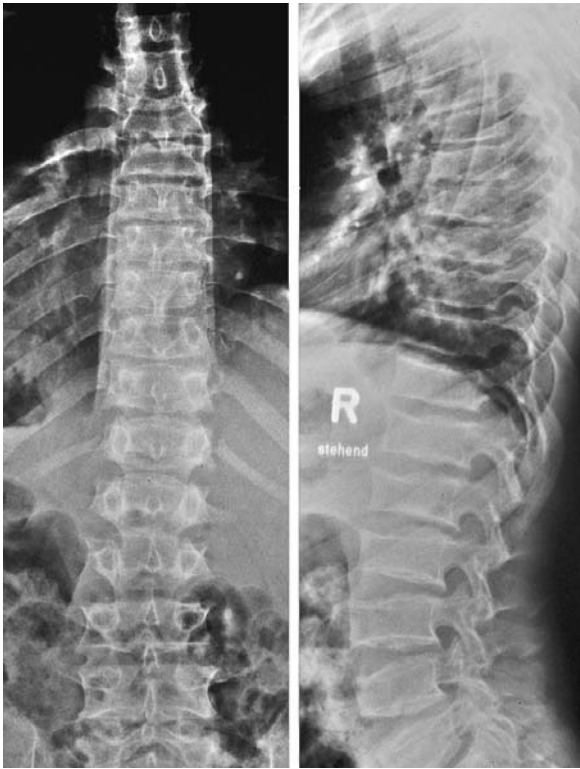
Scolioses occur, albeit unusually so, in association with spondyloepiphyseal dysplasia. Another not infrequent finding is hypoplasia of the dens, which occurs predominantly in the congenital type of the disease. Since this can result in atlantoaxial instability, an occipitocervical fusion often has to be performed at a very early stage. A new, autosomal-dominant form of the condition with atlantoaxial instability was recently described [31]. On the whole, the spinal problems in spondyloepiphyseal dysplasia are similar to those in mucopolysaccharidosis.



■ Fig. 3.107. MRI of a 14-year old male patient with *diastrophic dwarfism* and very severe thoracic kyphosis



■ Fig. 3.108. Three-dimensional reconstruction of an upper cervical spine in a 5-year old boy with *diastrophic dwarfism*. Note the lack of arch closure, particularly of the atlas



■ Fig. 3.109. AP and lateral x-rays of the spine in a 17-year old male patient with *spondyloepiphyseal dysplasia*. Typical findings are the long-drawn out, flat and biconvex vertebral bodies with subchondral irregularities



■ Fig. 3.110. Lateral view of the cervical spine of a 1-year old child with *Larsen syndrome*. Note the kyphosing of the cervical spine, which occurs very frequently in Larsen syndrome

### 3.1.11.11 Larsen syndrome

Segmentation defects in the cervical spine are a particular feature of this autosomal-dominant or autosomal-recessive hereditary condition with multiple congenital dislocations (► Chapter 4.6.4.16) [14, 19]. The segmentation, and occasionally also formation, defects can also lead to the development of a cervical kyphosis. Torticollis is also invariably present (■ Fig. 3.110). Early posterior fusion is indicated particularly if a cervical kyphosis has formed (in some cases even during the first year of life) [15]. If lordosing subsequently occurs, an additional anterior stiffening procedure may be needed. Atlantoaxial subluxation also occurs occasionally and must be treated by occipitocervical fusion.

### 3.1.11.12 Kniest syndrome

Kniest syndrome involves a defect of enchondral ossification (► Chapter 4.6.4.7). The patients show disproportionate dwarfism, and the spinal changes are similar to those seen in spondyloepiphyseal dysplasia. Problems can occur particularly with atlanto-occipital instability [23], in which case an occipitocervical fusion may be indicated.

### 3.1.11.13 Osteopetrosis

Osteopetrosis is a metabolic bone disorder characterized by a systemic increase in skeletal mass (► Chapter 4.6.4.12). Thickening of the vertebral body endplates is observed in osteopetrosis [4], but this does not have any clinical implications.

### 3.1.11.14 Chromosomal anomalies

Chromosomal anomalies are described in detail in ► chapter 4.6.5. The commonest anomaly is Down syndrome (trisomy 21). The principal problem in relation to the spine is atlantoaxial instability, which is observed in approx. 9% of cases [27]. Children with atlantoaxial instability frequently show abnormalities of the cervical spine.

! **Atlantoaxial instability is a serious problem that must be borne in mind in patients with Down syndrome. Before a general anesthetic is administered the existence and extent of any such instability must be clarified [13].**

If significant instability is present, an occipitocervical fusion or atlantoaxial screw fixation is indicated. Two-thirds of patients with atlantoaxial instability have



positive neurological findings [27]. Scolioses are also observed in patients with Down syndrome, though not very frequently.

### 3.1.11.15 Klippel-Trenaunay-Weber syndrome

This rare congenital anomaly is characterized by large, hemangiomatic nevi and unilateral hypertrophy of soft tissues and bone (► Chapter 4.6.8.7). A study investigating 28 patients with this syndrome found eight with scoliosis, two with kyphosis and two with hemivertebrae [10]. The scolioses are connected with the, often very pronounced, leg length discrepancy. The treatment criteria match those of idiopathic scolioses.

### 3.1.11.16 Fibrous dysplasia

This condition is characterized by hamartoma-like fibrous changes that can occur in polyostotic (► Chapter 4.6.8.5) or monostotic (► Chapter 4.5.2.3) forms. In the polyostotic form, the spine is occasionally affected, potentially resulting in severe scolioses and/or kyphoses [11]. Scolioses are treated according to the usual guidelines. The kyphosis requires combined ventral and dorsal correction, and the insertion of a stable fibular graft is required ventrally because of the extensive bone defect (■ Fig. 3.111).

### 3.1.11.17 Prader-Willi syndrome

This hereditary condition is characterized by hypotonia, mental retardation and obesity. In one study involving

12 patients, a scoliosis was observed in three cases and a kyphosis in one patient [32]. Treatment is based on the same guidelines applicable to idiopathic scoliosis and kyphosis.

### 3.1.11.18 Williams syndrome

Very pronounced kyphoscolioses can occur in this very rare hereditary disorder with characteristic facial changes, mental retardation and impaired growth [26]. Combined ventral and dorsal correction is occasionally required. Brace treatment is less effective.

### 3.1.11.19 Goldenhar syndrome

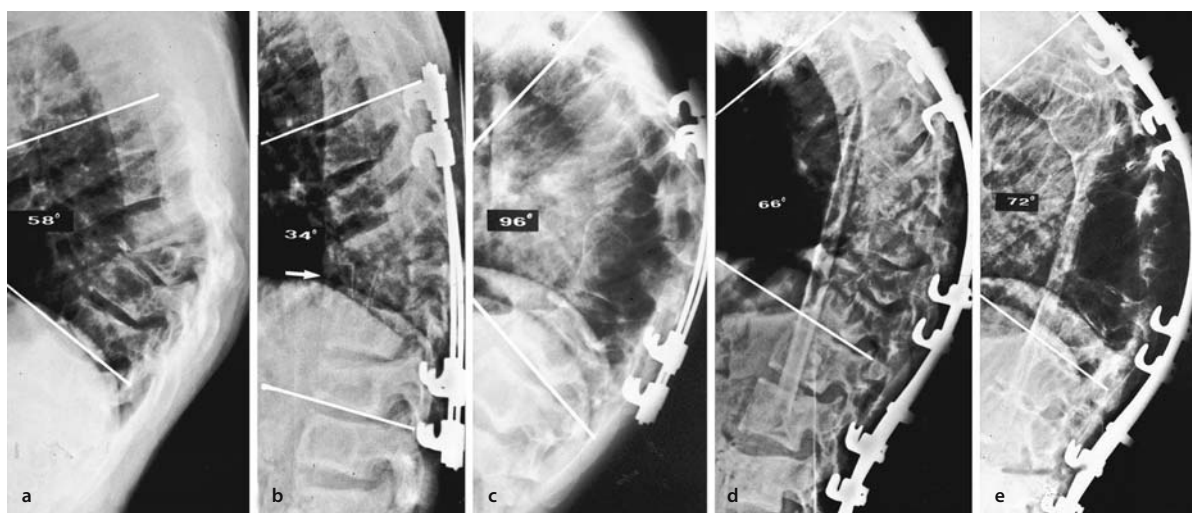
This syndrome is characterized by multiple anomalies. Vertebral formation or segmentation defects are also found in two-thirds of cases [9].

### 3.1.11.20 Prune-belly syndrome

In this syndrome the abdominal wall muscles are absent, producing the wrinkled, prune-like belly implicit in the name. The lack of any force to counter the spine can promote the formation of a kyphosis [18]. Children with this syndrome should be fitted with a compressing lumbar corset to avoid this problem.

### Spinal deformities in systemic diseases

■ Table 3.9 provides an overview of spinal deformities in systemic diseases.



■ Fig. 3.111. Lateral x-rays of the spine of a male patient with *polyostotic fibrous dysplasia*. **a** By the age of 14 wedge vertebrae had formed as a result of fibrous dysplasia at the level of T8. **b** Situation after ventral correction and dorsal tension-band wiring. **c** 3 years later: renewed severe kyphosing due to spread of the disease. **d** Status after ven-

tral correction with fibular graft and dorsal stabilization with Cotrel-Dubousset instrumentation. **e** 6 years later further slight kyphosing had occurred as a result of progression of the disease, although the situation has remained reasonably stable



■ Table 3.9. Spinal deformities associated with systemic diseases

Disease	Typical spinal deformity	Frequency within the syndrome	Severity	Treatment
Neurofibromatosis	Type I: »normal« scoliosis	+++	+	»Anterior release« and posterior correction  Possibly occipitocervical stabilization
	Type II: lordoscoliosis	+	++	
	Type III: kyphoscoliosis	++	++	
	Type IV: kyphoscoliosis with gibbus	++	+++	
	XR: Wedge vertebrae, depressions, pencil-thin ribs, also cervical deformities			
Marfan syndrome	Thoracic scoliosis, occasionally with kyphosis	+++	++	Posterior correction, poss. additional »anterior release«
Osteogenesis imperfecta	Biconcave vertebral body, thoracic and lumbar scolioses, poss. with kyphoses	+++	+++	Posterior correction, poss. additional »anterior release«
Ehlers-Danlos syndrome	Thoracic and lumbar scolioses, poss. with kyphoses	+	++	Posterior correction, poss. additional »anterior release«
Apert syndrome	Cervical segmentation defects	+++	+	Generally no treatment required
Fibrodysplasia ossificans progressiva	Cervical anomalies (small vertebral bodies, large pedicles)	+	+	No treatment required
Mucopolysaccharidoses	Platyspondylia, thoracolumbar kyphosis	+++	++	Posterior tension-band wiring Possibly occipitocervical fusion
	Atlantoaxial instability with missing dens	+++	++	
Achondroplasia	Lumbar kyphosis, spinal stenosis	+++	+++	Laminectomy –
	Occipitalization of the atlas	+	+	
Diastrophic dwarfism	Cervical kyphosis	++	+++	Posterior, poss. also Anterior fusion, poss. correction osteotomy, poss. Anterior and Posterior correction and stabilization
	Scoliosis	+++	+++	
Spondyloepiphyseal dysplasia	Platyspondylia, thoracolumbar kyphosis	+++	++	Posterior tension-band wiring Possibly occipitocervical fusion
	Atlantoaxial instability	++	++	
Larsen syndrome	Segmentation defects in the cervical spine	+++	+	Possibly Posterior and anterior fusion Possibly occipitocervical fusion Possibly early posterior spinal fusion
	Atlantoaxial instability	+	++	
	Cervical kyphosis	+	+++	
Kniest syndrome	Atlantoaxial instability	+	++	Possibly occipitocervical fusion
Osteopetrosis	Thickening of the vertebral body endplates	++	–	–
Trisomy 21 (Down syndrome)	Atlantoaxial instability	+++	++	Possibly occipitocervical fusion Possibly brace or scoliosis operation
	Scoliosis	+	++	
Klippel-Trenaunay-Weber syndrome Fibrous dysplasia	Scoliosis, kyphosis, hemivertebra	++	+	Possibly brace or scoliosis operation Anterior and posterior correction and stabilization
	Scoliosis, kyphosis	+	+++	
Prader-Willi syndrome	Scoliosis, kyphosis	++	++	Treatment as for idiopathic forms
Williams syndrome	Severe kyphosis	++	+++	Anterior and posterior correction and stabilization
Goldenhar syndrome	Formation and segmentation defects	+++	++	Possibly hemivertebrectomy, spondylodesis

Frequencies: + rare, ++ occasional, +++ common. Severity: + mild, ++ moderate, +++ severe.

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### 3.1.12 Spinal injuries

#### Definition

Fractures of the vertebral bodies and/or vertebral arches, ligament injuries and/or dislocations of the axial skeleton with or without neurological lesions.

#### Occurrence

A Canadian study investigated 3,200 patients, including 174 who were still in the growing phase (5.4%). Almost half (45%) of these had a neurological lesion [12].

**While children are less likely to suffer a spinal injury than adults, when a child does sustain such an injury, the risk of an associated neurological lesion is much higher than for adults.**

The incidence of spinal cord injury is around 30–40/1,000,000 inhabitants [9, 22]. Another study on cervical spine injuries found that these occurred less frequently in children under 11 than in adults, but were associated with a high mortality. The incidence of cervical spine injuries in over-11-year olds matches that in adults and was cited as 74/1,000,000 of the population/year [16].

#### Etiology

Traffic accidents and falls from a great height are the predominant causes of injury in children under 10 years [7, 16, 21]. In adolescents, on the other hand, sporting accidents are the commonest cause. In our own investigation [14], the sporting activity that resulted in (severe) spinal injuries was skiing in 33% of cases, swimming in 13%, horse riding and gymnastics both in 12% of cases, mountaineering in 8%, paragliding in 4% and diving in 3% of cases. The remaining 16% occurred during a variety of other sports. An increased frequency of accidents has also been reported for trampolining [4]. The risk of spinal injuries during skiing is higher in adolescence than either before or after this period [23]. By contrast, the currently popular youth-oriented sport of snowboarding does not appear to involve an increased risk of spinal injuries (in contrast with injuries to the upper extremities) as the speeds achieved with a snowboard tend to be lower than with skis.

#### Localization

The principal sites of injury in adults are the lower cervical spine and the thoracolumbar junction (T11–L3). In general, lesions of the lumbar spine are more common than cervical injuries. With the exception of vertebral bodies T11 and T12, fractures of the thoracic section are extremely rare. By contrast, in our own study [20] with 51 children and adolescents with 113 fractures we found that the thoracic spine was actually the most frequently affected site of injury (Fig. 3.112). This, of course, is due

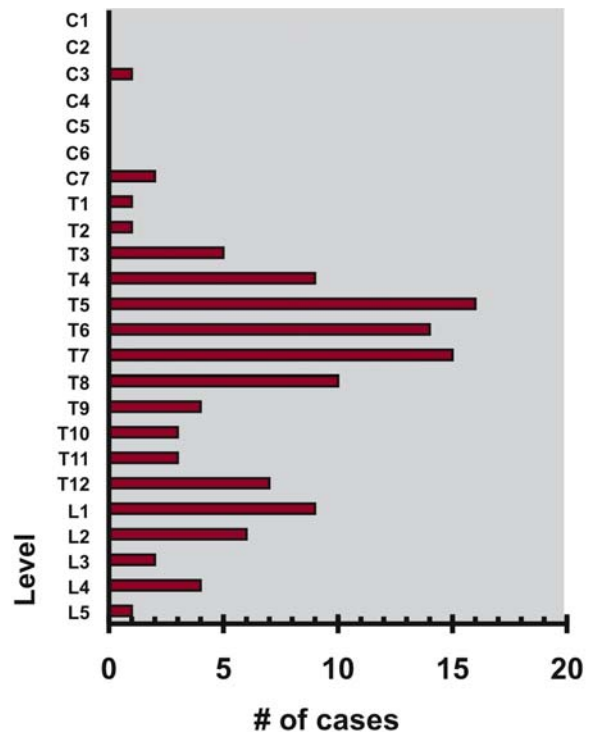


Fig. 3.112. Distribution of fracture levels in children and adolescents (after [20]). Fractures at the thoracic level are commonest in this age group, but rarely occur at this site in adults

to the fact that the thorax is much more elastic in children and adolescents than in adults. A second frequency peak for the pediatric age group was observed for the thoracolumbar junction, where most of the adult fractures also occur.

#### Classification

A special feature of pediatric spinal trauma is traumatic paraplegia without any detectable changes on the x-ray (known as SCIWORA syndrome, which stands for spinal cord injury without radiographic abnormality [3]). Such injuries are not included in the usual classifications since they do not produce any radiographically visible lesion. Nor does the MRI scan show any clear findings, even though the diagnosis is made much easier with this type of imaging. The injuries with radiographically visible fractures can be classified as for adult fractures.

To this end we use the AO classification, in which the fractures are subdivided according to the mechanism of injury [15]:

- A: Compression
- B: Distraction
- C: Torsion

**Table 3.10. AO classification of spinal trauma**

Type	Features
A: Compression	
A 1:	Impaction
A 2:	Split fracture
A 3:	Burst fracture
B: Distraction	
B 1:	Distraction with transosseous injury
B 2:	Distraction with intra-articular injury
B 3:	Distraction and extension
C: Rotational	
C 1:	Rotational + type A
C 2:	Rotational + type B
C 3:	Special cases (e.g. »slice-fracture«)

Each group has 3 subgroups (Table 3.10)

Torsion injuries are identified e.g. by a displacement of the spine or by a break in transverse processes, ribs, etc.

In a group of over 1,400 fractures, type A dominated with 74% of cases, followed by types B and C in 10% and 16% of cases respectively. Over half of the type A injuries were pure compression fractures (A 1).

### Clinical features, diagnosis

If a spinal injury is suspected, AP and lateral radiographs must be recorded. In addition, meticulous neurological examination is required [1]. Interpreting the x-rays, particularly those of the cervical spine, is not always easy. On the one hand, a distinction needs to be made between incomplete ossification, particularly in the upper cervical spine, and fractures or even pseudarthroses. Thus, synchondroses are found in children between the dens and corpus or on the atlas arch and are of no clinical significance. Os odontoideum is common and can be mistaken for a dens fracture [17]. On the other hand, the relatively substantial mobility of the upper cervical spine also needs to be interpreted correctly. The anterior subluxation of the 2nd vertebral body over the 3rd is normal up to the age of 8, and the gap between the dens and atlas arch can be over 3 mm in small children.

However, genuine tears of the transverse ligament with atlantoaxial subluxation also occur [10]. The interpretation of cervical x-rays is hampered by the fact that the pain usually causes the patient to assume a position of torticollis with rotation of the whole cervical spine. Provided no neurological lesion is present, temporary im-

mobilization with a cervical collar and re-evaluation after one week can prove helpful in uncertain cases. In particular, the presence or absence of any instability can then be established with a (careful) functional x-ray in inclination and reclination.

Radiographs of the thoracic and lumbar spine are easier to evaluate than those of the cervical spine. Compression fractures can be differentiated from wedge vertebrae in Scheuermann disease since the endplate of compressed vertebral bodies tends to overlap the anterior edge slightly. Moreover, the intervertebral disk space is normal in contrast with the situation in Scheuermann's disease. One should not overlook injuries of the vertebral arches and pedicles (type B and type C fractures). On an AP x-ray, which must also be recorded in every case, we look for asymmetry of the endplates, i.e. a collapse on one side, and examine the alignment of the vertebral bodies and any lateral dislocation. The latter is evidence of a (usually severe) torsion injury. Other indications of such a lesion also include fractures of the ribs or transverse processes and dislocations generally.

Myelography or a CT scan (a CT-myelogram) can provide further information in uncertain cases. Under no circumstances, however, should such an investigation be allowed to delay the reduction of a dislocation with a neurological lesion. Myelography can show the level of any impairment of the spinal canal. Fragments in the spinal canal are best viewed by CT. The MRI scan has little place in acute diagnosis and is primarily suited to the imaging of soft tissue injuries in those patients with neurological changes but no visible radiographic lesion.

### Prognosis

**! Although spinal injuries are rarer in children than in adults, they are more commonly associated with neurological lesions. The chances of recovery are greater in children than in adults [12].**

Of 174 children with spinal injuries 45% had a neurological lesion. 74% of these subsequently showed a marked improvement in the neurological findings, while 59% even experienced a complete recovery [12]. Another more recent study confirms the high rate of neurological improvement following severe traumatic pediatric spinal cord injury [24].

Children with permanent neurological lesions are at great risk of scoliosis formation. In 55 prepubertal children and 75 adolescents significant scoliosis occurred in 97% and 52% of cases respectively [5]. Children with neurological lesions should therefore be supported with a brace even before any scoliosis has become pronounced [1, 5].

Deformities can also occur without neurological lesions. This is particularly the case when the growth zone of an endplate is affected. Establishing whether such an

injury has occurred or not is not always easy. In pure compression fractures (type A), the growth usually remains unaffected. Injuries of the apophyseal ring tend to occur in connection with a torsion element. In such cases, an asymmetrical wedge shape is usually observed on the AP view [20]. Whether pure wedge vertebrae can be straightened out also depends on the resulting pressure. Provided the pressure is not too great, the wedge vertebra will straighten out, depending on the growth potential in each case [18, 20].

**! Expressed simply, vertebral bodies with a wedge vertebra of less than 10° will straighten out spontaneously (■ Fig. 3.113), while vertebral bodies with a wedge angle of 10° or more can only be corrected with external support (brace or cast treatment, possibly surgery) [18].**

Naturally, this straightening process also depends on the available growth potential, and a spontaneous correction can only be expected if the Risser sign has not progressed beyond stage II [18]. If the apophyseal plate is injured, increasing deformity rather than correction will result.

#### Treatment

The following options are available:

- mobilization and functional treatment
- cast treatment
- brace treatment
- surgical treatment

#### Conservative treatment

Over a third of spinal injuries involve simple compression fractures. No specific treatment is required for a single compression fracture with a wedge angle of less than 10°. After a few days' bedrest and painkillers, early

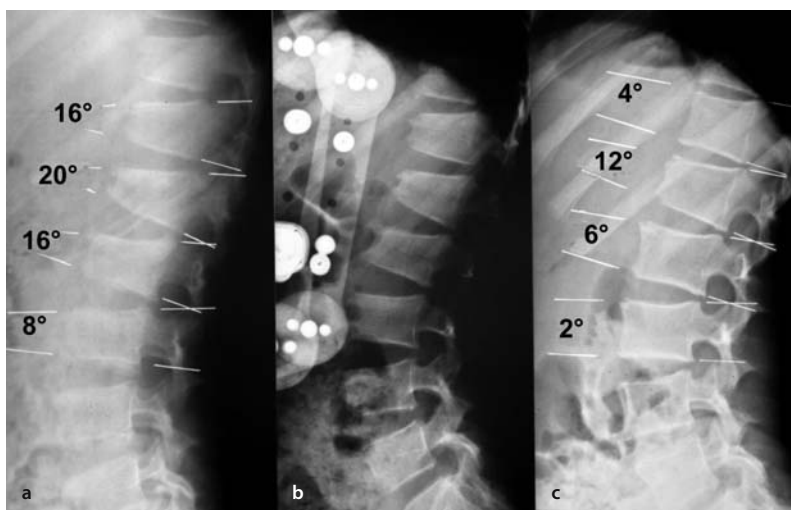
mobilization can begin without a brace [18]. This particularly applies to fractures of the thoracic spine. Since a kyphosis is much less well tolerated at lumbar level than at thoracic level, a plaster cast or brace treatment should be prescribed in doubtful cases for lumbar injuries.

For fractures with a wedge angle greater than 10°, cast treatment followed by a brace should be prescribed [13, 20]. We initially lay patients down with a padded roll underneath the fractured vertebral body. When the pain has subsided we fit a plaster brace while the patient is in ventral suspension (■ Fig. 3.114). We follow a similar procedure if several vertebral bodies with a wedge angle of more than 6° are present. The patient is mobilized with the plaster (or plastic) brace. After 6 weeks the cast is changed and a removable brace is fitted after 3 months, which is then worn for a year.

Fractures of the cervical spine are treated with a cervical collar. A Minerva cast is fitted in the event of significant instability or a dens fracture.



■ Fig. 3.114. Preparation of a cast brace in ventral suspension: The patient lies on his front with shoulders and legs on separate tables and is held by the hands and feet. The spine is freely suspended between the two tables



■ Fig. 3.113a–c. a Lateral x-ray of the spine in a 12-year old boy after compression fractures at several levels. In contrast with a wedge vertebra in Scheuermann disease, the vertebra after a fracture does not show intervertebral disk narrowing. b View with brace. c 2 years later: The multiple wedge vertebrae have largely straightened themselves out



## Surgical treatment

Surgical treatment is indicated in:

- unstable fractures,
- neurological lesions,
- secondary deformities.

At the level of the *cervical spine*, atlantoaxial instabilities and dens fractures are the main indications for surgical treatment. Dens fractures occurring in adolescence, as in adults, can be managed with screw fixation [2]. Atlantoaxial instability can be managed by screw fixation according to Magerl's method [11]. Occasionally an occipitocervical arthrodesis proves necessary. Postoperatively a halo fixed to a cast brace must be fitted. Fixation with the halo is generally well tolerated, although minor complications can occur (particularly infections at the nail insertion points [8]).

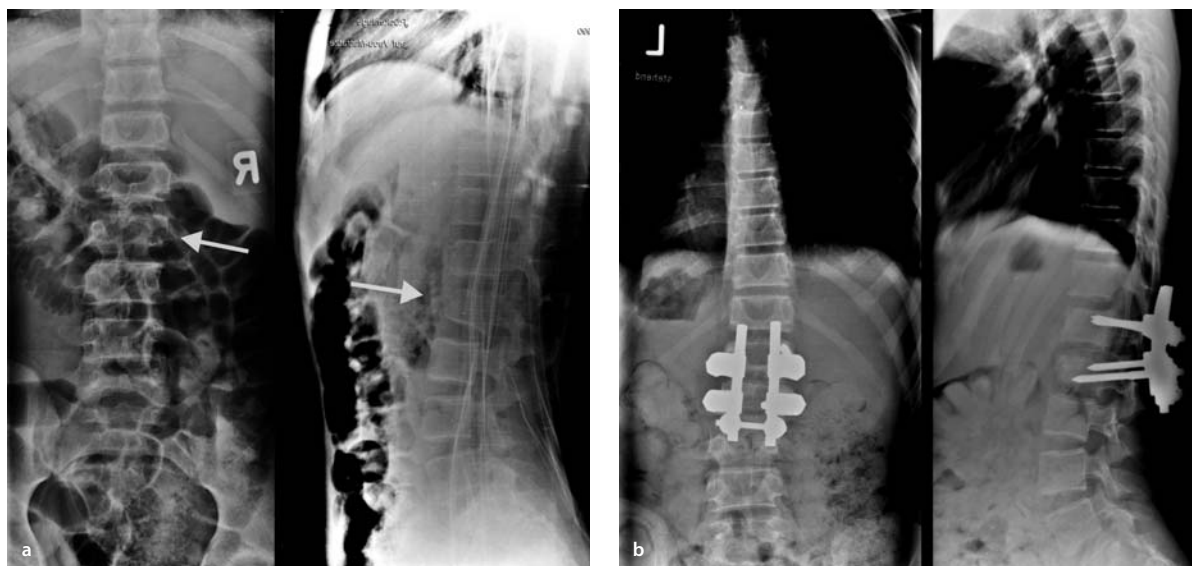
Dorsal instrumentation is primarily used for managing injuries at the *thoracic level*. We tend to use the (pediatric) USS instrumentation. The fact that a number of segments around the fracture will need to be stiffened at the same time does not matter greatly in adolescents at thoracic level. Rather, the important requirements are, firstly, to avoid using excessively large instrumentation since, particularly in small patients, little soft tissue is present and, secondly, to implement any decompression quickly enough. In small children, plates are sometimes better than rods with hooks or screws for stabilization as they take up less space.

At the *lumbar and thoracolumbar junction levels*, the same principles apply to adolescents as for adults. We use

a rod system with transpedicular screws that was originally developed by Dick [6] as a »fixateur interne«. Surgeons can now choose from a variety of modified instruments that are somewhat easier to manage, though still based on the same principle. These systems can be used for the efficient management of fractures in the lower thoracic spine and the lumbar spine (■ Fig. 3.115). The procedure of ligamentotaxis is used to reduce bone fragments in the spinal canal spontaneously, usually by distraction. If, in exceptional cases, this does not prove possible, the spinal canal must be revised, in which case intraoperative myelography will demonstrate the patency of the canal.

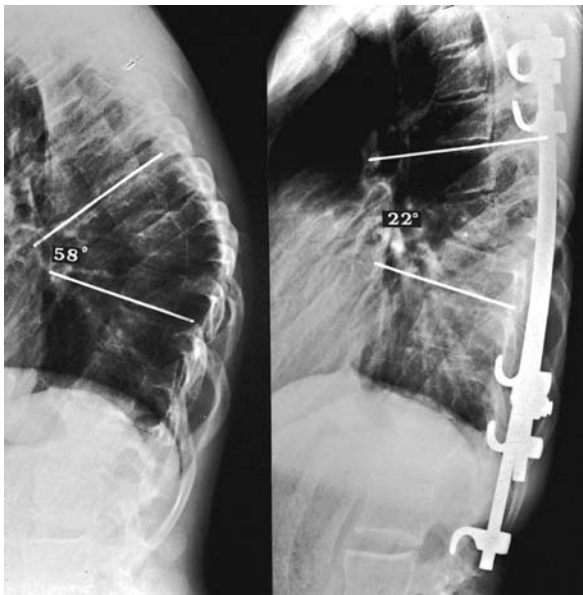
**! Under no circumstances should a laminectomy on its own be performed as treatment for a vertebral fracture with a neurological lesion. This is especially out of the question for small children as the risk of a severe posttraumatic kyphosis developing at a later date is very great. Stabilization of the affected segment is required in every case. This is almost always possible, even in small children, with plates and screws.**

*Secondary deformities* after vertebral fractures occasionally require surgical management. On the one hand this applies to neurogenic scolioses in para- or tetraplegic children, in which case the treatment is similar to that for neurogenic scolioses resulting from other causes (► Chapter 3.1.10). On the other hand, a posttraumatic kyphosis will occasionally require straightening. Exclusively dorsal correction is not sufficient for a badly collapsed wedge vertebra. The vertebral body must always be osteotomied from the anterior side, although this is also perfectly fea-



■ Fig. 3.115a, b. Surgical correction of a burst fracture of L2 with neurological deficits in a 13-year old boy using the USS instrumentation . a AP and lateral x-rays after trauma. b after correction

### 3.1.13 · Inflammatory conditions of the spine



■ **Fig. 3.116.** Posttraumatic kyphosis in a 16-year old male patient (left). Right 1.5 years after surgical correction with ventral disk removal (from the dorsal side) and dorsal tension-band wiring

sible from a purely posterior approach. A good correction can be achieved with a dorsal wedge osteotomy down to the anterior longitudinal ligament (■ Fig. 3.116).

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### 3.1.13 Inflammatory conditions of the spine

#### ► Definition

Inflammatory conditions of the spine resulting from infection and changes associated with rheumatic disorders.

#### 3.1.13.1 Spondylitis, Spondylodiscitis

#### ► Definition

Acute or chronic pyogenic infection of the intervertebral disk or the adjacent vertebral body by non-specific (usually staphylococci) or specific (TB) pathogens. In addition to the destructive form, there is also a benign, self-limiting form restricted to the disk. The disk is almost always affected in this condition. Pure spondylitis without involvement of the disk hardly ever occurs in growing patients.

#### Etiology

Spondylodiscitis is induced by specific or non-specific pathogens. The commonest non-specific pathogen is *Staphylococcus aureus* [2, 3, 10]. Other non-specific

pathogens include *Streptococcus*, *Escherichia coli*, *Salmonella typhosa* and, very rarely, *Kingella kingae* [2], *Brucella abortus* or *Coxiella burnetii* [1]. Specific spondylitis is caused by *Mycobacterium tuberculosis* (human type, rarely bovine type). Although tuberculosis has become a rare illness in Central Europe since BCG vaccination, it remains as common as ever in developing countries. Skeletal tuberculosis is a typical illness suffered by children, and the spine is particularly frequently affected [5, 6, 11, 12].

### Pathogenesis

In small children, blood vessels pass from the endplates into the disks, allowing bacteria to enter the disk by hematogenous transmission. During the course of childhood these vessels are obliterated, thereby preventing any further direct hematogenous infection of the disk. Consequently, the infection always begins in the bone next to the disk in adolescents and adults, but usually directly in the disk itself in small children.

### Occurrence

Spondylodiscitis is a rare condition. We observe about one case a year. Fairly large series of spondylitis TB have been described in Hong Kong [12], India [5] and South Africa [6].

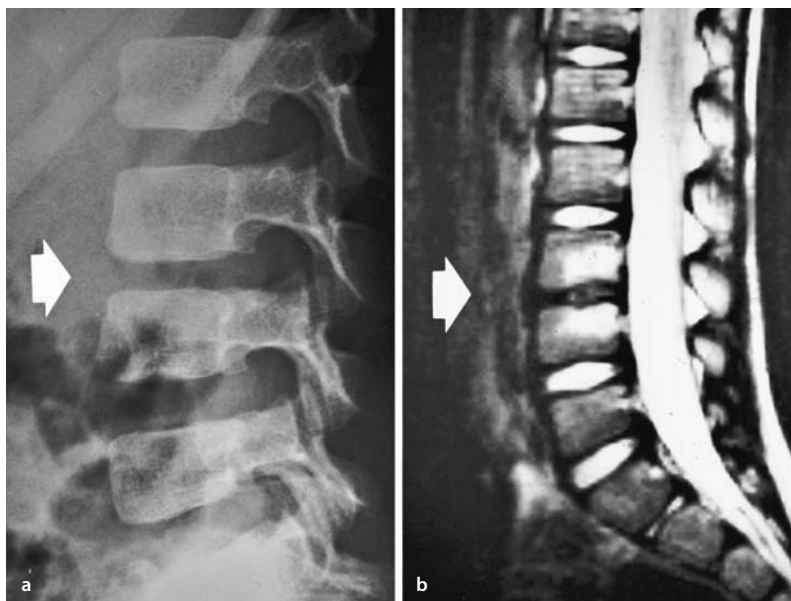
### Clinical features, diagnosis

**! Spondylodiscitis generally occurs in small children under 10 years of age. In this age group one should always consider the possibility of spondylitis when back pain occurs. Even tuberculous spondylitis can occur at a very early age.**

The pain is usually acute and occurs at the level of the affected disk or vertebral body, occasionally radiating caudally and possibly also into the legs or the abdomen. Small children refuse to walk or sit, and often are no longer able to stand up straight. During the clinical examination, the child prefers to adopt a lying position. Severe general symptoms are not usually present, and a high fever is not especially typical. The erythrocyte sedimentation rate and the C-reactive protein level may, but need not be, elevated. A leukocytosis with left shift is not necessarily present. In cases of specific spondylitis, in particular, laboratory parameters of infections are almost invariably absent. If septic temperatures are measured, blood cultures should be taken to identify the pathogen.

### Radiographic findings

The first sign on the *x-ray* is usually the narrowing of the intervertebral space at the affected level (■ Fig. 3.117a), although it takes from 2–3 weeks from the onset of the disease until this finding is visible on a plain *x-ray*. Increased uptake on a *bone scan*, and particularly a *leukocyte scan*, can be detected much sooner. Consequently, a scan should always be recorded in the early stage if the *x-ray* is normal and the clinical findings are suggestive of spondylodiscitis. If increased uptake is found on the scan, an *MRI scan* is recommended. The hyperemia near the affected disk leads to changes that typically appear hypointense on T1-weighted images and hyperintense on T2-weighted images [7] (■ Fig. 3.117b). The disk narrowing visible on the *x-ray* as an early sign is not usually detected on the MRI scan, but only becomes apparent after the inflammatory edema has regressed. An important complication of spondylodiscitis is a psoas



■ Fig. 3.117a, b. 6-year old child with spondylodiscitis L3/L4 in the early stages. a Lateral *x-ray*: note the slight narrowing of the intervertebral disk space. b MRI ( $t_2$ -weighting): in addition to dehydration of the disk at L3/L4, the cord edema in the adjacent vertebral sections is clearly visible

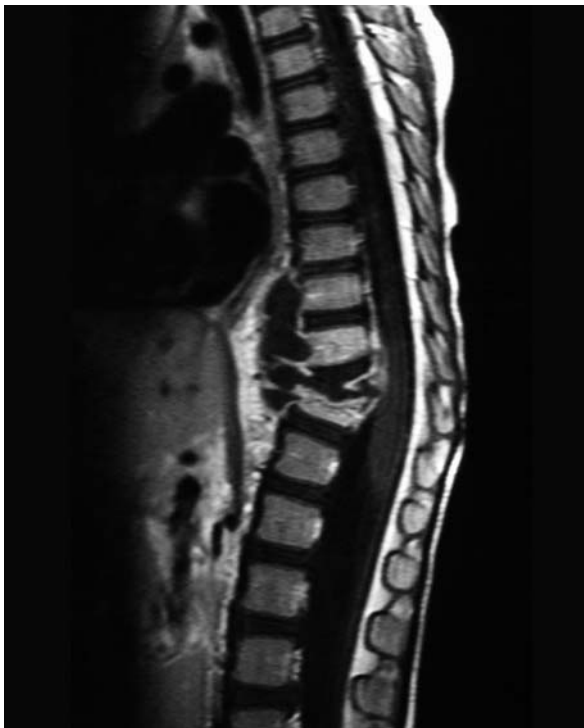
### 3.1.13 · Inflammatory conditions of the spine

abscess [10], which can very easily be seen on the MRI scans. *CT* does not make any further contribution to the diagnosis.

A potentially important diagnostic tool is *vertebral puncture*, which is always indicated if spondylitis TB is suspected (■ Fig. 3.118). This suspicion is then confirmed if, despite a history of several weeks, all laboratory results are negative (apart from a possible slight lymphocytosis) and the TB test is strongly positive. Additionally, the patient's history itself can provide helpful information. A puncture biopsy is also indicated if the infection fails to respond to antibiotic treatment. The puncture is performed under anesthesia from the dorsal side with a wide-bore needle and with image intensifier control. The affected vertebral body is approached on the lateral side of the pedicle and punctured directly. The collected pus is investigated as a direct preparation and in an animal test. However, we do not consider that vertebral body puncture is indicated generally in spondylodiscitis. Providing a specific inflammation is not suspected and the illness responds well to the antibiotics, this investigation – which is not without its risks – is unnecessary.

#### Differential diagnosis

Spondylodiscitis tends to occur in the lumbar area and must be differentiated primarily from tumors and tumor-like lesions, particularly *Langerhans cell histiocytosis*,



■ Fig. 3.118. MRI in a 2-year old child with *spondylitis TB*. The vertebral body T10 is completely collapsed and a large prevertebral abscess is present

although this condition primarily affects the vertebral body and not the intervertebral disk. Other tumors that typically occur in the pediatric spine are *aneurysmal bone cysts* and *Ewing sarcoma*. *Osteoblastoma* is also common, but is generally located in the pedicle and is therefore unlikely to be confused with spondylodiscitis.

#### Treatment

Children with spondylodiscitis should be admitted to hospital. Ideally the spine is immobilized in a plaster brace or a body cast. The main purpose of the brace or cast is to alleviate the pain by immobilization. Since the spondylodiscitis is frequently located in the lumbar area, the brace also helps prevent kyphosis. Since the pathogen usually remains unknown, the anti-infective treatment cannot be targeted specifically and the antibiotics should be administered intravenously in high doses. They should be broad-spectrum preparations with proven efficacy particularly against staphylococci. The intravenous treatment should be continued until the clinical findings and laboratory parameters have returned to normal. In view of the poor circulation in the disk we continue the antibiotic treatment orally for a further three months. During this time, the child must wear a lordosing plaster brace to relieve the affected disk and adjacent apophyseal plate.

*Surgical treatment is indicated in the following situations:*

- a psoas abscess,
- spondylitis TB,
- postinfective kyphosis.

A psoas abscess must always be surgically relieved and drained. Operation is also indicated for a case of spondylitis TB. The affected vertebral body must be removed and the gap bridged with a bone graft. Small children also require dorsal spondylodesis in order to prevent further kyphosing during dorsal growth. If a secondary kyphosis does occur, this may occasionally require surgical correction. The affected section usually has to be straightened simultaneously from the ventral and dorsal sides. The *prognosis* of spondylodiscitis in childhood is quite good. Although approximately 40% sustain a spontaneous fusion of the vertebrae, most patients do not have symptoms according to a long-term follow-up study [8].

#### 3.1.13.2 Spinal changes associated with juvenile rheumatoid arthritis

Changes to the upper cervical spine occur especially in the polyarticular form of juvenile rheumatoid arthritis (► Chapter 4.4.1). One study involving 159 patients with



juvenile rheumatoid arthritis found inflammatory changes of the cervical spine in 98 cases (62%), including ankylosis (41%), atlantoaxial subluxation (17%) and atlantoaxial impaction (25%) [9]. Despite the occasionally extensive radiographic changes of the cervical spine, patients generally suffer few symptoms in this area. Radiographic investigation of the cervical spine is therefore always indicated in the polyarticular form. In particular, the orthopaedist should look for *atlantoaxial instability*, which may occasionally require atlantoaxial screw fixation [4]. The status of the cervical spine should always be checked before operations as the risk of dislocation during intubation is not negligible.

**! The most important complication of the polyarticular form of juvenile rheumatoid arthritis is atlantoaxial instability, whereas iridocyclitis tends to occur in the oligoarticular type.**

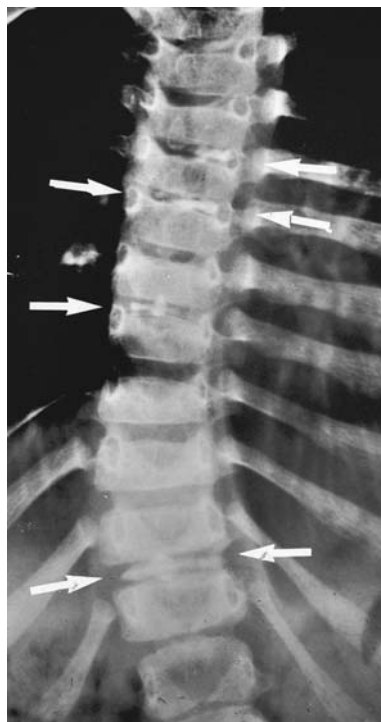
In addition to problems affecting the cervical spine, scolioses can also occur in connection with juvenile rheumatoid polyarthritis, although these are rarely severe enough to require treatment. Brace treatment is indicated in restricted cases, since rheumatic patients always suffer from osteoporosis, which would be further exacerbated by a brace. Surgical correction is also occasionally required.

### 3.1.13.3 Juvenile ankylosing spondylitis

Ankylosing spondylitis (Bechterew disease) is a condition that typically starts in the iliosacral joints during the patient's twenties or thirties. But the disease can also occur as early as the second decade in a small proportion of patients, usually boys. In adolescents the lower extremities tend to be affected rather than the iliosacral joints. The disease shows a distinct familial tendency, and the genetic marker HLA-B 27 has been detected in serological tests. In addition to the major joints, the cervical spine is also frequently affected. Radiographs show arthritis of the iliosacral joints (usually starting on one side). Since these joints already show increased uptake on a bone scan in healthy adolescents, this investigation contributes little to the diagnosis. While drug treatment is rarely indicated in adolescents, physical therapy is particularly important for preserving mobility and avoiding kyphosis. Physical therapy must be continued throughout life in patients with severe ankylosing spondylitis. Efficient gymnastic exercise therapy can prevent stiffening of the spine in a case of severe kyphosis. Surgical measures are rarely indicated at this age. Spinal wedge osteotomies are occasionally required in adults with established severe kyphosis.

### 3.1.13.4 Intervertebral disk calcification

Calcification of several intervertebral disks, particularly in the cervical and thoracic spine, can occur in small



**■ Fig. 3.119.** AP x-ray of the thoracic spine in a 7-year old boy with several spontaneous disk calcifications. These have no pathological significance

children either spontaneously or as the result of an accident. This usually manifests itself on x-rays for only a few months or years and subsequently disappears spontaneously (■ Fig. 3.119). Diffuse symptoms occasionally occur, although the laboratory tests and bone scans show normal findings. Treatment is not required in view of the spontaneous resolution. Temporary immobilization may prove helpful if the patient is experiencing severe symptoms.

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## 3.1.14 · Tumors of the spine

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## 3.1.14 Tumors of the spine

### Definition

Primary bone tumors originating in the vertebral bodies or vertebral arches, or soft tissue tumors arising from muscles, connective tissues, blood vessels or nerve tissues in the immediate vicinity of the spine.

### Occurrence

Only 10% of all primary bone tumors are located in the spine, 85% of which are benign.

In order to obtain an indication of the distribution of tumor types affecting the spine in children and adolescents we have been registering cases in the Basel Bone Tumor Reference Center since 1972. A total of 80 primary bone tumors have been registered in under-20-year-olds during this time, compared to 183 primary bone tumors in adults. Table 3.11 shows the diagnostic distribution. Only four tumors were malignant (3 Ewing sarcomas, 1 osteosarcoma). The commonest tumors were osteoblastoma and aneurysmal bone cyst, which each occurred in around a quarter of cases. All other tumors only occurred sporadically. Even giant cell tumor of the sacrum and chordoma, which were fairly common in adults, were extremely rare in children and adolescents. Osteoblastoma was slightly less predominant in adults than in adolescents, while aneurysmal bone cysts were hardly observed in adults at all.

The tumors are distributed very regularly across all segments, without any one preferred region. Only the sacrum is affected to a slightly greater extent (particularly by giant cell tumors and chordomas). Osteoblastomas are slightly more likely to affect the lumbar area than the thoracic or cervical spine [26].

Of the malignant tumors, osteosarcomas and chondrosarcomas are less commonly seen in children and ado-

Table 3.11. Diagnoses of primary tumors of the spine in children and adolescents (n=80) compared to adults (n=183) (Basel Bone Tumor Reference Center)

	Children and adolescents		Adults	
Osteochondroma	3	3.8%	11	6.0%
Chondroblastoma			1	0.5%
Enchondroma	1	1.3%	1	0.5%
Aneurysmal bone cyst	18	22.5%	4	2.2%
Non-ossifying fibroma of bone	1	1.3%		
Giant cell tumor	5	6.3%	23	12.6%
Fibrous dysplasia	1	1.3%	1	0.5%
Osteoid-osteoma /osteoblastoma	27	33.8%	24	13.1%
Hemangioma	3	3.8%	15	8.2%
Other benign tumors	4	5.0%	5	2.7%
<i>Langerhans cell histiocytosis</i>	3	3.8%	1	0.5%
<i>Unicameral bone cyst</i>			2	1.1%
<i>Other tumor-like lesions</i>	2	2.5%	2	1.1%
<b>Osteosarcoma</b>	<b>2</b>	<b>2.5%</b>	<b>16</b>	<b>8.7%</b>
<b>Ewing sarcoma/PNET</b>	<b>4</b>	<b>5.0%</b>	<b>5</b>	<b>2.7%</b>
<b>Chondrosarcoma</b>	<b>2</b>	<b>2.5%</b>	<b>23</b>	<b>12.6%</b>
<b>Chordoma</b>	<b>2</b>	<b>2.5%</b>	<b>21</b>	<b>11.5%</b>
<b>Plasmacytoma</b>			<b>11</b>	<b>6.0%</b>
<b>Other malignant tumors</b>	<b>2</b>	<b>2.5%</b>	<b>17</b>	<b>9.3%</b>
<b>Total</b>	<b>80</b>	<b>100.0%</b>	<b>183</b>	<b>100.0%</b>

lescents than Ewing sarcomas. Our register only records two primary osteosarcomas in this age group, whereas this tumor occurs in adults with above-average frequency in the spine. In an overview of the European multicenter study on the treatment of osteosarcoma (COSS), 22 of these tumors were located in the spine (including 15 in the sacrum). Out of 975 Ewing sarcomas recorded in the European multicenter EICESS study, 78 (8.0%) were located in the spine [12].

### Diagnosis

We know from a study on benign tumors of the cervical spine, that only 70% of the tumors are visible on a conventional x-ray, even when other imaging techniques have shown a tumor to be present [21]. It is all the more difficult, therefore, to diagnose tumors of whose existence the doctor is completely unaware.

**! Severe back symptoms are rare in children and adolescents. In view of the difficulty with radiographic diagnosis, a bone scan should be arranged within a reasonable period if the patient complains of pain that is *not* load-related.**

Just how difficult it can be to diagnose a spinal tumor in young patients is shown by a study involving 22 patients with osteoblastomas of the spine, which were only diagnosed, on average, some 16 months after the start of symptoms [26].

A bone scan is a cost-effective investigation for demonstrating the presence of a neoplastic process with a very high probability and also for indicating its location. Only if the scan shows a positive uptake should further investigations such as MRI and CT scans be arranged. An MRI scan is essential in malignant processes. As a general rule, an MRI scan is indicated for all tumors that emerge from the bone and show significant soft

tissue infiltration (particularly intraspinal infiltration). Tumors that remain within the bone, on the other hand, are better visualized in the spine by CT. This particularly applies to the relatively common spinal tumor of osteoblastoma.

### Differential diagnosis

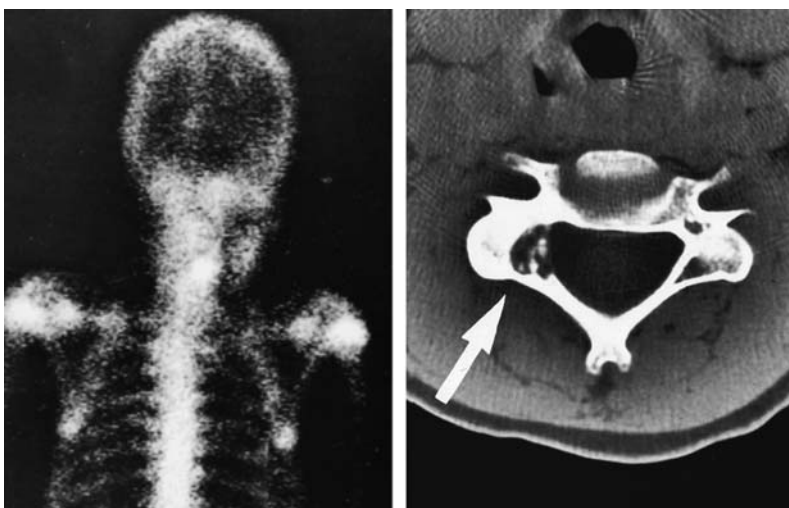
The most important differential diagnosis in relation to tumors of the spine is infection, i.e. spondylitis and spondylodiscitis. The common symptom is non-load-related pain that can also occur at night. The laboratory results usually indicate whether an infection is present or not, but it should be borne in mind that chronic infections often show only minimal, or even no, changes in the blood film. This also applies to tuberculosis and brucellosis.

### Typical features of the principal tumor types

#### Benign and semi-malignant tumors

##### Osteoblastoma

Next to aneurysmal bone cysts, osteoblastomas are the commonest bone tumors found in the spine in children and adolescents. They occur 2.5 times more frequently in male patients than females [24] and tend to be located at lumbar level [27]. Osteoblastomas cause diffuse pain that frequently occurs at night. As with osteoid osteomas, the pain responds well to aspirin, although the effect is not as strong in osteoblastoma. In histological respects, an osteoblastoma is identical to an osteoid osteoma. An osteoid osteoma is located in the cortex of long bones and produces substantial amounts of osteoid. The tumor itself remains as small as a grain of rice («nidus»). An osteoblastoma is located in cancellous bone and forms less osteoid, though the tumor itself can be much larger. A typical site in the spine is the pedicle or vertebral arch (■ Fig. 3.120). Occasionally, the tumor also occurs in the



■ Fig. 3.120. Bone scan (left) and CT (right) in a 7-year old girl with osteoblastoma in the vertebral arch of C6

vertebral body [6]. Certain authors also make a distinction in the spine between clearly demarcated osteoid osteomas and osteblastomas with their more diffuse margins and which can also project from the bone into the soft tissues [29]. Making this distinction can often prove difficult in individual cases. However, since both tumors are histologically identical, this distinction is not particularly useful.

Apart from pain, the tumor can also lead to antalgic scoliosis of the spine. In this case the osteblastoma is usually located at the apex of the curve, and the tumor always affects the pedicle on the convex side of the scoliosis. The prognosis for the scoliosis depends on the duration of symptoms until treatment [9]. A spontaneous course is characterized by pain that can persist for years. Cases of neurological complications resulting from the penetration of the tumor into the spinal canal have been described in the literature [6, 24], even including a fatal outcome in a patient with cervical tetraplegia caused by an osteblastoma. Spontaneous recoveries also occur.

### Treatment

Treatment of the osteblastoma of the spine consists of simple curettage, usually without the need for more extensive treatment [9]. At least a marginal resection is recommended for larger osteblastomas with diffuse margins, and possibly with soft tissue components [6]. If intralesional excision only is possible, subsequent radiotherapy is indicated [6]. We consider the use of a gamma camera during the operation [25] to be unnecessary as the lesion can be visualized very accurately on the CT scan, and locating the tumor poses little difficulty during the operation. A CT-controlled percutaneous excision of the tumor seems an elegant option. However, since the tumor is located in the thin pedicle, which is easily perforated by the drill, at the spinal level (in contrast with the long bones) we prefer open curettage. Since the recurrence rate is low even after simple curettage and malignant degeneration of the spine has not been reported, we would warn against overtreatment.

### Aneurysmal bone cysts

Aneurysmal bone cysts can occur primarily or secondarily in association with other tumors. When they occur secondarily, the possible primary tumors include osteoblastomas, giant cell tumors and osteosarcomas [35]. Around one quarter of aneurysmal bone cysts are the secondary form [35], although these hardly ever occur in the spine. 75% of aneurysmal bone cysts occur in under-20-year olds [8]. As regards etiology, they are thought to result from an arteriovenous fistula within the bone. Genetic factors also appear to be involved and a familial tendency has been described [13].

A solid form of aneurysmal bone cyst that is histologically very similar to an extragnathic giant cell reparative

granuloma has also been described [4]. The recurrence rate for the solid variant appears to be rather lower than that for a standard aneurysmal bone cyst [4]. Active aneurysmal bone cysts can grow very rapidly and destroy the bone within a correspondingly short period (■ Fig. 3.121). Simple curettage is often not sufficient in such cases and frequently results in recurrences. On the other hand, cases of spontaneous recovery in the spine have also been reported [23]. Neurological lesions as a complication of an aneurysmal bone cyst in the spine have also been observed in isolated cases [10], although these appear to be extremely rare.

### Treatment

Since aneurysmal bone cysts are associated with a relatively high rate of recurrence after simple curettage [8, 15], a marginal resection at least should be attempted. The quality of the resection crucially affects the recurrence rate. If the cyst cannot be resected en bloc, the margins must be drilled out. We consider the use of liquid nitrogen, phenol, methyl methacrylate etc. to kill the cells at the margins of the resected cyst to be too dangerous in the spine as there is no enclosed tumor cavity. However, the use of a Cavitron ultrasonic unit can enhance the quality of the resection. Campanacci recommends simple curettage for inactive cysts (types I–III, ► see chapter 4.5.2) and en bloc resection (where this is possible) with additional irradiation for types IV and V.

### Langerhans cell histiocytosis

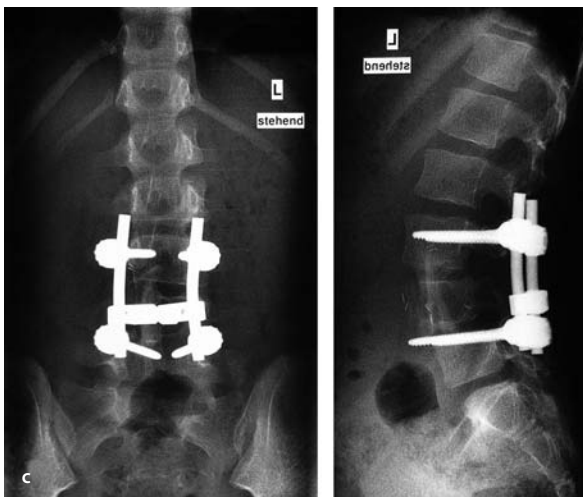
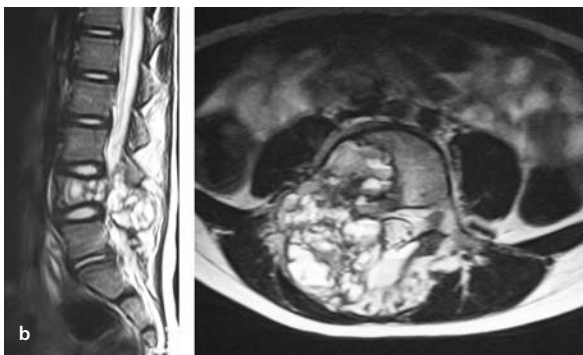
Langerhans cell histiocytosis can, in principle, occur in any bone, including the bones in the spine. Langerhans cell granulomas occur in monostotic or polyostotic forms, with or without visceral involvement. In the spine, the vertebral body is usually affected, less commonly the vertebral arch. Of the 139 cases of Langerhans cell granulomas recorded in the Basel Bone Tumor Reference Center, six involved the spine [19]. While the granulomas can essentially occur at any age, they tend to be more common in the first and second decade of life (■ Fig. 3.122). They can be located in any vertebral body, with no preference for a particular part of the spine.

Typical findings in Langerhans cell histiocytosis in the spine are the collapse of vertebral bodies and the formation of a vertebra plana (■ Fig. 3.123). A vertebra plana develops because both the anterior and posterior parts of the vertebral body are affected, hence the absence of a wedge shape. The formation of a vertebra plana is *not* associated with neurological complications, and only very isolated cases involving neurological symptoms have been described in the literature [1, 36]. The fact that neurological complications rarely occur despite the dramatic change in the shape of the vertebral bodies is partly explained by the absence of wedge vertebrae and consequent kyphosis and partly by the consistency of the

granuloma itself, which consists of a soft, almost liquid, mass distributed around the spinal cord without exerting any compression.

If a focal finding in the spine suggests the presence of histiocytosis, the diagnosis should be confirmed by

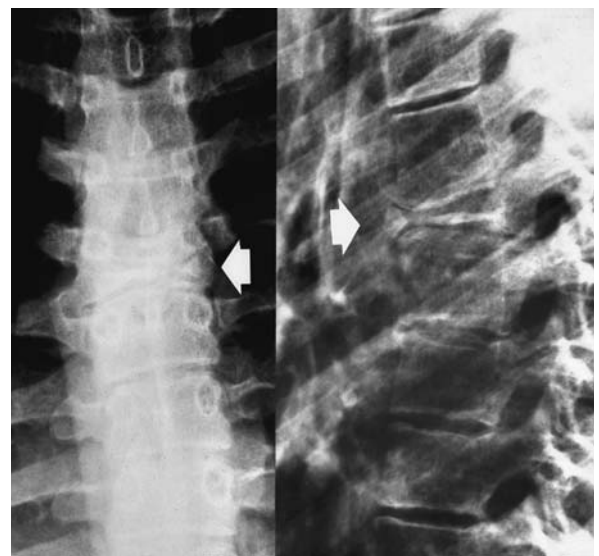
a biopsy, which can be performed either as an open procedure or transcutaneously as a needle biopsy under image intensifier control. If a polyostotic form of Langerhans cell histiocytosis has already been diagnosed on the basis of a biopsy from another focus, a further biopsy



**Fig. 3.121a–c.** Aneurysmal bone cyst in a 9-year old girl in the area of the vertebral body and arches of L4. **a** The right pedicle is missing on the AP x-ray, while the tumor is barely visible on the lateral view. **b** MRI: sagittal and horizontal views in the same female patient. Large tumor masses in the vertebral body on the right, in the pedicle and the vertebral arch with fluid levels. **c** Situation after en-bloc resection, bridging with autologous fibula and stabilization with USS instrumentation



**Fig. 3.122.** MRI scans of the lumbar spine in Langerhans cell histiocytosis of the vertebral body of L4 in a 2-year old girl. Despite the constriction of the spinal canal, no neurological symptoms were present



**Fig. 3.123.** X-rays of the thoracic spine in a 17-year old female patient with a history of Langerhans cell histiocytosis 10 years ago and a *vertebra plana*. The female patient is symptom-free



### 3.1.14 · Tumors of the spine

is not necessary. Solitary foci do not require treatment since they recover spontaneously without any clinical sequelae. The vertebral bodies do not correct themselves as growth continues and the vertebra plana persists throughout life. However, since no kyphosis or structural deformity ensues, the patients remain asymptomatic.

#### Other benign tumors

While almost any benign tumor can occur in the spine in principle, most very rarely do so. Apart from the types already described, we have personally encountered osteochondromas, desmoplastic fibromas, giant cell tumors and fibrous dysplasia. Treatment is based on the symptoms and the functional impairment. A typical benign tumor in children and adolescents is the chondroblastoma, which occurs almost exclusively in the epiphyses of long bones and is therefore extremely rare in the spine.

#### Malignant tumors

##### Chordoma

Chordomas are tumors that develop from the remnants of the embryonal notochord. Such remnants are found in approx. 2% of all autopsied individuals, but result in tumor growth in only very few cases [34]. The tumor itself is a primarily malignant, slowly growing neoplasm that rarely metastasizes. It is located in the sacrum in 50% of cases, at the base of the skull in 35%, and in the mobile part of the spine (cervical, thoracic or lumbar spine) in only 15% of cases. At the sacral level the chordoma accounts for 40% of all primarily malignant tumors [34]. While the tumors can occur at any age, they are rare in childhood and adolescence. They are typically seen in patients aged from 40 to 60. Males are twice as likely to be affected as females [5].

The primary clinical finding in a patient with this tumor tends to be pain. A radiograph shows non-specific osteolysis. A widening of the interpedicular distance on the plain x-ray can provide an initial indication of the presence of a chordoma since it is a highly typical finding for this tumor. Neurological problems occur at a relatively late stage. Additional diagnostic techniques include a bone scan, CT and MRI scans. On an MRI scan, the tumor shows relatively low signal intensity on the T1-, T2- and proton-weighted images.

Current recommended treatment is the widest possible surgical excision and subsequent irradiation of the tumor with approx. 50 Gy [3, 5, 30, 31]. Five-year survival rates ranging from 78% [5] to 84% [3] and ten-year survival rates ranging from 36% [5] to 64% [3] have been reported after this combined treatment. Since half of the tumors occur in the sacrum, individual sacral roots often need to be sacrificed during the resection [31].

##### Osteosarcoma

While the spine is not a typical site for an osteosarcoma in young people, the tumor can occur in this part of the body (■ Fig. 3.124). Treatment is essentially based on the guidelines described in ► chapter 4.5.6. Osteosarcomas are initially treated for 3 months by chemotherapy according to the EURAMOS protocol. The tumor is then resected. Since the wide resection required for an osteosarcoma is only possible in very few cases in the spine, given the need to preserve the cord, the response to the chemotherapy is crucial for the prognosis. A wide resection at the cost of spinal cord function would probably not be acceptable to most patients. Radiotherapy of a spinal osteosarcoma is recommended even though this tumor is not very sensitive to radiation [27].

##### Ewing sarcoma

Ewing sarcoma (and the related *primitive neuroectodermal tumor*; PNET) is the commonest malignant spinal tumor in children and adolescents (■ Fig. 3.125). Ewing sarcomas are primarily treated by chemotherapy according to the EICESS protocol. Although surgical resection is not generally recommended, it may be appropriate in individual cases (solitary tumor in a relatively favorable location, no metastases detected). Since Ewing sarcomas are radiosensitive, radiotherapy is indicated after the initial chemotherapy. Chemotherapy is subsequently continued until its conclusion after a total of one year.

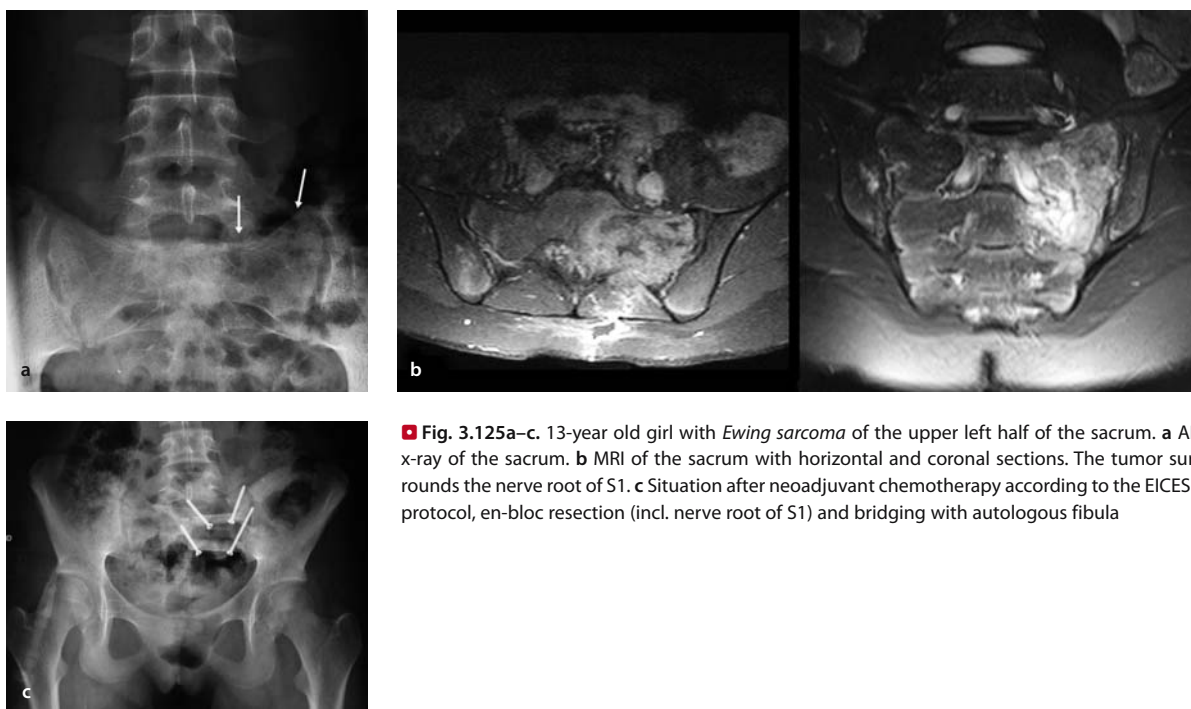
##### Chondrosarcomas

Chondrosarcomas rarely occur in children and adolescents. As for chondrosarcomas in other areas of the body, treatment is primarily surgical for the spinal forms, although the required wide resection is likely to prove feasible only in the rarest of cases. Consequently, sub-



■ Fig. 3.124. AP and lateral x-rays of a 12-year old girl with an osteosarcoma of the vertebral body of L2





**Fig. 3.125a–c.** 13-year old girl with *Ewing sarcoma* of the upper left half of the sacrum. **a** AP x-ray of the sacrum. **b** MRI of the sacrum with horizontal and coronal sections. The tumor surrounds the nerve root of S1. **c** Situation after neoadjuvant chemotherapy according to the EICESS protocol, en-bloc resection (incl. nerve root of S1) and bridging with autologous fibula

sequent radiotherapy is required following a marginal resection [7].

### **Intraspinal tumors and neoplasms associated with congenital anomalies**

#### **Systemic diseases**

##### **Neurofibromatosis**

The commonest intraspinal tumor is *neurofibromatosis*. This hereditary disorder is described in detail in ► chapter 4.6.8.6, while the secondary spinal deformities are discussed in ► chapter 3.1.11.1. The neurofibromas located in and around the spinal canal can lead to a characteristic scoliosis: short-curved, strongly progressive and often with a pronounced kyphotic component, the vertebral bodies are indented and the ribs drawn out. Particular risk factors for strong progression are the early onset of the scoliosis, a pronounced kyphotic component and an apex located in the lower thoracic spine [16]. Intraspinal neurofibromas are present in what is known as the dystrophic type, which accounts for approx. 70–80% of the scolioses associated with neurofibromatosis [33]. On the other hand, only around 15% of patients with this disease suffer a scoliosis [2]. Neurological symptoms occur in 15% of cases, and paravertebral soft tissue tumors are also occasionally observed. Atlantoaxial abnormalities are also not infrequently seen [14]. Solitary intraspinal neurofibromas that are unconnected with a generalized neurofibromatosis occasionally occur [32]. Neurofibromas can lead to neurological problems. The most serious complication is degeneration into a malignant schwannoma [11]. We ourselves have had the misfortune to observe four such

cases. Malignant schwannomas are relatively slow-growing tumors, but hardly respond at all either to chemotherapy or radiotherapy. Since the surgical options in the area of the spinal canal are very limited, the patients die after a few years. The patients treated by us also suffered this fate.

##### **Local disorders**

Intraspinal tumors occur either as independent neoplasms of the nerve tissue and spinal meninges or in association with other anomalies of the spine or certain hereditary disorders.

In patients with congenital scoliosis, concealed intraspinal anomalies are present in approx. 20% of cases, and 10% of these are tumors (teratomas, lipofibromas, etc.) [28]. Intraspinal anomalies are particularly common in unilateral segmentation defects with a contralateral hemivertebra. Intraspinal tumors have also been described in connection with the Klippel-Trenaunay-Weber syndrome [17].

Other local intraspinal tumors (which occur primarily in the area of the cauda equina): ependymomas (commonest), astrocytomas, lipomas and teratomas.

**! Particular caution is always indicated when scolioses occur in children under 10 years [18]. Intraspinal anomalies are found in approx. 20–30% of these patients and intraspinal tumors are not infrequently observed as well. An MRI scan should always be recorded before therapeutic measures are initiated (especially of course before surgical correction) for such scolioses.**

### 3.1.15 · Why do backs that are as straight as candles frequently cause severe pain?

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### 3.1.15 Why do backs that are as straight as candles frequently cause severe pain? – or: the differential diagnosis of back pain

It is a popular misconception to believe that a crooked back is associated with corresponding pain. But this is certainly not the case with children and adolescents. Insofar as the shape of the back can be used as a criterion at all, it tends to be the strikingly straight back that gives rise to pain in the young, since the commonest cause of serious symptoms in this age group is (thoraco-) lumbar Scheuermann's disease, which is associated with a flat back.

#### Occurrence

Whereas back pain in children and adolescents was once thought to be a very rare phenomenon, more recent studies have shown that symptoms in the region of the spine are not especially rare. An epidemiological study in Den-

mark with 29,000 individuals showed that, by the age of 20, 50% of the interviewees of both sexes had experienced back pain [5]. The risk of further back pain in the future is twice as high for this group as for asymptomatic subjects [4]. Earlier studies showed a lower incidence of back pain in adolescents [1, 2, 6].

### Clinical features

#### History

When a patient attends a consultation with back pain, we must ask the *following questions* (■ Fig. 3.126):

- *When did the symptoms start?*
- *Has trauma occurred? If so – nature of the trauma, precise circumstances?*
- *Is the pain related to certain activities? – Does it occur, for example, after prolonged sitting, standing or sporting activity?*
- *Does the pain occur at night?*
- *Duration of the pain? – Does it only last for a few minutes or hours each time or does it continue for days?*
- *Frequency of the pain? – Every day, once a week, once a month?*

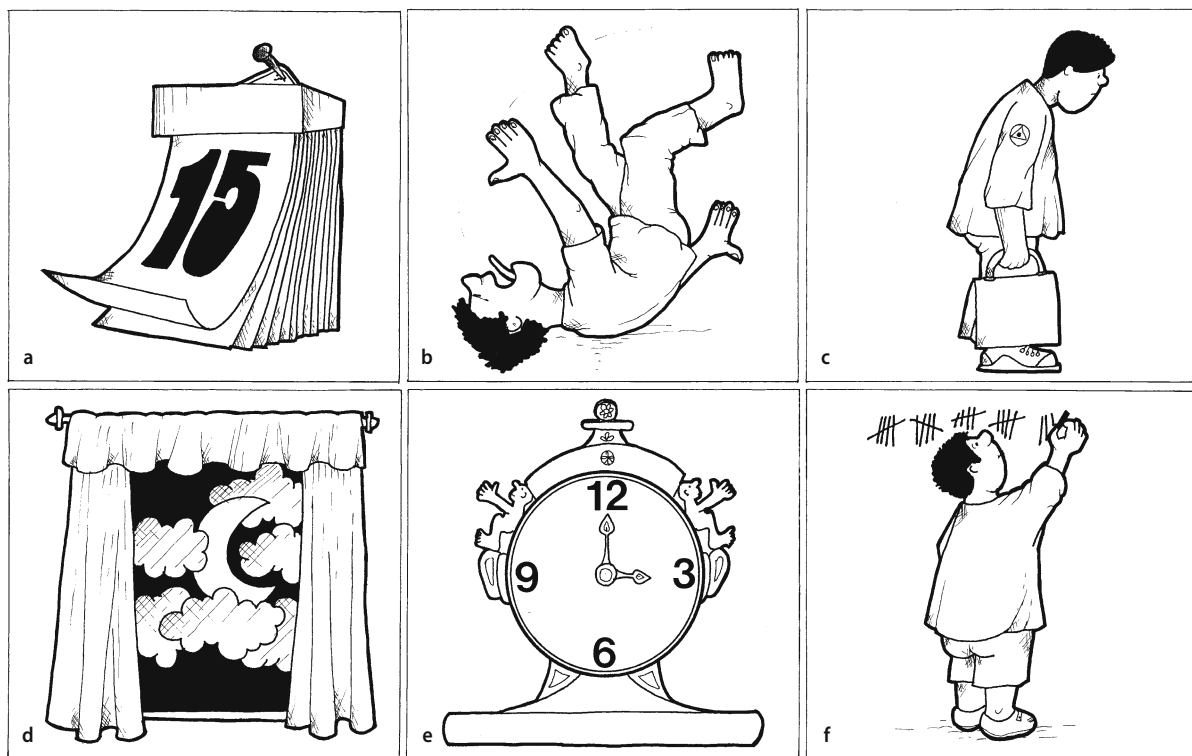
#### Examination findings

During the general *back examination* (▶ Chapter 3.1.1) we pay particular attention to the following points:

- *Restriction of mobility:* Increased finger-floor distance, reduced inclination, reclination or lateral inclination to the left and right
- *Is antalgic scoliosis present?*
- *Muscle tenderness?* In particular, we palpate the back for any muscle spasm (myogelosis): This can be felt as a very rough and painful cord within the paravertebral muscles. An experienced examiner usually knows exactly when the patient is hurting and not hurting during this procedure, even when the patient keeps quiet.
- *Pain on pressure and vibration of individual spinous processes?*
- *Pain on reclining?*
- *Pain during the heel-drop test?*
- *Pain on coughing or sneezing?*
- *Neurological symptoms?*

#### Differential diagnosis

A particular problem when assessing back pain in children and adolescents is the fact that, not infrequently, a diagnosed spinal condition is not actually responsible for the pain. Many – in some cases impressive – spinal diagnoses rarely, or never, cause symptoms in young patients, although they can produce pain at a later stage during adulthood. This is most conspicuously the case with thoracic scoliosis.



■ Fig. 3.126a–f. While taking a history we must ask the following questions: a *When did the symptoms start?* b *Has trauma occurred?* c

Is the pain *related to certain activities?* d *Does the pain occur at night?* e *Duration of the pain?* f *Frequency of the pain?*

### 3.1.15 · Why do backs that are as straight as candles frequently cause severe pain?

❗ **Even a very severe case of thoracic scoliosis does not usually elicit any pain during adolescence. The pain only arises when decompensation occurs, i.e. when the spine falls out of alignment.**

Some deformities of the spine also follow a completely benign course and do not cause any pain. What is less well known is that this also applies to *thoracic Scheuermann's disease*. Even a very pronounced case of fixed thoracic kyphosis with clearly visible Schmorl nodes on the x-ray and wedge vertebrae these changes are not generally responsible for back pain, nor does such pain occur more frequently.

❗ **By contrast, patients with Scheuermann disease located at the thoracolumbar or even lumbar level suffer very severe back pain on a regular and frequent basis.**

Another finding that is likewise not responsible for back pain is a *leg length discrepancy*. This is very common in children and adolescents but can never be blamed for causing the lumbago.

Those disorders that are actually the cause of symptoms are listed below.

#### Disorders that can trigger severe back pain

- *Lumbar or thoracolumbar Scheuermann's disease*  
This is probably the commonest cause of severe, in some cases very severe, back pain in adolescents. The pain is generally related to activity, but can also persist at night. Such pain can cause major problems particularly during the florid phase of the disease. The diagnosis is easily confirmed on the basis of AP and lateral x-rays of the spine.
- *Spondylodiscitis*  
Spondylodiscitis is a diagnosis that should always be borne in mind, even though it is a rare condition. It occurs in children and adolescents by hematogenous transmission and can be extremely painful. The pain tends to be stronger at night than during the day. In many cases, the children are hardly able to walk. The x-ray is not particularly striking in the initial stages, and even the laboratory test results frequently fail to show any impressive changes, although the erythrocyte sedimentation rate, leukocyte count and C-reactive protein are usually elevated. If the plain x-ray is not unequivocal and if this diagnosis is suspected then (leukocyte) scintigraphy should be arranged. If the scan shows highly localized substantial uptake, an MRI scan is indicated. The most important differential diagnosis is a tumor (▶ see chapter 3.1.13 for further details).
- *Tumors*  
Tumors of the spine are not all that rare in children. Two tumors in particular are found: *osteoblastoma*

and *aneurysmal bone cyst* (▶ Chapter 3.1.14). The osteoblastoma especially is very painful, and nocturnal pain is a very typical symptom. Fairly small tumors, and thus osteoblastomas as a rule, are relatively difficult to see on plain x-rays. Since they are usually located in the pedicles these must be scrutinized very carefully. If a tumor is suspected, a bone scan should be arranged. The laboratory findings are always negative. If the bone scan is positive, an MRI scan can provide further useful information, although intraosseous tumors are better viewed on a CT scan. In addition to these two tumors, the tumor-like lesion of Langerhans cell histiocytosis is also not infrequently seen. This generally leads to the collapse of the vertebral body to produce a vertebra plana.

#### — *Fractures*

Fresh fractures can, of course, also be responsible for very intense back pain. These are usually compression fractures. Thoracic fractures can be fairly difficult to detect on the x-ray, and it is not always easy to distinguish them from wedge vertebrae in a case of Scheuermann's disease (▶ see chapter 3.1.5). The patient's history usually proves helpful, although not every trauma necessarily results in a fracture and sometimes even relatively violent traumas are not reported as such (e.g. if they occurred during a »forbidden« activity).

#### Disorders that can be responsible for mild or moderately severe back symptoms

##### — *Spondylolysis, Spondylolisthesis*

Spondylolysis is a common condition (occurring in approx. 5% of the population), but is asymptomatic in most cases. It can occasionally cause pain in young patients and remain symptomatic over a fairly prolonged period [3]. The cause of the pain in this age group is usually the spondylolysis itself rather than the disk degeneration (in contrast with the situation in adults). The spondylolysis is usually clearly visible on lateral x-rays of the lumbosacral junction (■ Fig. 3.68).

In case of doubt, oblique x-rays are worthwhile. In the initial stages, however, the lysis may not yet be properly visualized even on these images. In such cases, a bone scan will show highly localized uptake at those points where the lysis has developed. On clinical examination the pain is found to be located at the lumbosacral junction. A very specific finding on palpation is pain on vibration of the spinous process L5. Reclination pain when leaning as far back as possible is another typical finding.

##### — *Severe lumbar scoliosis*

These scolioses can, particularly in cases of loss of balance, lead to lumbar symptoms during adolescence, although this rarely occurs.



### — *Herniated lumbar disk*

Herniated disks are very rare during adolescence [7]. The pain tends to be located in the hip rather than the back, and is occasionally combined with neurological deficits. The diagnosis can usually be confirmed by an MRI scan or myelography. Since a herniated disk in adolescents generally responds well to conservative treatment, surgical interventions are rarely required.

### — *Intraspinal anomalies*

Anomalies such as a  *syrinx*  or bar can occasionally cause symptoms, although they are usually pain-free and primarily cause neurological deficits, e.g. in a  *tethered-cord syndrome* . Since a syrinx can also be responsible for an unusually shaped »idiopathic« scoliosis, we arrange an MRI scan for all atypical idiopathic scolioses (e.g. if they show a left-convex curve at thoracic level).

### — *Muscle spasms (myogeloses)*

Muscle spasms can occur even in adolescents after unusually strenuous physical exertion. These can persist for weeks or sometimes even for months. They are readily palpated during examination and respond rapidly to physical heat treatment and physical therapy exercises. Myogeloses are the commonest tangible cause of back pain in adolescents.

In an investigation of 100 consecutively examined patients attending our office or outpatient clinic for back pain we were able to establish the cause in 43 cases (■ Table 3.12). Although we diagnosed a spinal condition in a third of the patients, this was probably not responsible for the symptoms. In the end we were unable to find any cause for the symptoms or make any diagnosis in just under a quarter of the patients. The patients were documented at the first visit at which they complained of back pain. Parents with children can attend our outpatient clinic for consultation without referral and without experiencing long waiting times. If we had registered them after a 4-week history the proportion of diagnoses that were actually responsible for the symptoms would certainly have been much higher. Nevertheless, the investigation shows that back symptoms without a discernible cause occur relatively frequently in children and adolescents as well, even though the symptoms are rarely as severe and persistent as they are in adults. Back pain in growing patients should always be taken seriously. If the pain is intense, immediate and targeted investigation is indicated. If the pain is only moderate, more specific diagnostic investigations should be initiated by 4 weeks, at the latest, after the onset of the symptoms (► Chapter 3.1.6, where the indications for more comprehensive diagnostic imaging investigations are listed according to the symptoms).

❗ **If you hear barking outside your front door, your natural and immediate inclination is to think that a dog is responsible for the barking, before considering the possibility that it might be a wolf. Among the causes of back pain (apart from myogeloses), spondylolysis and lumbar Scheuermann disease are the dogs...**

Regardless of the triggering pathology (instability in spondylolysis or fractures, adverse statics in lumbar Scheuermann disease or scolioses), most back pain is attributable to muscle spasms. All passive measures can, at best, merely relieve the spasm temporarily, but will not be able to prevent a further contracture shortly thereafter. In the long term, the muscles can only be relaxed by  *cyclical exercise* , i.e. regular muscle training (with alternating contraction and relaxation of the muscles). Making patients understand this fact and encouraging them to undertake daily exercise is one of the doctor's most important tasks.

### ***Basic reflections on therapeutic measures***

Nowadays it is not so easy to make it clear to patients that they themselves must actively do something to remain free of symptoms. There is a prevailing attitude that the medical industry is responsible for making people healthy by prescribing easily consumed drugs. In our low-exercise era, muscles are subjected almost exclusively to static loads (sitting, standing). While comparable studies may not be available, there is no doubt that back pain was very rare in young people 100 years ago. At that time children had to walk long distances to school, whereas children nowadays are transported to school by their parents even over distances as short as 500 yards. Over the course of the last 100 years we have replaced almost all muscle activity by machines.

This situation is paralleled by the growth of sport. But sport is essentially a voluntary activity, and exercise-averse children always manage to find ways of avoiding sport at school. On the other hand, sport can also lead to muscle spasms if it involves uncoordinated, excessive activity, which makes it difficult for people to appreciate that (appropriate) muscle activity is the solution for avoiding pain in the long term. Moreover, the pain can be aggravated at the start of cyclical exercise of a spastic muscle. Patients often complain that the pain has got worse after physiotherapy. While this doubtless applies to most patients, this phase must be overcome. The muscles cannot be trained sufficiently with just half an hour of physiotherapy exercises once or twice a week. The training must take place on a daily basis. Our musculoskeletal system is designed to move 12 hours a day. These days, adolescents hardly have time to activate their limbs for half an hour each day.



## 3.1.15 · Why do backs that are as straight as candles frequently cause severe pain?

■ **Table 3.12. Causes of back pain in 100 patients in our outpatient clinic**

	Number	Intensity
Diagnoses that were probably the cause of symptoms		
Myogelosis	17	+
Spondylolysis/spondylolisthesis	9	++
Lumbar or thoracolumbar Scheuermann disease	7	+++
Fracture	2	+++
Severe lumbar scoliosis	2	++
Tumor	2	+++
Intraspinal anomaly	1	+
Juvenile rheumatoid arthritis	1	+
Herniated disk	1	++
Spondylodiscitis	1	+++
<b>Total</b>	<b>43</b>	
Diagnoses that were probably not responsible for the symptoms		
Leg length discrepancy	12	
Thoracic Scheuermann disease	7	
Mild lumbar scoliosis	6	
Thoracic scoliosis	5	
Congenital anomaly of the spine	4	
<b>Total</b>	<b>34</b>	
Patients without diagnosis		
<b>Pain of unknown origin</b>	<b>23</b>	

The problem starts at school. Children and adolescents sit for 45 minutes. The original aim of the break was to let the students exercise. But nowadays the youngsters simply lug their books to another classroom, smoking a cigarette or a joint on the way. Playful exercise is increasingly rarely seen in school playgrounds. The two weekly sports lessons are »skipped«, or the student presents a medical note stating that gymnastics would be dangerous in the child's present condition. One increasingly observes two groups of children: those who are enthusiastic about exercise and those who are exercise-averse. While this development has genetic and social causes, even exercise-averse children should be encouraged to undertake pleasurable exercise from a young age.

A child can never make up for what it has missed in terms of motor development between the ages of 7 and 12. At this age, the brain can learn complex movement

sequences in a way that is not possible in later life. There are no successful athletes and no great musicians that did not acquire the crucial motor skills at this age. But each person has his own level of learning ability. A less gifted child should not let itself be demotivated by the rivalry of more gifted individuals. The doctor can help by finding appropriate types of exercise for acquiring these skills.

**! Watch out for school sports exemptions! These can be a particularly insidious form of poison...**

We are currently seeing a new phenomenon among the young that used not to occur two decades ago: chronic back pain. Nevertheless, we pediatricians have it much easier than those who have to treat adults as back patients. Three factors rarely need to be considered when it comes to young patients: depression, problems in the workplace or a refusal to work and the poorer recuperative powers of

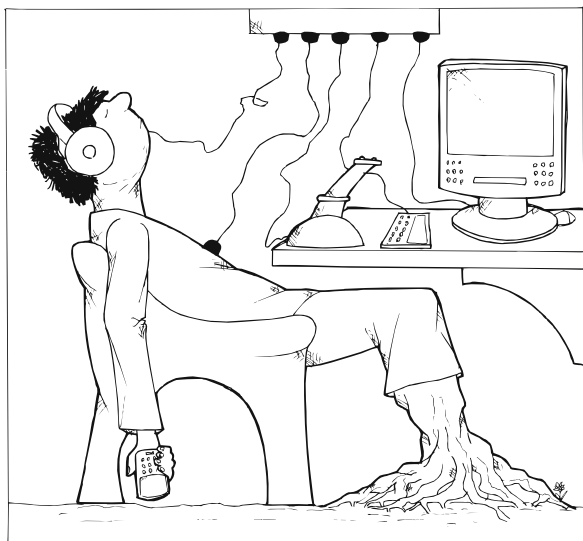
muscles. Because of pension expectations, doctors treating adults often have to struggle (in vain) against the fact that patients – despite their protestations – do not actually want to be healthy.

Young patients with chronic back pain generally *want* to be healthy. It is up to us to persuade them to take responsibility for their own health and do something for themselves, in the form of activity (i.e. cyclical exercise), in order to get rid of the pain. We should help them to practice exercise in a pleasurable way – in this context an appropriate sport is usually better in the long term than years of physiotherapy. How much exercise is needed to remain free of pain depends on the deformity and the associated statics. A decompensated spine requires much stronger muscles than a normally shaped spine. A flat back is also disadvantageous since it can lead to a forward shift in the center of gravity that is difficult to offset. However, sporting patients with such back shapes do not generally suffer pain.

I offer my patients with chronic back pain (in which the pain rather than the deformity is the predominant factor) three options:

1. Live with the pain and find ways of coping with its constant presence (possibly relieve the pain with pain-killers).
2. An operation with an extremely uncertain prognosis in terms of freedom from pain.
3. Train the muscles by intensive daily cyclical exercise.

Almost all patients appreciate that the third option is the best. Some select alternative types of treatment as a fourth option: chiropractic, Rolwing, atlas therapy, shiatsu,



Many children no longer exercise their back muscles regularly because they don't have the time...

acupuncture, osteopathy, etc. These are popular treatments because they can produce immediate freedom from pain without patients having to undertake any significant activity themselves. However, the duration of pain relief is usually short and the treatment must be repeated constantly. The secret of these treatments lies in the fact that the spastic muscle groups are relaxed (which could also be achieved with any form of heat). However, since the muscles are not strengthened and the cause of the tension is not eliminated, the pain recurs at the next (slightest) exertion.

**! Ultimately, therefore, there is no way of avoiding the daily cyclical exercising of the muscles.**

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### 3.1.16 Summary of indications for imaging investigations for the spine

**! The pain »descends« – lumbar pain can be caused by thoracic problems.**

An overview of the indications for spinal imaging procedures is provided in **■** Table 3.13.

### 3.1.17 Indications for physical therapy for back problems

An overview of the indications for physiotherapy for back problems is provided in **■** Table 3.14.

## 3.1.17 · Indications for physical therapy for back problems

■ **Table 3.13.** Overview of the indications for spinal imaging procedures

Symptom?	Where?	Circumstances?	Tentative diagnosis?	When to x-ray?	What is x-rayed?
Local pain	Cervical spine	Acute, without trauma	Torticollis	After 4 weeks	Cervical spine, AP/lateral
		Acute, with trauma	Fracture	Directly	Cervical spine, AP/lateral
		Acute or chronic, without trauma	Tumor, inflammation	Directly	Cervical spine, AP/lateral, poss. bone scan, MRI
	Thoracic spine	Acute, without trauma	Myogelosis	After 4 weeks	Thoracic spine, AP/lateral
		Acute, with trauma	Fracture	Directly	Thoracic spine, AP/lateral
		Acute or chronic, without trauma	Tumor, inflammation	Directly	Thoracic spine, AP/lateral, poss. bone scan, MRI
	Lumbar spine	Acute, without trauma	Myogelosis	After 4 weeks	Thoracic spine + lumbar spine, AP/lateral
		Acute, with trauma	Fracture	Directly	Thoracic spine + lumbar spine, AP/lateral
		Acute or chronic, without trauma	Tumor, inflammation	Directly	Thoracic spine + lumbar spine, AP/lateral, poss. bone scan, MRI
		Chronic, without trauma	Scheuermann's disease	–	Thoracic spine + lumbar spine, AP/lateral
Lumbosacral	Chronic, without trauma	Spondylolysis	Directly	Lumbosacral junction, AP/lateral, poss. bilateral oblique	
Sacrum/coccyx	With or without trauma	Coccygodynia	After 4 weeks	Sacrum and coccyx lateral	
Radiating pain	Legs	Positive neurology	Herniated disk	Directly	Lumbar spine, AP/lateral, poss. MRI or myelogram
Local pain	Thigh	Psoas is spared	Tumor, inflammation	Directly	Lumbar spine, AP/lateral, poss. bone scan, poss. MRI
Deformity	Cervical spine	Oblique position at birth	Congenital (muscular) torticollis	No	–
	Cervical spine	Oblique position without muscle contraction	Klippel-Feil syndrome	Occasionally	Cervical spine, AP/lateral, dens transbuccal
	Thoracic spine	Rib prominence <5°	Thoracic scoliosis	Directly	Thoracic spine + lumbar spine, AP/lateral
	Thoracic spine	Fixed kyphosis	Scheuermann's disease	Directly	Thoracic spine + lumbar spine, AP/lateral
	Lumbar spine	Lumbar prominence <5°	Lumbar scoliosis	Directly	Thoracic spine + lumbar spine, AP/lateral
	Lumbar spine	Tissue anomaly	Spina bifida	–	Lumbar spine, AP/lateral, poss. MRI

■ **Table 3.14.** Overview of indications for physical therapy for back conditions

Disorder	Indication	Goal/type of therapy	Duration	Other measures
Spondylolysis/-olsthesis	If symptoms are present (pain)	Strengthening of back and abdominal muscles (»muscle corset«). Stretching of hamstrings. No lordosing exercises	While symptoms continue	No P.E. exemptions. If the olisthesis progresses or neurological symptoms occur or if the pain fails to respond to treatment, possibly operation. Sport: Not recommended: gymnastics, figure skating, ballet
Thoracic Scheuermann disease	Fixed kyphosis >40°	Straightening, strengthening of paravertebral muscles, stretching of pectoral and hamstrings	Until completion of growth or cure	If kyphosis >50° poss. brace treatment. Operation only poss. if kyphosis >80°. Sport: Not recommended: cycle racing, rowing
Thoracolumbar or lumbar Scheuermann disease	If diagnosed during pubertal growth spurt (regardless of symptoms)	Straightening, strengthening of paravertebral muscles	Until completion of growth or cure	No P.E. exemptions. Sport: Not recommended: cycle racing, rowing. For severe forms, poss. cast brace in ventral suspension. For severe lumbar kyphosis, poss. operation
Scolioses	Fixed scolioses from 15° if growth potential still present	Strengthening of paravertebral muscles, especially on convex side, stretching of muscles on concave side, kyphosing to reduce the lordosis, prevent asymmetry	Until completion of growth	No P.E. exemptions. Sport: Everything permitted, although ballet, gymnastics, figure skating not advisable. From 25° poss. brace treatment. From 40° poss. operation. Continuation of physical therapy important even with brace or surgical treatment
Postural anomalies	None	Motivating patient to take up sporting activity more useful than physiotherapy	–	No P.E. exemptions
Fractures	Only after fracture has healed, if symptomatic (pain, deformity)	Relaxation treatment for muscle spasms (heat); strengthening of paravertebral muscles	While symptoms continue	–

## 3.2 Pelvis, hips and thighs

### 3.2.1 Examination of hips

#### History

- Birth and family history
- Start of walking
- Has trauma occurred?
- Pain history: Where is the pain located? When does it occur? Is it related to loading or movement, or does it also occur at rest (e.g. while sitting) or even at night? If so, does the pain occur only during a change of position, or does the pain cause the patient to wake up at night? Does the pain radiate to the knees or lower legs? Is the pain constant, decreasing or progressive

#### Inspection

##### Examination of the walking patient

Is a *limp* present? What kind of limp is involved: Duchenne/Trendelenburg limp, shortening limp, antalgic limp

We observe the *rotation of the leg* while walking, particularly whether the knees rotate inwardly or are abnormally rotated outwards.

##### Examination of the standing patient

We examine the *pelvic tilt*: see ► chapter 3.1.1 for the measurement of pelvic obliquity. Pelvic rotation is present if, with symmetrically positioned feet, the pelvis is not parallel with both heels. If this is not the case, a rotational deformity is present somewhere. The commonest cause is differing degrees of femoral anteversion (■ Fig. 3.127).

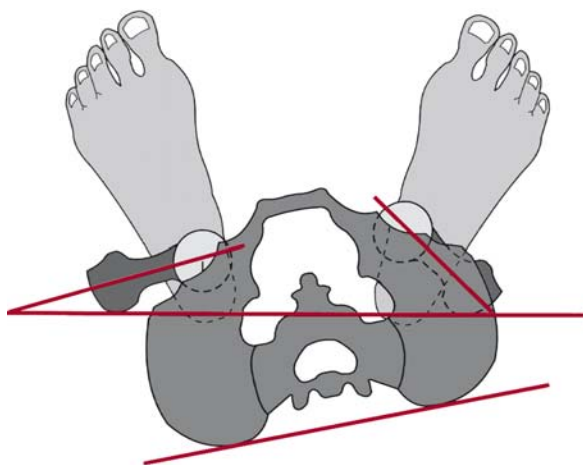
##### Investigation of the Duchenne and Trendelenburg signs

During single-leg stance the pelvis is slightly raised on the side of the free leg under normal circumstances

whereas, in cases of insufficiency of the gluteus medius and minimus muscles, the pelvis drops on the side of the free leg. The patient is able to compensate for a slight insufficiency by shifting the upper body towards the stance leg (Duchenne sign, grade I). If the insufficiency is more severe, however, the pelvis drops on the side of the free leg (Trendelenburg sign, grade II). If the muscle decompensation is even more pronounced, the free leg has to be supported against the stance leg in order to maintain the single-leg stance (grade III). In a grade IV insufficiency, the patient cannot even maintain a single-leg stance without holding onto some kind of support (■ Fig. 3.128).

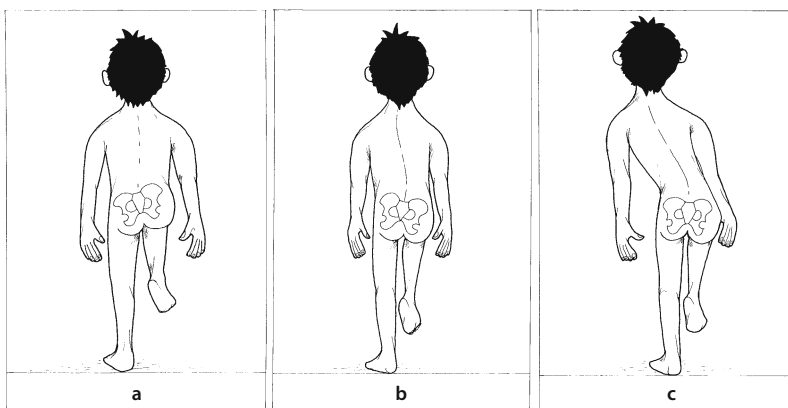
#### Palpation

Palpation primarily serves to establish any *tenderness*. Typical pain points include: the greater trochanter, the soft tissues beneath the inguinal ligament (iliopectineal



■ Fig. 3.127. Pelvic torsion due to differing degrees of anteversion (from above): The pelvis is rotated when the feet are parallel because of the differing degrees of anteversion of the femoral necks. Pelvic rotation can simulate pelvic obliquity

■ Fig. 3.128. Investigation of the Duchenne and Trendelenburg signs during single-leg stance. **a** Normal: The pelvis is slightly raised on the side of the free leg. **b** In cases of insufficiency of the abductors, the pelvis drops on the side of the free leg (Trendelenburg sign). **c** The abductor insufficiency can be compensated to some extent by shifting the body's center of gravity towards the stance leg (Duchenne sign)



### 3.2.1 · Examination of hips

bursitis), the psoas muscle, the attachment of the adductors and the gluteus medius and minimus muscles above the greater trochanter. Pain originating from the joint itself is usually diffuse and cannot be elicited by palpation. An effusion is equally difficult to palpate.

#### Range of motion

❗ **Both sides must always be measured when investigating hip mobility.**

##### — Flexion/extension:

Only flexion can be measured if the patient is in a supine position (■ Fig. 3.129). If we wish to measure the precise degree of extension the patient must be examined in the lateral position (■ Fig. 3.130). In most cases, however, the main objective is to detect the presence of any flexion contracture. Since the lack of extension at the hip can be compensated by hyperlordosis of the lumbar spine, this lordosing option must be excluded. The Thomas grip is a suitable maneuver

for this purpose (■ Fig. 3.131). Flexion and extension should always be tested in respect of rotation in the neutral position. If the leg is forced into a gradually increasing external rotation with increasing hip flexion, this is termed the »Drehmann sign«. This typically occurs after slipped capital femoral epiphysis, but is also observed in other hip disorders.

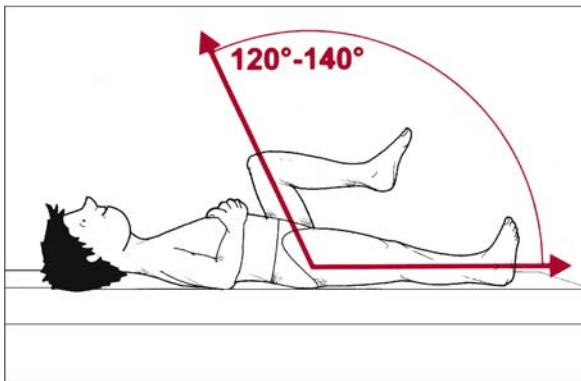
##### — Abduction/adduction:

This is normally measured in the supine position with the hip extended. For the correct measurement of adduction, the opposing leg must be raised (■ Fig. 3.132).

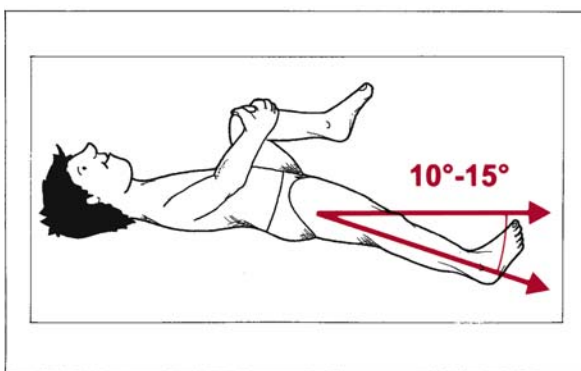
##### — External/internal rotation:

This measurement can be performed with the hips in 90° flexion and in the extended position.

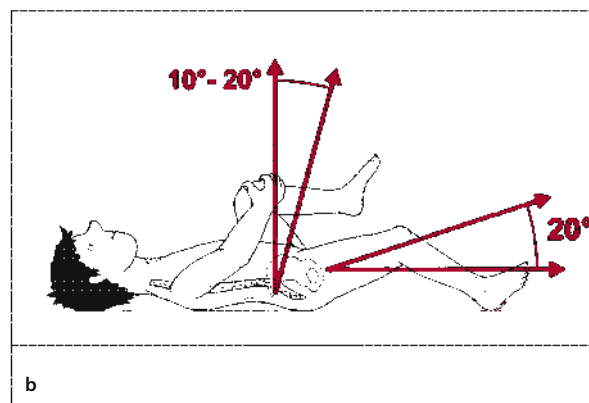
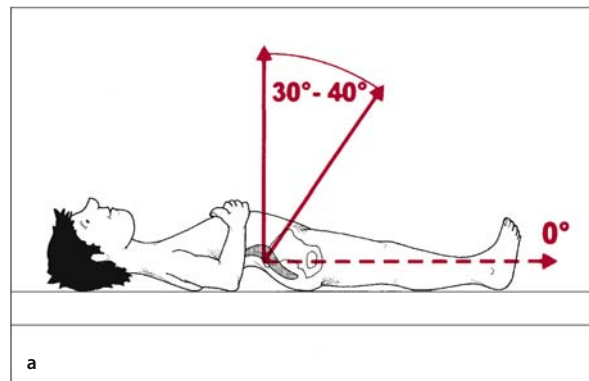
— *Measurement in 90° flexion:* This measurement can be performed with the patient lying in the supine position on the examination table (■ Fig. 3.133). The extent of external and internal rotation should be roughly equal in each case. If internal rotation greatly predominates, an anteverted hip is present.



■ Fig. 3.129. Measurement of hip flexion in the supine position



■ Fig. 3.130. Measurement of extension in the lateral position: The pelvis is fixed by flexing the hip on the other side. This prevents any hyperlordosis of the lumbar spine. The extent of extension in relation to the neutral position is measured



■ Fig. 3.131a, b. Thomas grip: a In the supine position, full extension of the hip can be simulated by hyperlordosis of the lumbar spine and increased pelvic tilting. b The patient holds the contralateral leg with maximum flexion at the hip. If a flexion contracture is present the affected knee is raised

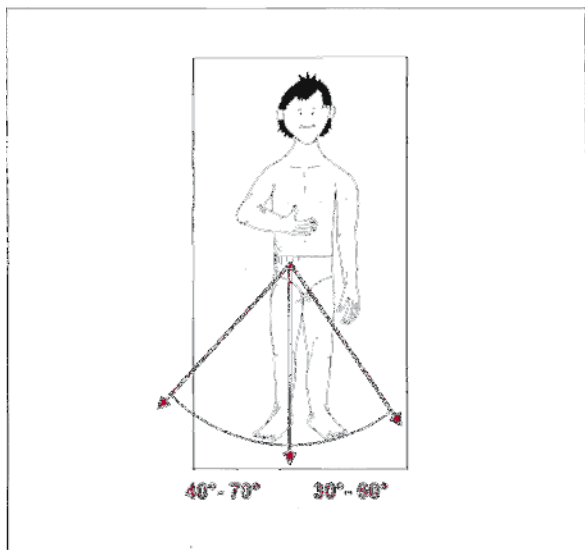


- *Investigation of impingement:* If the hip is adducted and internally rotated while in 90° flexion, a typical pain is provoked if impingement exists between the femoral neck and the acetabulum.
- *Measurement with the hip extended:* This is the more important investigation and can be performed either with the patient in the supine position at the end of the examination table (in which case the lower legs hang down at right angles over the end of the table) or in the prone position (■ Fig. 3.134). With the hips

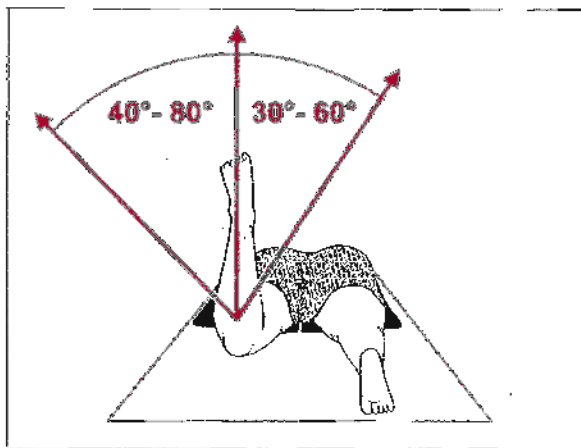
extended, the extent of external rotation is usually less than with the hips flexed.

**Femoral anteversion**

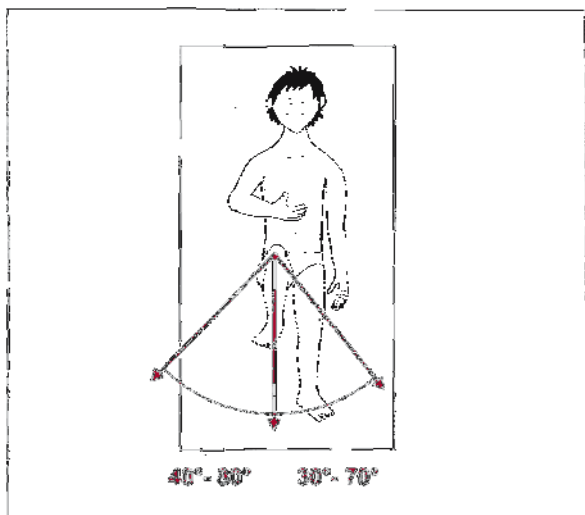
*Anteversion* is also clinically examined in the prone position (■ Fig. 3.135) [2]. The examiner palpates the greater trochanter with one hand and then rotates the lower leg outwards until the maximum lateralization of the greater trochanter is felt. In this position, the angle formed by the lower leg with the vertical is measured. This corresponds



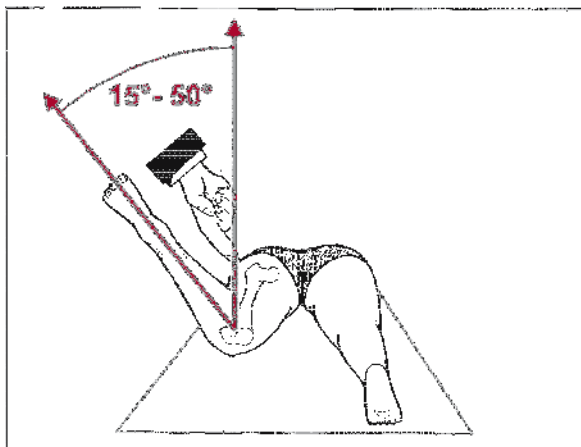
■ Fig. 3.132. Investigation of ab- and adduction in the supine position: The opposite leg must be raised for the adduction measurement



■ Fig. 3.134. Investigation of external and internal rotation in the prone position with extended hips: The lower leg is used as a pointer in this test in the prone position



■ Fig. 3.133. Investigation of external and internal rotation in 90° flexion of the hips in the supine position: The lower leg is used as a pointer. Note that external swiveling of the lower leg produces internal rotation at the hip



■ Fig. 3.135. Clinical measurement of anteversion: Anteversion is examined in the prone position. The examiner palpates the greater trochanter with one hand to detect the position of maximum lateral protrusion. The other hand moves the lower leg (knee in flexion). When the greater trochanter is in maximum lateralization, the anteversion can be measured directly by the deviation from the vertical. In this position, the femoral neck is horizontal, while the knee condyles and lower leg show the anteversion angle

## 3.2.1 · Examination of hips

to the degree of anteversion. In experienced hands, the accuracy of this measurement is just as good as x-ray measurement or measurement by ultrasound [2].

**Examination protocol for the hips**

Our examination protocol for the hips is shown in

■ Table 3.15.

**References**

1. Debrunner HU (1982) Orthopädisches Diagnostikum. Thieme, Stuttgart New York, S 102–105
2. Ruwe PA, Gage JR, Ozonoff MB, De Luca PA (1992) Clinical determination of femoral anteversion. A comparison with established techniques. J Bone Joint Surg (Am) 74: 820–30

■ Table 3.15. Examination protocol for the hips

Examination position	Examination	Questions
<b>I. History</b>	Birth, family history? Pain (hip/groin/knee)? How long? Load-related? Radiating? Increasing or decreasing? Trauma?	Hip dysplasia or other hip disorder? Fracture, dislocation?
<b>II. Walking</b>	Limp? – Duchenne/Trendelenburg – Shortening limp – Stiff limp – Paralytic limp – Antalgic limp  Rotational position of the leg (kneeing-in, kneeling-out?)	Insufficiency of the gluteal muscles? Leg length discrepancy of >2 cm? Restricted hip range of motion? Neurological problems? Anteverted hip?  Rotational deformity of the lower leg?
<b>III. Standing</b>	Atrophy/asymmetry Pelvic obliquity Pelvic rotation Trendelenburg sign	Contractures, neurological problems? Leg length discrepancy? Differing degrees of anteversion? Insufficiency of the gluteal muscles?
<b>IV. Sitting</b>	Sitting with legs straight Cross-legged, cross-legged with alternative leg position	Contracture of the hamstrings? Increased anteversion?
<b>V. Lying (infant)</b>	Buttock folds, Ortolani sign, leg lengths, inhibition of abduction, instability	Hip dysplasia or dislocation?
<b>VI. Lying (older child)</b>		
<b>Supine</b>	Flexion/extension Internal/external rotation Ab-/adduction with extended hip Drehmann sign Forced adduction and internal rotation at approx. 90° flexion	Restricted movement? Pain (particularly during rotation)? Slipped capital femoral epiphysis?  Examination for impingement
<b>Prone</b>	Extension Rotation with extended hip Estimate anteversion angle	Full extension possible? Restricted rotation? Pain? Anteverted hip?
<b>Lateral</b>	Extension	Full extension possible?

### 3.2.2 Radiographic techniques

The following standard x-rays of the hip are recorded:

- AP overview of the pelvis and both hips with lower legs suspended:

The patient lies in the supine position at the end of the table with both lower legs hanging freely over the edge of the table. The lower legs are parallel and the patella points forwards, the buttocks lie directly over the grid cassette. The central beam is 5 cm above the symphysis in the mid-line (■ Fig. 3.136).

- AP view of the pelvis and hips in the infant with parallel lower legs and slightly flexed thighs:

Although diagnostic imaging techniques have been superseded by ultrasound in the infant hip, this view is still indicated occasionally, e.g. as a final check after treatment for a hip dysplasia, when child abuse is suspected or in septic arthritis with destructive bone changes in the vicinity of the hip.

- Pelvic overview in the infant according to Andren/von Rosen, both hips held in 45° abduction and maximum internal rotation:

This view is likewise required nowadays only in rare instances, for example if ultrasound is not available and hip dysplasia in young infants is strongly suspected, particularly also if hip dislocation is suspected.

- Lateral view of hips with vertical x-ray beam (according to Lauenstein; frog view):

The patient lies in the supine position with the relevant hip in 45° flexion and 45° abduction. If abduction is inhibited by pain, the other leg will need to be elevated. A suitable alternative to the Lauenstein view is a lateral x-ray of both hips in the »frog position« with a vertical beam, e.g. in cases of suspected Legg-Calvé-Perthes disease or other lesions of the proximal femoral epiphysis.

- Anteversion view of both hips according to Dunn and Rippstein:

Patient in supine position, both hips and knees flexed at 90°, both hips abducted by 20° (■ Fig. 3.137).

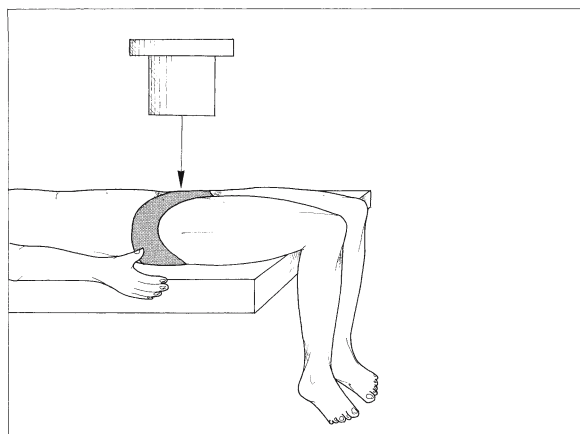
- Oblique view of the hip in the standing position (»faux-profil view« according to Lequesne):

For this x-ray the patient stands obliquely in front of the x-ray stand so that the angle between the back and the stand plane is 65°. If the right hip is being x-rayed, the right foot is parallel to the film plane so as to obtain a lateral view of the upper end of the femur. In case of doubt, the forward-pointing patella is selected as a reference point (■ Fig. 3.138).

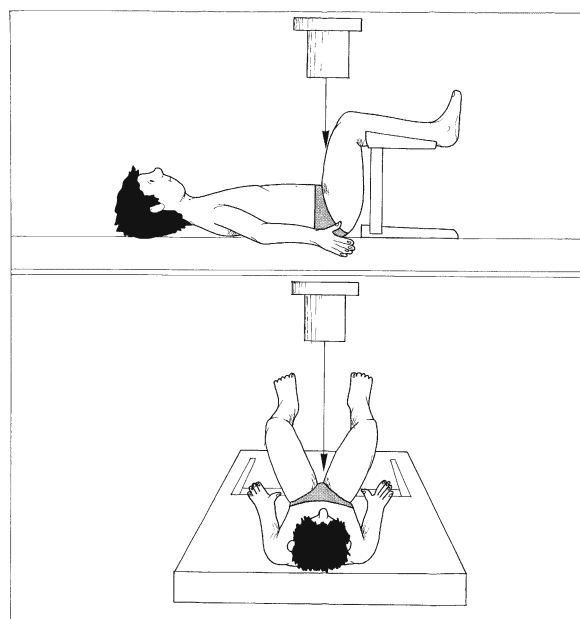
A gonad shield is not used for this view, but must be used for all other x-rays.

- Ultrasound of the hip:

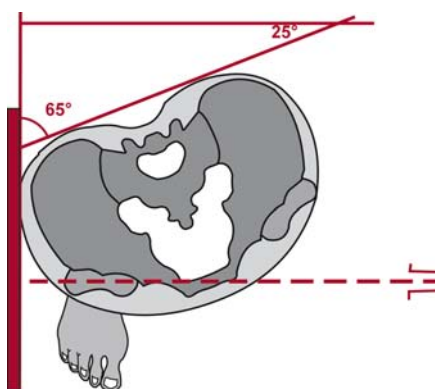
The ultrasound examination of the infant hip is described in ► chapter 3.2.4. The sonogram can also be useful in cases of hip joint effusion (■ Fig. 3.240b) or



■ Fig. 3.136. Recording the AP view of the pelvis with hanging lower legs



■ Fig. 3.137. Recording the anteversion view according to Dunn



■ Fig. 3.138. Position for the »faux profil view« according to Lequesne

### 3.2.3 · Biomechanics of the hip

in Legg-Calvé-Perthes disease for the evaluation of lateral calcification (■ Fig. 3.188).

#### — Arthrography of the hip:

The conventional arthrogram may be indicated in hip dislocation (■ Fig. 3.157), Legg-Calvé-Perthes disease (■ Fig. 3.193b) or a proximal femoral focal deficiency (■ Fig. 3.205).

#### — Computed tomogram (CT) of the hip:

A CT scan is primarily indicated if the mechanical situation needs to be assessed, for which the three-dimensional reconstruction is often helpful (example in ■ Fig. 3.151), or for evaluating intraosseous lesions (examples in ■ Fig. 3.210b).

#### — MRI and arthro-MRI of the hip:

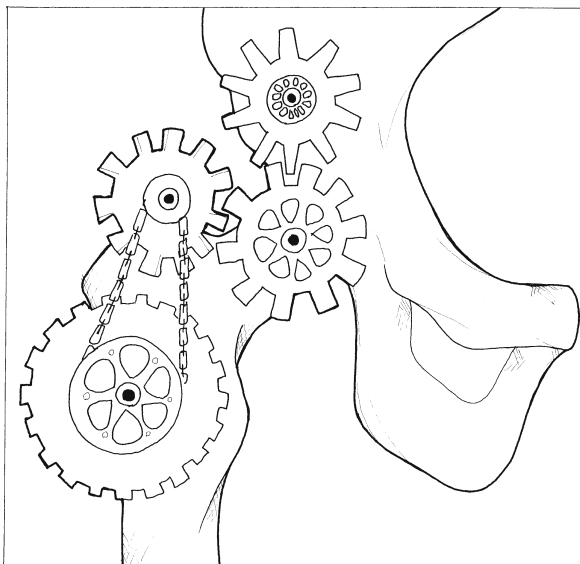
MRI is useful for identifying tumors (example in ■ Fig. 3.247). An arthro-MRI scan with intra-articular administration of contrast medium is needed to assess the intra-articular situation and the labrum (example in ■ Fig. 3.150).

### 3.2.3 Biomechanics of the hip

» *Without research, and particularly without basic research, there can be no progress. The great strides forward in orthopaedic surgery over the course of the last three decades are primarily attributable to the efforts and successes of biomechanical research [16].* «

#### ► Definition

Biomechanics is concerned with the reactions of the living body to physical forces. It involves the study of a biological system using the resources of mechanics. Biomechanics is an interdisciplinary field between medicine and engineering.



### Historical background

Preliminary biomechanical observations were made by Fick back in 1850 [5]. Braune and Fischer presented ideas about the center of gravity of the human body in 1889 [2]. Fischer was also the first to investigate the human gait scientifically. In 1899 he published a precise analysis of a soldier's walk [6]. In 1935, Pauwels published major studies on the mechanical loading and stressing of the hip 1935 [20]. Other important findings were made by Amtmann [1], Kummer 1968 [13], Legal from 1977–1980 [18] and by Tönnis in 1976 [25].

### Anatomical and radiological principles

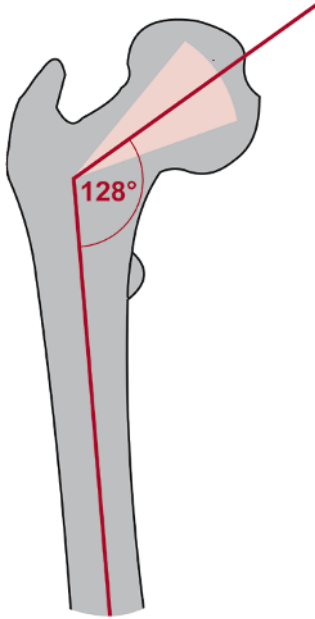
The hip is a modified ball joint with 3 main axes at right angles to each other and thus 3 degrees of freedom of movement. The femoral head is largely spherical, while the acetabulum is horseshoe-shaped with a central depression and a caudal recess. The femoral head tapers where it joins the femoral neck in the section of bone not covered by cartilage. The direction of the femoral neck is characterized by two factors: its inclination and anteversion. The femoral neck inclination (or femoral neck-shaft angle) is also known as the CCD angle (Center-Collum-Diaphysis angle) (■ Fig. 3.139). This decreases during human development from birth to adulthood from 150° to approx. 130° [14] (■ Fig. 3.140). Anteversion refers to a forwardly projected open spatial angle formed between the femoral neck and the frontal plane or the plane of the knee condyles (■ Fig. 3.141). This angle also declines during life from approx. 30° at birth to around 15° in adulthood (■ Fig. 3.142).

Anteversion cannot be measured directly. The axial x-ray only provides a projected anteversion angle. We use the technique suggested by Dunn (► Chapter 3.2.2). The actual angle is calculated on the basis of the figures for the projected neck-shaft angle and anteversion angles using a conversion table. While not a highly accurate method, it is just as accurate as measurement by ultrasound [4] (► Chapter 4.2.1) or clinical measurement (► Chapter 3.2.1) [23].

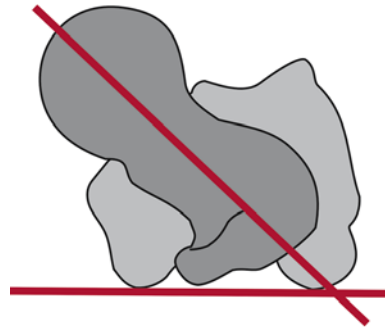
The bone structure of the proximal end of the femur is characterized by typical compression and tension trajectories formed, according to Wolff's law of transformation, by bending stresses acting on the femur at the hip (■ Fig. 3.143). The alignment of these trajectories depends on the loading and the shape of the proximal end of the femur. The bone structure at the acetabulum is also influenced by shape and loading. The smaller the loaded area and the greater the overall load, the more likely it is that a sclerotic section will form.

As regards the acetabulum, the acetabular roof angle (AC angle) is crucially important in the evaluation of the infant hip. The measurement of this angle and its values in normal and pathological hips are described in detail in ► chapter 3.2.4. In the evaluation of the adolescent or adult hip, the Center-Edge angle (CE angle; ■ Fig. 3.149) proposed by Wiberg is the most important factor these days [29]. This angle is a numerical value reflecting the

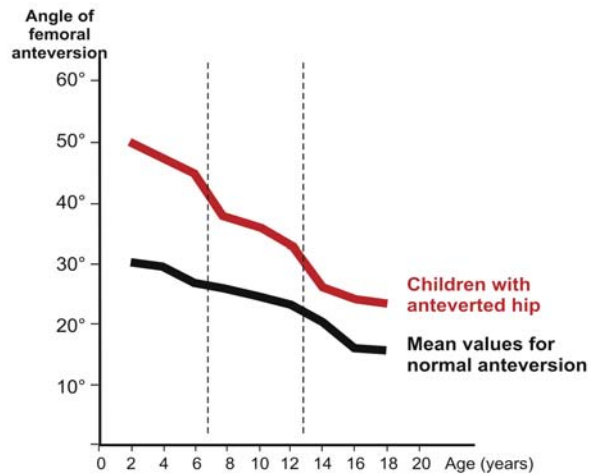
extent of the support for the femoral head provided by the acetabular roof. Wiberg observed an average value of  $26^\circ$  for adults, with a range of variation of  $20^\circ$  to  $46^\circ$ . An angle of  $20^\circ$  is currently cited as the borderline between normal and abnormal in adults [26]. However, the Wiberg angle should be viewed in conjunction with the head radius. With large radii, smaller CE angles are more acceptable than with a small head diameter [15].



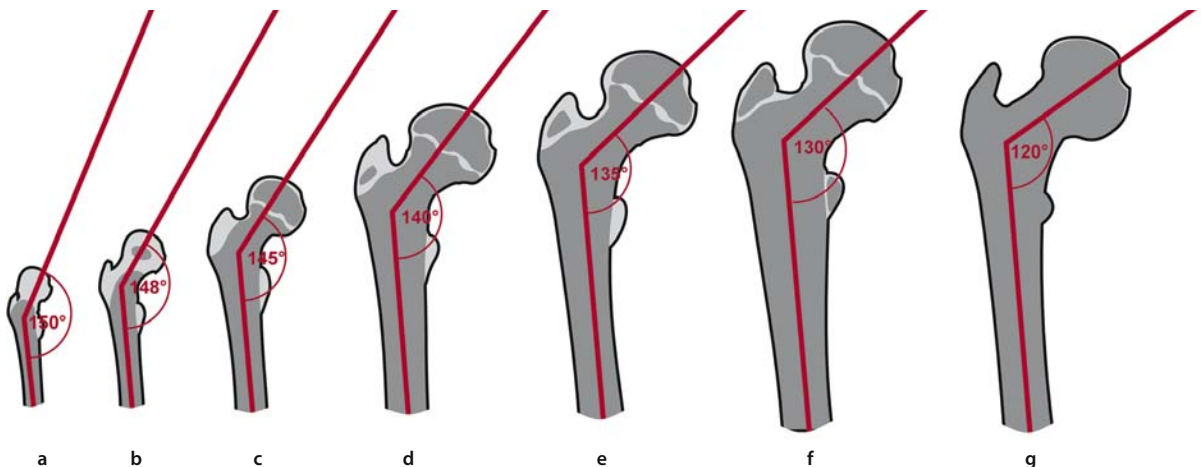
**Fig. 3.139.** Measurement of the neck-shaft angle: The center of the femoral head is located with the aid of a circular template. The center of the femoral neck is also determined at its narrowest point. The two points are joined, and the resulting straight line forms the femoral neck axis. The angle formed by this line with the shaft axis constitutes the neck-shaft angle (the colored zone shows the normal range)



**Fig. 3.141.** Presentation of the anatomical anteversion angle: Anteversion refers to the spatial angle formed between the horizontal plane and the plane of the femoral neck. The knee condyles and femoral neck are projected in an overlapping position



**Fig. 3.142.** Development of femoral anteversion angle. Mean values for normal anteversion (black curve) and children with an anteverted hip (red curve). Following two derotation phases (dotted lines), anteversion reaches its normal adult angle by the completion of growth. (After [10])



**Fig. 3.140a–g.** Development of the neck-shaft angle: a at birth, b at the age of 1 year, c 3 years, d 5 years, e 9 years, f 15 years, g in adulthood. (After [14])





■ Fig. 3.143. The *compression and tension trajectories* at the proximal end of the femur can be seen on any AP x-ray of the hip

Anteversion of the acetabulum plays an important biomechanical role. It refers to the angle formed by the acetabular opening and the horizontal plane. Note that the plane of the acetabular opening does not correspond to a closed ring, but incorporates an anterolateral recess to allow for flexion of the femur. If the anterior and posterior contours overlap on the AP x-ray (»cross-over sign«) and the posterior rim of the acetabulum is medial to the center of the femoral head in a well-centered hip (»posterior wall sign«), acetabular retroversion is suspected [24] (■ Fig. 3.150).

Apart from the shape of the femoral head, its position in relation to the femoral neck crucially affects the play between the femur and acetabulum. The head must be centered over the neck so that it projects beyond the neck anteriorly. This is known as the offset of the head [9, 19, 21].

### Calculation of loading

In a double-leg stance, only external forces act on the hip via the weight of the body. The pelvis rests on both femoral heads. No muscle forces are required in the frontal plane. The situation is different for a single-leg stance or during the stance phase while walking. In the latter

case, the hip of the stance leg bears the weight not only of the head, trunk and arms but also of the swing leg. Because of this one-sided supporting of the pelvis by the hip of the stance leg, muscle forces must act to prevent the dropping of the pelvis on the swing leg side, thereby substantially increasing the joint forces compared to the double-leg stance scenario. The maintenance of balance requires a lever system with a pivot point located in the center of the head. The acting forces are greatly dependent on the anatomical circumstances. Initial calculations with a two-dimensional model were first made by Pauwels [20]. In a normal hip, the force resultant acting on the hip is approximately four times the body weight. If the femoral neck angle is greater than normal, and the lever arm of the abductors correspondingly shorter, the force resultant increases. The reverse situation applies with coxa vara, in which the femoral neck angle is smaller, thus lengthening the lever arm of the abductors (■ Fig. 3.144).

However, Pauwels' calculations are based on a two-dimensional model and can only provide rough approximations. By no means should one conclude that an increased femoral neck-shaft angle necessarily results in overloading of the hip. Firstly, the valgus position of the proximal end of the femur does not exist in reality, but only produces a coxa valga in projection if the anteversion angle is elevated. Secondly, the loading of the hip cannot be determined from just *one* single parameter since a wide variety of factors are involved. The varus osteotomy that often used to be implemented on the basis of theoretical considerations resulted in the recurrence of a valgus position during subsequent growth, as this was required by the anatomical circumstances in a given hip.

Recent investigations with cadaver bones have shown that the trochanteric apophysis is primarily loaded in compression, and that the force resultant is approx. 1.7 times the body weight. This stimulates cranio-lateral growth and therefore also influences the development of the neck-shaft angle [8].

### Loading – three-dimensional analysis

A three-dimensional view of the anatomical situation is needed to calculate the *loading* of the hip joint. Only if the loaded area is known can the pressure distribution and loading be determined, and this is usually possible only with complex mathematical calculations [3, 8, 13, 15, 22, 26]. The author has developed a relatively simple method for determining the contact area between the acetabulum and femoral head [7], subject to the requirement that the femoral head and acetabulum are roughly spherical and that the bony parts of the hip are largely fully developed. The method can be applied to girls from a skeletal age of 10 years and, correspondingly, to boys from 12 years of age. The method involves the placement of a template

over normal AP x-rays (good quality) of the hip on which the anterior and posterior rims of the acetabulum must be visible. The circular template is marked with projected spherical areas that are all equal in size and each representing 0.5% of the total surface area of the sphere. By counting the rectangles and triangles located under the anterior or posterior rim of the acetabulum, the percentage of the covered area in relation to the total surface area of the sphere can be determined. Finally, the value

in square centimeters can be calculated on the basis of the radius.

Fig. 3.145 shows the geometrical design of the template, which always presents equally sized areas in 3D projection. The various sizes of the template are shown in Fig. 3.146. This pattern can be copied onto a sheet of transparent film. The sheet with the template of the appropriate size is placed over the hip x-ray (Fig. 3.147). The percentage and area can be determined very simply

Fig. 3.144a–c. Schematic view of the forces in the hip according to Pauwels [20] a in the normal hip, b valgus hip and c varus hip. The diagram shows the effect produced by a change in the lever arms on the acting forces (*G* Center of gravity, *W* Body weight, *R* Force resultant in the hip, *M* Forces of the abductors)

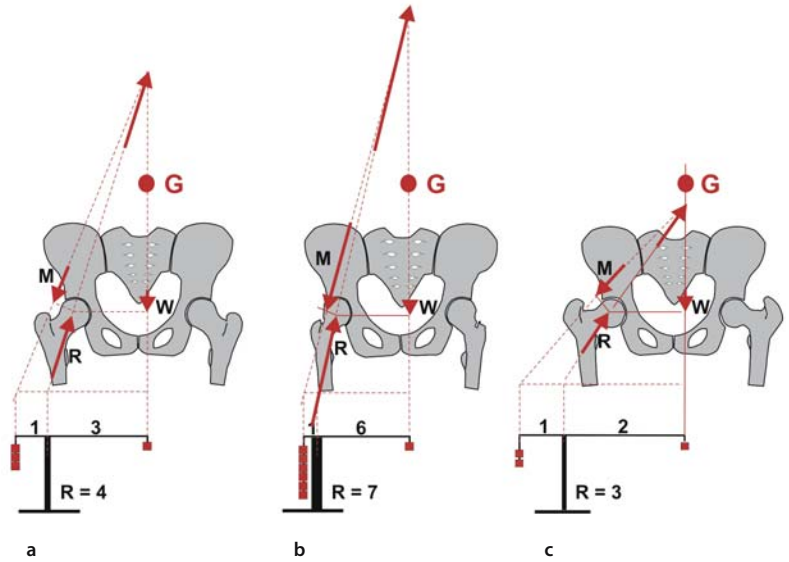
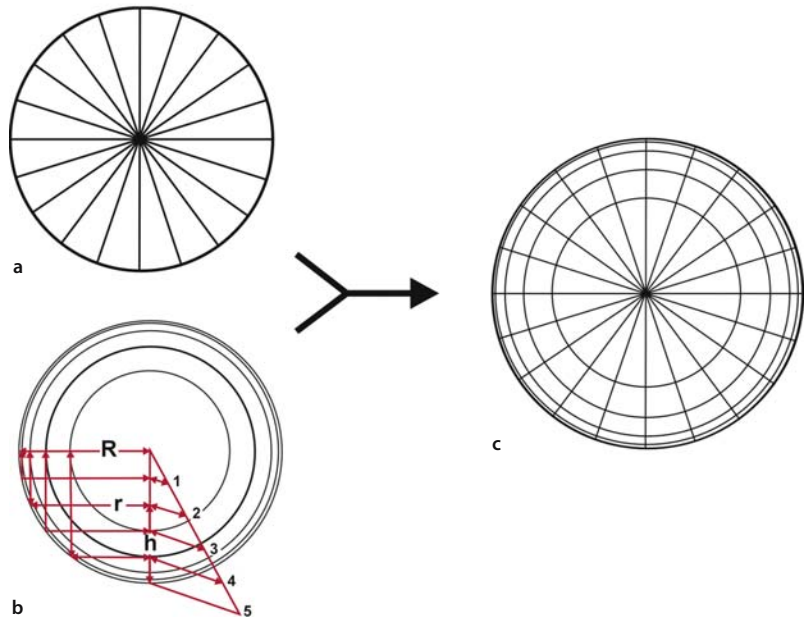
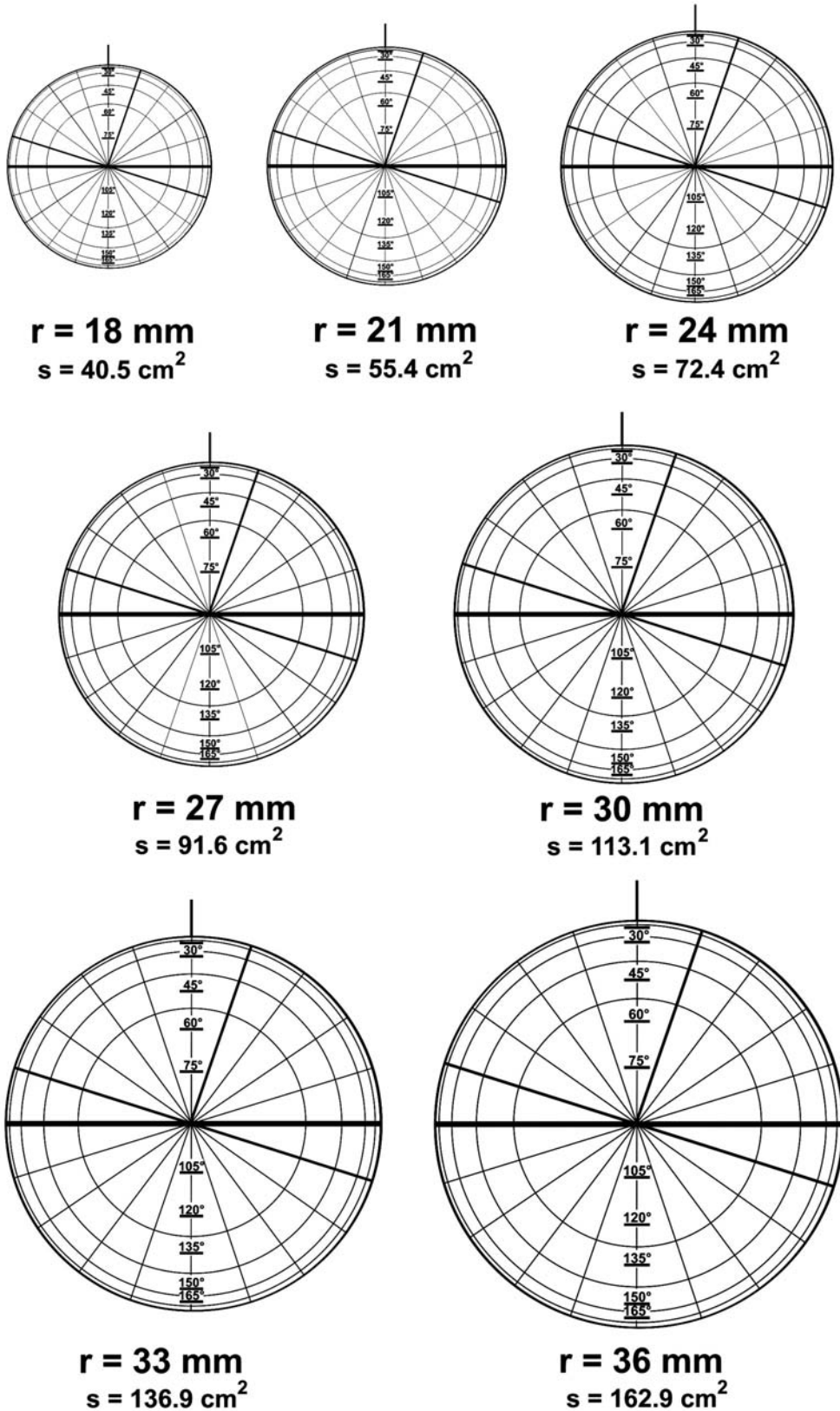


Fig. 3.145. a Subdivision of a sphere into 20 equally sized sectors each with an angle of 18°. b Geometrical calculation of the segment boundaries for the template, allowing the contact area to be determined between the acetabulum and femoral head. In projection, each segment covers the same spherical surface area. c Overlapping the sector and segment boundaries produces a total of 100 triangles and rectangles on a hemisphere (R Radius of the overall sphere, h Height of the segment boundaries, r Radius of the segment boundaries)





■ Fig. 3.146. Templates in various sizes. These can be copied onto transparent film, and the template of the appropriate size is placed over the x-ray. The percentage in relation to the total surface area of the sphere (lower figure) can be calculated by counting the segments

(each corresponding to 0.5% of the surface of the sphere) on the anterior and posterior hemisphere. The  $\varphi$  angles between the center of the femoral head and the anterior and posterior acetabular rims can also be entered on the templates (also Fig. 3.149)

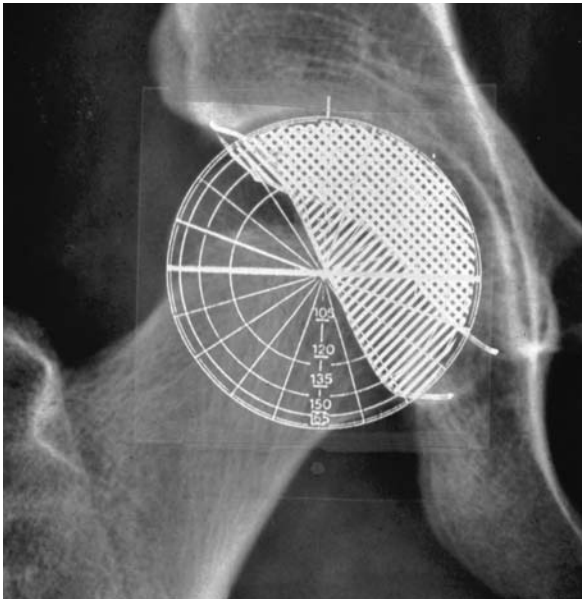
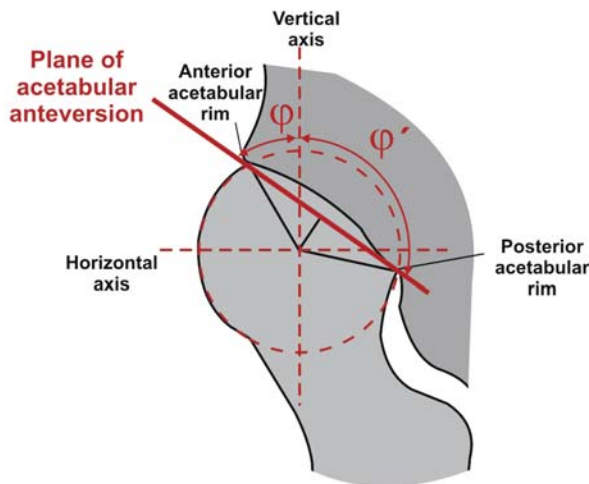


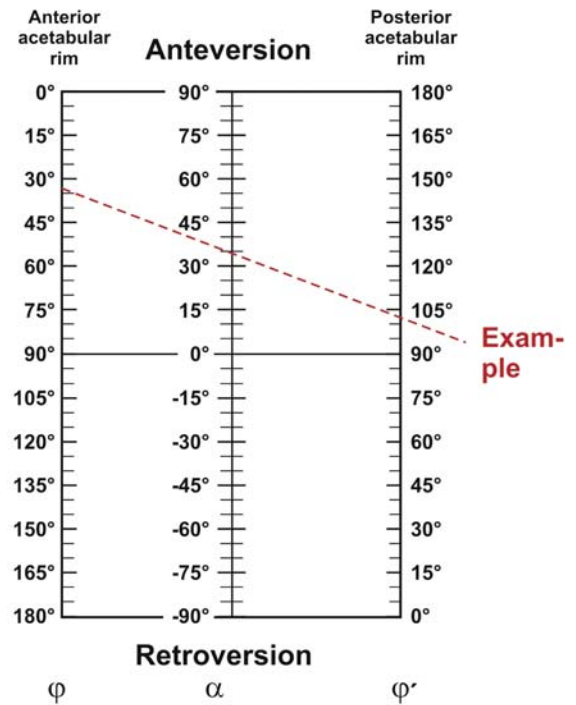
Fig. 3.147. Example of a contact area calculation using a template placed on an AP x-ray of the hip. The sections bounded by the anterior and posterior rims of the acetabulum are counted and converted into the percentage of the total surface area of the sphere

by counting the rectangles and triangles within the anterior and posterior rims of the acetabulum. A method based on the same principle but employing more sophisticated computer calculation was recently described [12]. The figures marked on the template also allow an estimate to be made of the angles between the center of the femoral head and the anterior and posterior acetabular rims. These two angles can be used to calculate the degree of acetabular anteversion/retroversion (Fig. 3.148). The two angles for the anterior and posterior sides are read off the template and then marked on the x-ray. The acetabular orientation in both the sagittal and anatomical planes can be determined by drawing a line between the two marks entered for the angles on the ventral and dorsal sides [18]. Anteversion remains very constant during the development phase [28].

The template can also be used to calculate the relevant loading of the hip [15]. Fig. 3.149 shows an auxiliary construction proposed by Legal for determining the relevant loading of the hip. Since the force resultant R generally forms an angle of 17° from the vertical, the nearest sector boundary to the vertical on the template can be used as an approximation, since the angle between the



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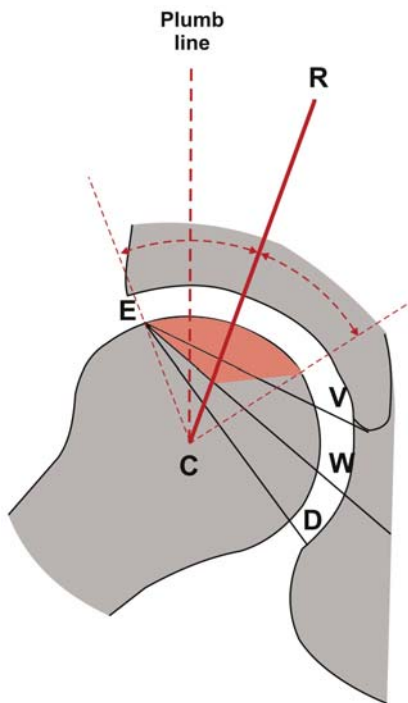


b

Fig. 3.148. Acetabular anteversion. a Drawing of a hip from the side. Angles between the center of the femoral head and the anterior ( $\varphi$ ) and posterior ( $\varphi'$ ) acetabular rims. The plane between these two points corresponds to the acetabular orientation or anteversion. The

angles of  $\varphi$  and  $\varphi'$  are determined on the AP x-ray using the template. b The nomogram can be used to determine the acetabular orientation (anteversion/retroversion) by drawing a line between these two scales for the angles  $\varphi$  and  $\varphi'$





■ **Fig. 3.149.** Auxiliary construction proposed by Legal [15] for determining the *loading of the hip*. The line *EW* bisects the angle between the lines *EV* and *ED*. The point at which this line crosses the force resultant *R* (at an angle of approx.  $18^\circ$  to the vertical) corresponds to the anterior boundary of the loading area. This extends symmetrically around the force resultant up to the lateral acetabular epiphysis (*E*) and up to the determined anterior boundary. This bounded area can be calculated very simply using the template (C Head center). The angle between the vertical and the line *CE* is termed the *Center-Edge angle (CE angle)* according to *Wiberg* [29]

sectors on the template is  $18^\circ$ . This load-relevant area can be calculated very simply by counting the rectangles and triangles in the hatched area of the sector.

### Joint play

The freedom of movement between the femoral head and acetabulum plays a – formerly underestimated – role in normal joint function. It was R. Ganz (Berne) who first highlighted the problem of impingement between the femoral neck and acetabulum, a problem that has been studied over the past 10 years. Various anatomical and biomechanical studies have identified the factors which, instead of a smooth sliding movement between the femoral head and acetabulum, cause the femoral neck to strike the acetabulum, in turn triggering a shear movement of the head in the joint [9, 19, 21, 24]. The cause of this »impingement« may lie in the acetabulum, the femur or both components together.

As regards acetabular causes, reduced anteversion (■ Fig. 3.150), a diminished anterolateral recess, an exces-



■ **Fig. 3.150a, b.** 19-year old female patient with bilateral *retroversion of the acetabulum*. **a** AP x-ray of the pelvis. Note the cross-over sign and the posterior wall sign on both sides (the corresponding contours are drawn in on the right hip: *solid line* anterior acetabular rim, *dotted line* posterior acetabular rim, *dot* head center). **b** *Arthro-MRI* of the same patient. Labral lesions are clearly visible on both sides

sively large and deep acetabulum can lead to impingement (known as a »pincer effect«). Such abnormal shapes can occur idiopathically, but are more commonly seen after incorrectly performed reorientation operations. They can result in injury to the labrum (■ Fig. 3.150b). A projecting anterior inferior iliac spine that is positioned too low (e.g. after an apophyseal avulsion) can also cause impingement (■ Fig. 3.151).

As regards femoral causes, the incorrect position of the femoral head with a reduced offset is primarily responsible for the femoral neck striking the acetabular rim, as typically occurs in slipped capital femoral epiphysis (■ Fig. 3.201a). On the one hand, this damages the acetabular labrum (known as a »cam effect«) and, on the other, produces a shear movement of the head within the joint. The shear movement mainly occurs during flexion, but can even be present during normal walking if the shape is severely abnormal [21]. The impingement can be reduced by external rotation of the leg during walking.





**Fig. 3.151.** CT with three-dimensional reconstruction in a 15-year old female athletic patient with *apophyseal avulsion* and excessively low growth of the anterior inferior iliac spine (arrow), resulting in impingement with the femoral neck during flexion

### Effects of incorrectly shaped bony components

The crucial question in every case is whether an incorrectly shaped component can lead to *premature osteoarthritis*. The answer is *clearly in the affirmative* if the following anatomical changes are present:

1. off-center femoral head [17],
2. excessively small acetabulum,
3. excessively steep acetabulum,
4. inadequate acetabular coverage laterally and anteriorly [17],
5. non-round acetabulum,
6. aspherical femoral head,
7. reduced anteversion of the acetabulum,
8. excessively large and deep acetabulum (acetabular protrusion),
9. reduced offset of the femoral head [9, 19, 24].

A *pre-arthritis condition* probably also exists in cases of:

10. shortening of the femoral neck,
11. retroverted hip [24, 27].

We would expect a *functional restriction without any risk of premature osteoarthritis* in the case of an:

12. elevated trochanter.

According to the latest findings, the following conditions do *not constitute pre-arthritis*:

13. anteverted hip,
14. coxa magna.

For biomechanical, rather than inflammation-related, reasons, the osteoarthritis can develop as a result of an excessively small loading area, a load transfer surface that is too steep and/or irregular, or the development of shear forces during impingement. The excessively small loading area is a factor in the above-listed situations 1, 2, 4, 5 and 6. We encounter the adverse load transfer orientation in situations 1, 3, 5 and 6. Impingement occurs in conditions 7–10. In many cases, the resulting shear forces cause arthroses that used to be described as »idiopathic«.

In a *triple osteotomy*, all three bones (ilium, pubis and ischium) are divided, while the cut in a *periacetabular osteotomy* goes around the acetabulum (and thus through the triadate cartilage, as well). The effect is similar in both osteotomies. The acetabulum is not actually enlarged but is rather rotated laterally and – if necessary – anteriorly, thereby enlarging the relevant loading area at the cost of the caudal sections. This operation is particularly suitable if the bony components are roughly spherical but inadequate lateral acetabular coverage exists. In this case the anterior coverage is improved at the expense of the posterior coverage.

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### 3.2.4 · Developmental dysplasia and congenital dislocation of the hip

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### 3.2.4 Developmental dysplasia and congenital dislocation of the hip

» *The discovery of bloodless hip reduction was an exquisite solution, in its classical simplicity so very much a product of the simple genius of Adolf Lorenz, a man whose mind was not befuddled with excessive book-learning and theory* «  
*(Albert Lorenz writing about his father Adolf Lorenz in the highly readable and amusing biography »Wenn der Vater mit dem Sohne...«).*

#### ► Definition

- *Developmental dysplasia of the hip (DDH):* Inadequate development of the hip with impaired ossification of the lateral acetabular epiphysis
- *Congenital dislocation of the hip (CDH):* Displacement of the femoral head from its central position in the acetabulum

#### Historical background

Even Hippocrates (approx. 390 BC) was aware of the existence of a congenital form of hip dislocation [87]. Ambroise Paré (1840) was the first to discover the importance of the role played by the inadequate development of the acetabulum.

#### Other important milestones in the development of its diagnosis

- 1846: Wilhelm Roser describes the »ilio-ischeal line«. This line, which passes through the iliac spine, the greater trochanter and the ischial tuberosity, is straight under normal circumstances. In a hip dislocation, however, the trochanter is well above the line, which thus provides a clinical diagnosis.
- 1895: A new era of diagnosis is opened up with the development of the examination technique discovered by C. Roentgen.
- 1937: M. Ortolani [63] describes the »Segno d'all scatto« (»clicking sign«).
- 1962: T.G. Barlow [5] describes a test similar to that of Ortolani.
- 1980: R. Graf [32] develops the ultrasound hip screening procedure.

#### Dates relating to treatment

- 1847: C. G. Pravaz [68]: Longitudinal traction.
- 1885: A. Lorenz [56]: Immobilization with a hip spica cast in the »frog position«.
- 1908: K. Ludloff [57]: Open reduction through a medial approach.
- 1925: H. Hilgenreiner [44]: Introduction of an abduction splint.
- 1955: W. A. Craig [17]: Introduction of overhead traction.
- 1957: A. Pavlik [65]: Treatment with a harness.
- 1968: E. Fettweis [27]: Immobilization with a cast in the squatting position.

#### Occurrence

Epidemiological figures relating to hip dysplasia should be viewed with caution, since both the screening methods and the interpretation of the findings vary greatly in some cases. Conclusions can be drawn about certain trends, however, on the basis of numerous studies [83]. The *dysplasia rate* in Central Europe (Germany, Czech Republic, Austria, Switzerland, Northern Italy) used to be from 2–4% until the late seventies. Today it is much lower. The *dislocation rate* (in historical studies) was 0.5–1%.

In the UK, the USA and Scandinavia, the dysplasia rate is 0.5–1%, and the dislocation rate less than 0.05%. In a recent study in the UK, 88 dislocations were found in 34'723 neonates (=0,25%) [64]. In Bulgaria, 124 cases of dislocation were found in a total of 20,000 neonates (0.6%) [18]. Dislocation of the hip is practically unknown in black populations. A study investigating almost 17,000 African neonates found not a single case of hip dislocation [24]. The absence of hip dysplasia among the primitive tribes of Africa is thought to be due to the fact that the infants are carried by the mother at the side, resting on the pelvis, or on the back with spread legs. Other – more northerly located – primitive peoples, for example the Lapps [31] or certain North American Indian tribes [16], tend to wrap their infants tightly and accordingly experience high dislocation rates. Frequencies as high as 5% have been reported. The female:male ratio is approx. 4:1. Regardless of the improved screen-

ing methods, a general decline in the incidence is nevertheless apparent.

As with other orthopaedic disorders with a genetic etiological component (for example clubfoot or idiopathic scoliosis), this is probably connected with the increased genetic intermixing of the population. The incidence in alpine countries and Central Europe is approaching that of the English-speaking countries. As we noted in an investigation of pediatric orthopaedic institutions in Switzerland, the decline in the incidence peaked between 1960 and 1980, and the subsequent reduction has been rather less pronounced.

### **Etiology and pathogenesis**

Since the introduction of the ultrasound screening method by Graf [32], we know that, in addition to dysplastic and dislocated hips, there are a large number of immature hips. Percentages as high as 30% have been reported. As part of the evolutionary development of humans, the upright gait led to a widening of the iliac wing to provide additional support for the abdominal organs. As intelligence developed, the brain and cranium grew in size while, at the same time, the birth canal became narrower. Humans solved this dilemma by bringing their children into the world in a physiologically immature condition. To this immaturity can be added a number of other factors:

- genetic,
- hormonal and
- mechanical.



Humans are enthusiastic »nest-sitters« and bring their offspring into the world in an immature condition...

Dunn [22] differentiated *two types* of hip dysplasia. The first group shows *general joint hypermobility*, which manifests itself at birth as hip instability. Girls are predominantly affected (the ratio of boys to girls in this group is 1:12). Hormonal, genetic and constitutional factors play a major role in this group.

The second group is characterized by *dysplasia of the acetabulum*, without any significant ligament laxity. Dysplasia is increasingly observed particularly in association with oligohydramnios. This acetabular immaturity is also observed in cases of breech presentation and in connection with other deformities or malformations, e.g. clubfoot, flat feet, facial asymmetries and muscular torticollis. The ratio of boys to girls in this group is only around 1:2, and the left side is twice as likely to be affected as the right side. Mechanical factors associated with the lack of space for the neonate in the uterus play a major role in this group. The consequence is delayed ossification of the lateral acetabular epiphysis, i.e. dysplasia, which leads to secondary dislocation as a result of the inadequate contouring of the acetabular roof. However, the dislocation itself very rarely occurs at birth, but tends to occur secondarily during the course of the first few months of life as a result of the increasing extension in the hip.

As the femoral head starts to be displaced from its central position, this exerts pressure on the lateral acetabular epiphysis, causing ossification and growth to be delayed. Spontaneous normalization is no longer possible by this stage. As the displacement progresses, the femoral head comes out of the acetabulum, usually in a cranio-dorsal direction. The acetabulum is secondarily filled with fatty and connective tissue. If the femoral head has left the acetabulum, shortening of the iliopsoas muscle will occur. The tendon, which is located right next to and partially fused with, the hip capsule, strangles the capsule and becomes an obstacle to reduction. The elevated position of the femoral head causes shortening of the leg. At the same time, the abductors (particularly the gluteus medius and minimus muscles) and the hip extensors (gluteus maximus) are shortened and weakened. This leads, on the one hand, to a flexion contracture of the hip and, on the other, to the inability to stabilize the pelvis when standing on one leg. The consequence is an abnormal pelvic tilt that is compensated by hyperlordosis of the lumbar spine.

If the ossification deficit is only slight, the displacement of the femoral head does not occur, and the acetabular dysplasia may heal up spontaneously during subsequent growth as the ossification catches up. There remains the risk, however, that the joint abnormality becomes exacerbated during the pubertal growth spurt [85]

(■ Fig. 3.167).

## 3.2.4 · Developmental dysplasia and congenital dislocation of the hip

**Diagnosis****Clinical diagnosis in the neonate****History**

- Family history (hip dysplasia or premature osteoarthritis of the hip)
- Firstborn child
- Amniotic fluid deficiency
- Breech presentation.

Hip dysplasia is more common if a corresponding family history exists [45, 64, 83]. Amniotic fluid deficiency and breech presentation are also associated with an increased incidence of hip dysplasia [64, 83].

**Clinical examination****Inspection**

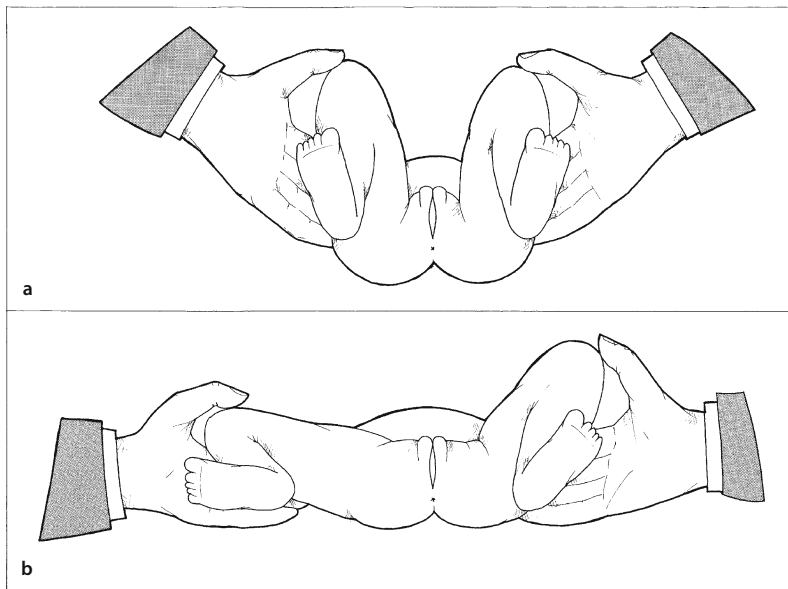
**Asymmetry of skin folds:** Pronounced asymmetry of the skin folds can be an indication of unilateral dislocation.

However, since skin folds in the infant are almost never completely symmetrical, this examination is not very informative.

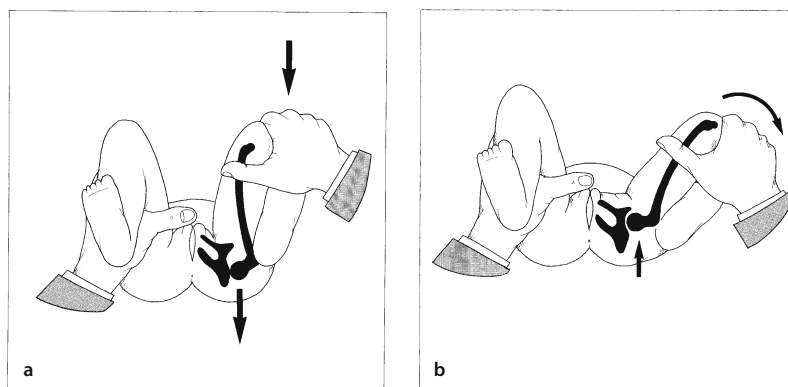
**Leg length examination:** With the hip and knee flexed at right angles, the thigh on the dislocated side is noticeably shorter (■ Fig. 3.152).

**Palpation**

**Examination according to Ortolani [63]:** The hip and knee are flexed at 90°. Grasp the knee, placing the thumb on the inside of the thigh and the index and middle fingers around the greater trochanter (■ Fig. 3.153). First hold the legs in an adducted position and apply gentle pressure in the dorsal direction. Then perform an abduction maneuver, applying slightly greater pressure to the greater trochanter. If the femoral head had been subluxated in the adduction position, a click is perceived as it snaps back into the acetabulum.



■ Fig. 3.152a, b. Testing for *shortening of the thigh* (a) in hip dislocation and for abduction (b)



■ Fig. 3.153a, b. Testing for the *Ortolani sign*

**Examination according to Barlow [5]:** Barlow's test is similar to that of Ortolani, but places less emphasis on the abduction/adduction maneuver, and more on the thumb pressure. Place the hips in a position of central abduction. First apply pressure to the greater trochanter to test the reduction maneuver. Then, from the same abduction position, try to dislocate the femoral head by applying pressure dorsally and laterally. If it snaps back into place, the hip is »dislocatable«. Stabilize the pelvis with the other hand by placing the thumb on the feet and encircling the sacrum with the other fingers. The Ortolani click (■ Fig. 3.153) and the Barlow sign remain positive for approx. 4 weeks in an unstable hip, and cannot be elicited thereafter regardless of the hip condition.

#### Examination of abduction

From a position of 90° flexion, the hips are simultaneously abducted and externally rotated. While the hips of a healthy neonate can almost always be abducted down to the examination table, abduction is inhibited in dislocation or subluxation of the hip in the first 3 months of life. (■ Fig. 3.152).

#### Examination of the range of motion

Neonates usually show a flexion contracture of around 30–40°. This is a physiological finding, since both hips are flexed more than 90° within the uterus. Since it is not possible therefore to examine rotation in the extended position, rotation is examined in the flexed position in the usual way.

**Ludloff's dislocation sign:** Extension of the knees is not normally possible if the hip is flexed by more than 90° because of the tensing of the hamstrings. If the hip is dislocated however, the knee can be extended in this position.

For further details on the examination of the hip in children and adolescents see also ► chapter 3.3.1.

#### Radiographic diagnosis

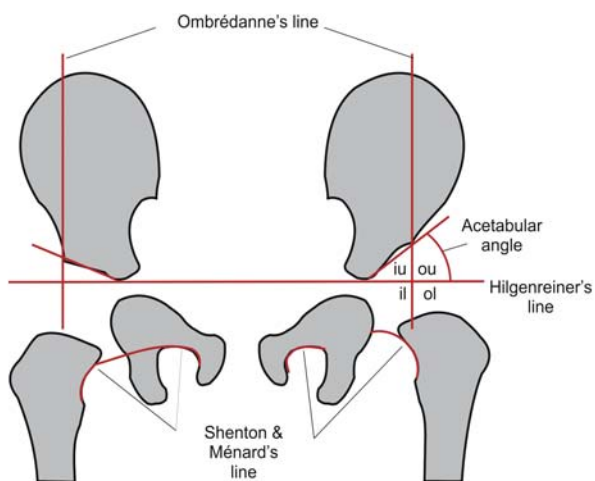
Radiographic diagnosis in infancy is almost completely irrelevant nowadays since it has been superseded by ultrasound, an examination that not only involves no radiation exposure but one that is also more informative. Since the femoral head center starts to ossify after a year or so, the diagnosis must then be made radiologically. At this age, only the AP view is normally recorded (► Chapter 3.2.2). Other x-ray views do not produce reproducible results since large sections of the skeleton are still cartilaginous at this stage and thus not radiopaque. The AP view in the infant should always be an x-ray of both hips so that the pelvic position and the horizontal situation can be evaluated.

A few *guide lines* will facilitate a general evaluation of the AP view of an infant (■ Fig. 3.154 and 3.155).

- The *Hilgenreiner line* [44] joins the two Y-lines of the triradiate cartilage and thus forms the horizontal on the pelvic view. This horizontal reference line is important because the baby does not always lie completely straight on the x-ray plate.
- The *Ombredanne line* [62] is drawn from the lateral edge of the acetabular roof, i.e. the lateral acetabular epiphysis (perpendicular to the Hilgenreiner line) and crosses through the Hilgenreiner line to form four quadrants. Normally the center of the femoral head is in the lower inner quadrant. In the early stages of a dislocation, the center is shifted to the lower outer quadrant and, in a high dislocation, to the upper outer quadrant.



■ Fig. 3.154. AP x-ray of the hips of an 18-month old infant with a high hip dislocation on the left side

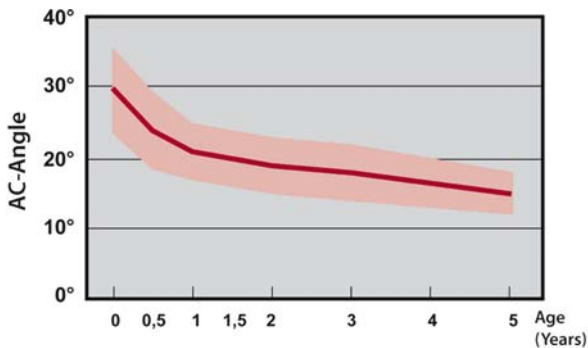


■ Fig. 3.155. Guide lines for evaluating x-rays of the infant hip (Hilgenreiner [44], Ombredanne [61], acetabular angle, Shenton-Ménard [80]); ou outer upper; iu inner upper; ol outer lower; il inner lower



### 3.2.4 · Developmental dysplasia and congenital dislocation of the hip

- **Orientation line according to Shenton and Ménard:** Normally the continuation of the medial femoral neck contour forms a smooth arc as it passes through the superior border of the obturator foramen. In a dislocated hip this arc is disrupted because the femoral neck is displaced upwards.
- **Acetabular roof angle = AC angle or acetabular index [44]:** angle between the horizontal (Hilgenreiner line) and the line joining the Triadiata cartilage and the lateral acetabular epiphysis. The average angle at birth is 30°, at 1 year slightly over 20° and at 3 years of age under 20°. ■ Fig. 3.156 shows the mean values for this angle in infancy and early childhood, although the accuracy of measurement for this angle is not very great ( $\pm 5^\circ$ ).



■ Fig. 3.156. Mean value for the acetabular angle [80]. Statistical distribution in 2,294 normal and questionably pathological hips (mean, single and double standard deviation)

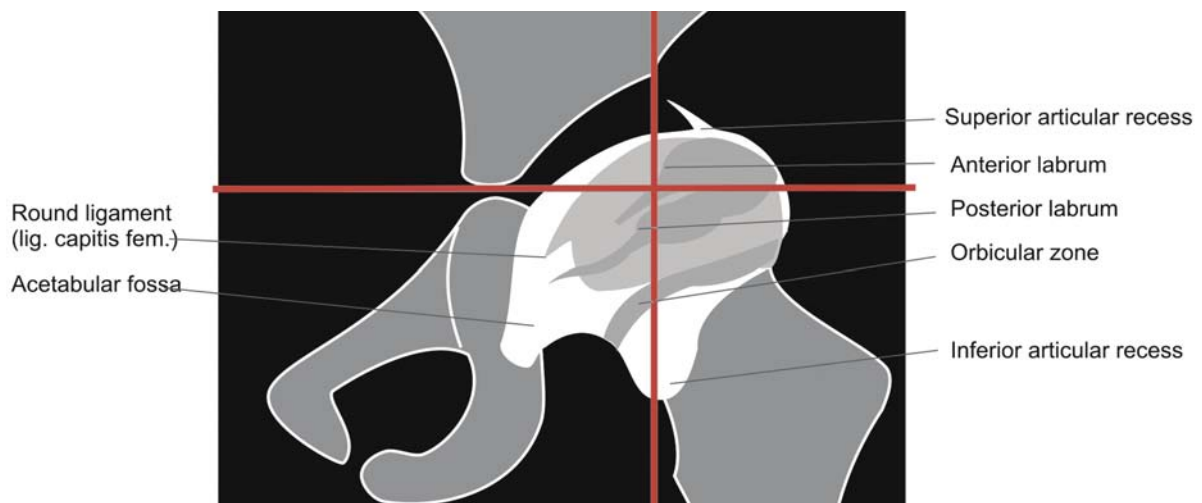
The gap between the femoral head and the radiographic teardrop should not exceed 4 mm up to the age of 4, otherwise instability will be suspected. The radiographic teardrop also deforms over time if dysplasia is present [2].

For details of the radiographic diagnosis of the hip in adolescents and adults see ► chapter 3.2.3.

#### Arthrography of the hip

Hip arthrography is suitable for evaluating the cartilaginous sections of the hip, the ligament of head of femur and other soft tissues. Although it has become less important since the introduction of ultrasonography, it is still valuable for checking the result of a reduction and the centering of the femoral head after a hip dislocation. In particular, soft tissue obstructions in the center of the acetabulum are better evaluated by arthrography than by ultrasound.

We use a caudal approach for the arthrography. The child is placed on a radiolucent table with the legs abducted. From the gluteal fold, a long needle is inserted under sterile conditions and advanced up to the hip under image-intensifier control. 2–3 ml of contrast medium (Jopamiro) are injected. ■ Fig. 3.157 shows an arthrogram of the hip. On the one hand it shows the whole femoral head down to the reflection of the joint capsule and, on the other, the acetabulum from the cranial labrum to the caudal acetabular rim with the transverse ligament. The ligament of the femoral head is also shown. We can readily assess the position of the femoral head in relation to the acetabulum and



■ Fig. 3.157. Schematic view of the arthrographic findings of an infant with a dislocated hip: the whole femoral head down to the reflection of the joint capsule and the acetabulum from the cranial labrum to the caudal acetabular rim with the transverse ligament, also showing the ligament of head of the femur (ligamentum capitis femoris).

The position of the femoral head in relation to the acetabulum and their demarcation are readily assessed; it is also possible to establish whether intra-articular soft tissue obstructions prevent the deep centering of the femoral head

their demarcation, the shape and position of the labrum and the caudal acetabular rim with the transverse ligament.

It is possible to establish whether intra-articular soft tissue obstructions or an hourglass-shaped constriction of the joint capsule interfere with the deep centering of the femoral head. Additionally, the shortened psoas tendon can leave an impression on the joint capsule and represent an obstacle to reduction. The labrum may not be able to open out correctly or may be pushed in, thereby preventing the deep centering of the head.

### Ultrasound examination

At the start of the 1980's, Graf developed a sonographic screening technique for the infant hip [32] that represented a significant advance in the diagnosis of congenital dysplasia of the hip. Before the era of sonography, the average age for starting treatment for a case of hip dysplasia or dislocation in German-speaking countries was over 8 months [50], compared to the current age of just a few weeks. The main contribution made by Graf was to establish a benchmark for examinations offering a high degree of reproducibility.

Sonography of the hip is performed from a lateral approach, and the ilium as displayed on the image must be parallel with the ultrasound head. If this is not the case, the ultrasound head is positioned either too anteriorly or too posteriorly. A linear scanner is required to produce an image allowing a proper assessment of the situation. The vector scanner frequently used in other investigations is not suitable for hip examination, since it produces a dis-

torted image and the parallel alignment of the iliac margin cannot be evaluated [34]. Suitable frequencies are the 7.5 MHz transducer head for small infants and the 5 MHz head for larger infants.

Fig. 3.158 presents the findings that can be viewed and interpreted on the ultrasound scan [32]. Graf introduced two angles as a guide to evaluation: alpha angle (angle between the lateral acetabular epiphysis and triadiate cartilage and the lateral margin of the ilium) and beta angle (angle between the lateral border of the ilium and a line joining the lateral acetabular epiphysis and labrum). Graf subsequently proposed a classification taking into account the various conditions of the hip according to the centering of the femoral head, maturation of the bony epiphysis, steepness of the acetabulum and the age of the patient. Fig. 3.158 and 3.159 illustrate this classification of the sonographic hip findings, including the morphological criteria, corresponding angles and the need for treatment. The nomogram in Fig. 3.160 allows a classification to be made on the basis of the alpha and beta angles.

Graf's ultrasound method has been criticized for a variety of reasons. On the one hand the classification with its combination of figures and letters is not very consistent, since the letters are repeatedly used according to different criteria: types Ia and Ib are differentiated according to the angle, types IIa and IIb according to age and types IIIa and IIIb according to the sonographic density of the cartilaginous epiphysis. The reproducibility of the angle measurements, particularly for the beta angle, is not very great ( $\pm 10^\circ$ ) [21]. But probably the most pertinent

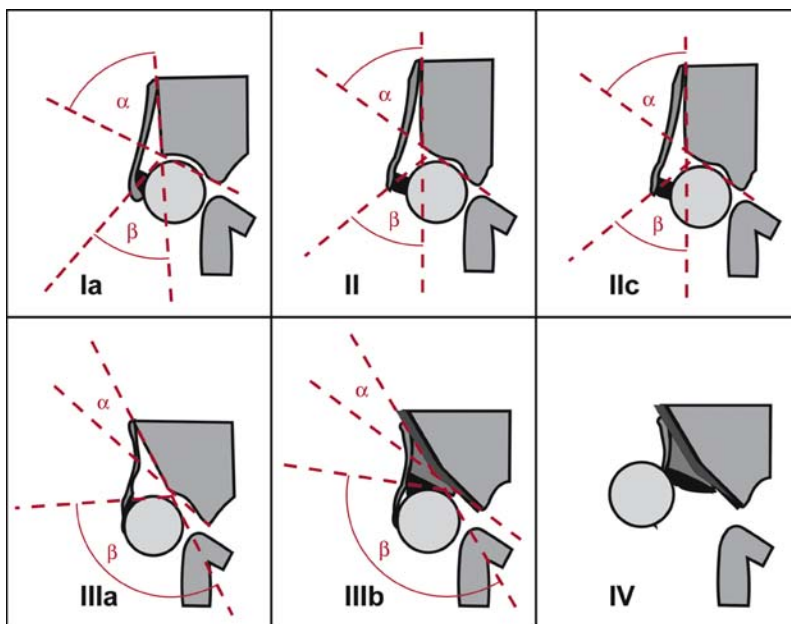


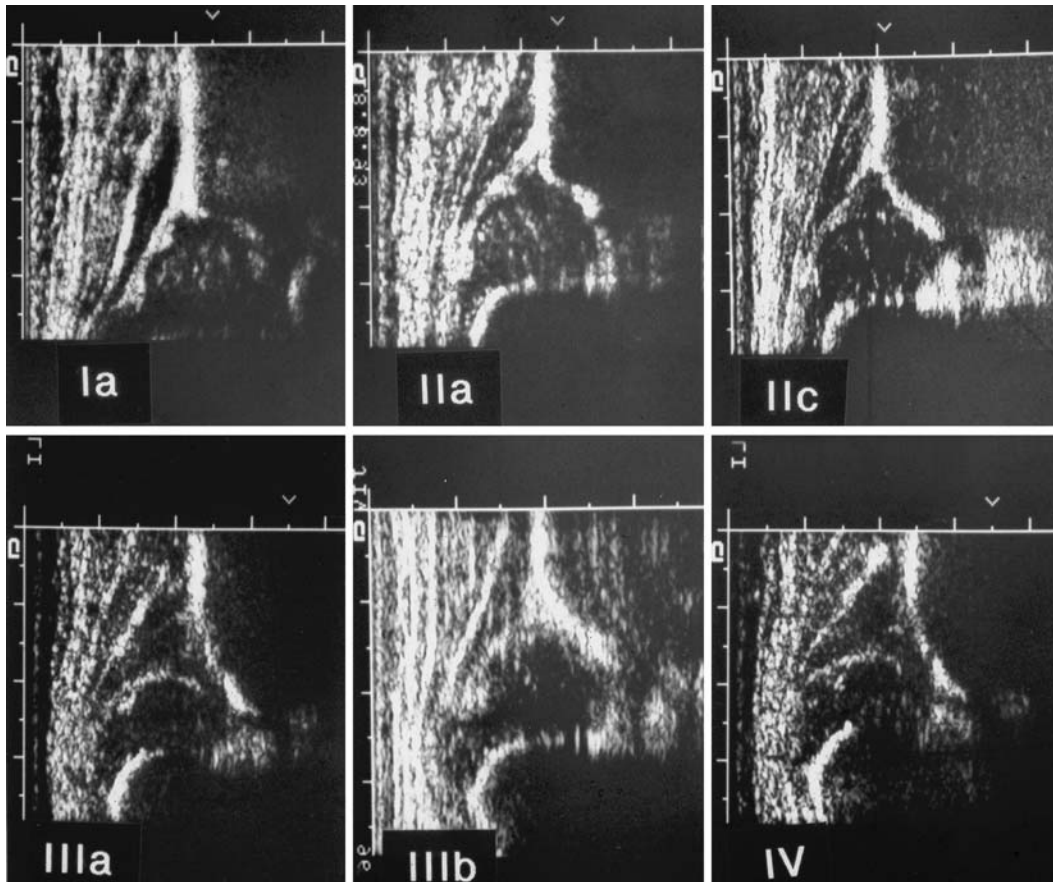
Fig. 3.158. Morphological classification of hip ultrasound findings according to Graf: I normal hip; IIa immature hip; IIc unstable immature hip; IIIa dislocated hip, cartilage extends in the cranial direction, IIIb as for a, but with thickened cartilage, IV dislocated hip, cartilage driven in the caudal direction

### 3.2.4 · Developmental dysplasia and congenital dislocation of the hip

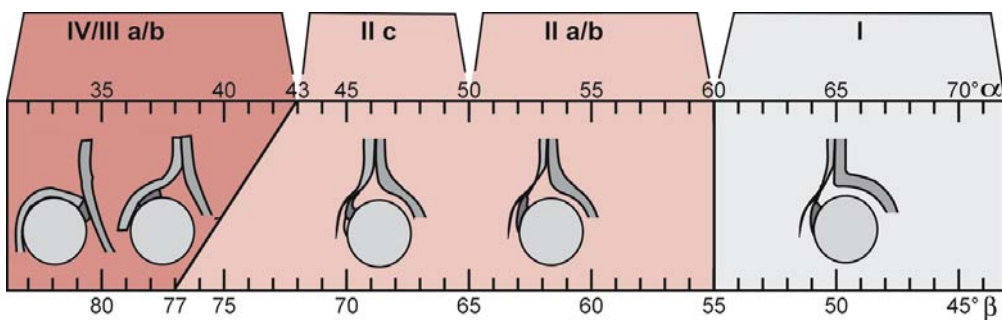
criticism is that this is a purely static examination with a purely morphological assessment and that an important element of hip dysplasia, i.e. the instability or ligament laxity, is disregarded.

As regards the unreliability of the measurements, both the angular measurements (particularly the beta angle) and the evaluation of the individual morphological criteria (shape of the cartilaginous epiphysis, labrum, etc.)

individually show poor reproducibility. If one assesses the overall picture however, the classification is easy, and experienced examiners show substantial agreement when it comes to establishing the type involved. The criticism of poor reproducibility therefore applies only to the consideration of individual parameters in isolation, but not to classifiability and thus the value of the method as a morphological evaluation of the hip.



■ Fig. 3.159. Clinical examples of the principal hip types in the ultrasound investigation of the infant hip



■ Fig. 3.160. Nomogram of  $\alpha$  and  $\beta$  angles and the correlation between these angles and hip types according to Graf

A more problematic aspect, in our view, is the fact that this is a static rather than a dynamic method. Various authors have proposed other, dynamic, ultrasound examination methods that provide a better assessment of joint instability and ligament laxity. The most popular is that described by Harcke [39]. The problem lies in the lack of standardization of these examinations. The room for subjective evaluation is much greater with these dynamic methods than with the purely morphology-based sonography according to Graf.

#### When is ultrasound examination appropriate?

There are numerous studies indicating that cases of hip dysplasia are repeatedly overlooked, and require subsequent treatment, with purely clinical screening of neonates [8, 9, 49, 78]. Ultrasound examination therefore seems a useful screening method for all neonates. In Austria this is largely the case in most of the country, while regional variations apply in German and Switzerland. Several studies also indicate that general screening is more cost effective than treating cases that are discovered too late [9, 51, 85].

The screening of neonates, on the other hand, uncovers a high proportion of immature hips (type IIa) that do not require treatment and usually resolve spontaneously. A recent Dutch study showed that 95.3% of the type IIa+ and 84.4% of the type IIa- hips develop normally if left untreated [70]. Nevertheless, such hips, accounting for approx. 30% of cases, do need to be monitored [25, 33]. It would be more effective, therefore, to implement general screening at the age of 4 weeks. The problem with this approach is that not all infants can be reliably tracked down at this age, whereas they are already in the maternity ward at birth and have to undergo a comprehensive examination in any case. The ultrasound scan is possible up until the time of ossification of the femoral head center, generally up to the age of 9, or a maximum of 12, months.

If general screening is not available, the ultrasound examination should at least be indicated if certain – broadly interpreted – risk factors are present. The corresponding *risk factors* are:

- a family history of hip dysplasia or coxarthrosis,
- premature birth,
- breech presentation,
- other skeletal anomalies,
- oligohydramnios,
- clinical suspicion of hip dysplasia.

These indications have become generally accepted throughout the German-speaking world, whereas ultrasound scanning is much less widespread in English-speaking countries. On the other hand, the incidence of hip dysplasia is also much lower in these countries, where the ultrasound method is only used in a few centers if risk factors are present. In such cases, dynamic examination

methods are generally used [8, 58, 85]. Some authors even consider ultrasound scanning to be wholly unnecessary [43].

A certain amount of rethinking is taking place however. An excellent study from the UK has shown how the treatment costs could be reduced from over £5000 per 1000 neonates after purely clinical screening to £3800 after ultrasound in the presence of risk factors and to £468 with universal ultrasound screening [15]. If the costs of sonographic screening are taken into account, the overall costs are no higher than with purely clinical screening. There is still some dispute, however, as to whether the ultrasound examination should be performed only if risk factors are present or on a universal basis [46, 64]. There is, of course, no 100% certainty. The above mentioned Dutch study also showed that a very small proportion of initially normal hips became abnormal at 3 months (0.4%) [70].

To sum up: ultrasound examination is a valuable addition to the diagnostic arsenal for investigating the hip in infants. Hip dysplasias can be detected at an early stage with a considerable degree of certainty with the Graf method.

**! Universal screening is essential in Central Europe in view of the relatively high incidence of hip dysplasia in these countries.**

If screening is not possible, sonographic examination is indicated in the presence of certain, broadly interpreted risk factors. If applied meticulously, the Graf technique provides a highly reliable overall picture, even if the correspondence in respect of individual parameters viewed in isolation is not particularly good.

#### Treatment

As ultrasound becomes more widespread, concerns are often expressed, particularly by health insurers, about the growing trend of the administration of unnecessary treatments.

**! It cannot be stressed too strongly that an immature hip of Graf type IIa does not require treatment. Abduction splinting should not be prescribed simply because of uncertainty about the interpretation of the ultrasound findings since it can also have side effects (femoral head necrosis). Only if a follow-up examination after 6 weeks shows no progress in terms of maturation (type IIa) may such treatment be introduced.**

#### Conservative treatment

The following *types of treatment* are differentiated:

- maturation treatment,
- closed reduction,
- immobilization.



### Maturation treatment

If an immature hip of type IIa or IIc is detected on the ultrasound scan, the femoral head is not dislocated and does not therefore need to be reduced. A maturation treatment with *abduction pants* or a *Tuebingen splint* (■ Fig. 3.161). The abduction pants were introduced by Frejka in 1941 [28]. These are made of a plastic material and incorporate a rigid bar placed between the legs. The pants hold the legs in abduction and are worn over the infant's normal clothes. The orthosis cannot be worn continuously since it must be removed for nursing care purposes or when changing the baby's clothes.

High rates of avascular necrosis were reported during the first few years of abduction splinting [83], at a time when these orthoses were used for reductions. Excessive abductions of up to 90° were also employed. We therefore use the *Tuebingen splint* developed by A. Bernau [6] for maturation treatment (■ Fig. 3.161). This produces less pronounced abduction but greater flexion than standard abduction pants. It is easy to handle and its size can be adjusted to fit the infant. Since it is made from plastic, hygiene is less of a problem than with the Pavlik harness, for example, which is made of fabric.

### Reduction methods

We differentiate between the following options:

- manual reduction methods,
- braces for reduction,
- traction methods.

### Manual reduction methods

Manual reduction methods are of historical significance only as the associated complication rates were far too high. Manual reductions were described by Lorenz 1895 [56] and Lange in 1898 [53].

### Reduction braces

The *Pavlik harness* [65] incorporates two shoulder straps that cross over at the back and are fastened to a broad chest strap which fastens at the front (■ Fig. 3.162). The lower legs are enclosed by stirrup-like straps, with the topmost strap encircling the leg just below the knee. From the chest strap the shoulder straps continue down to the lower legs. The distance between the chest strap and the lower legs can be adjusted separately by means of buckles at the front and back. The legs are first placed in a flexion position of approx. 110°, which should then be gradually supplemented by increasing abduction. An additional transverse strap can prevent the distraction from exceeding 60°.

This repositioning of the dislocated hip can take a few days in some children, but may require several weeks in others. The dislocated hips reduce themselves spontaneously as a result of the baby's thrashing about, and no actual reduction maneuver is needed. Naturally, this assumes that the infant possesses normal motor skills. The use of this harness beyond the age of 9 months is not recommended [83]. In the hands of skilled practitioners, reduction with the Pavlik harness is a reliable method with few complications [11, 40]. Certain authors, how-



■ Fig. 3.161. Infant with a *Tuebingen splint*. The splint is easy to manage and holds the hips in over 90° flexion and an abduction of approx. 60°



■ Fig. 3.162. Child with a *Pavlik harness*: The harness straps can be adjusted to place the hip in the desired position



ever, report a high number of unsuccessful reductions and complications [55, 60, 91].

On the one hand, these findings were very probably the result of inadequate compliance on the part of the mothers. The Pavlik harness is relatively complicated and the numerous straps can be confusing for the parents. For hygienic reasons, the harness has to be changed frequently, and the constant readjustments can be problematic. The main problem is that the harness very easily becomes soiled by the child and cannot then simply be wiped down like a plastic splint. Accordingly, one study has shown that plastic splints are much easier to manage [3].

On the other hand, the Pavlik harness is more suitable for reducing subluxated (Graf type III) hips than completely dislocated (Graf type IV) hips [60]. Another study has also reported a relatively high necrosis rate of 33% after reduction with the Pavlik harness [80].

### Traction methods

We make a basic distinction between two methods:

- longitudinal traction,
- overhead traction.

**Longitudinal traction:** Longitudinal traction for reducing the hip is the first known therapeutic procedure and was described by Pravaz in 1847 [68]. It is still used today, in some cases as a home-based treatment. The traction is achieved with plaster strapping affixed to the legs. A board placed beneath the feet is designed to avoid pressure on the malleoli. The traction weight is initially 1/7 of the infant's weight, but can subsequently be increased to 1/4 or more. The skin should be monitored carefully. Triangular pants can be used to provide counterforce, or else the foot of the bed can be elevated so that the weight of the body is shifted towards the head. The legs are abducted by approx. 20°.

**Overhead traction:** Overhead traction was introduced in 1955 by Craig [17], and remains a widely used method even today. This traction can also be employed for older children for whom a Pavlik harness is no longer appropriate. This treatment remains the standard method in our hospital. Overhead traction requires the fitting of two bars at the side of the bed which are linked together above the bed by a crossbar. A weight of 1–1.5 kg is attached to the child's legs with strapping and exerts traction via a cord that runs over pulleys. The degree of traction should initially be adjusted to produce a flexion of over 90°. The pulleys are then shifted laterally to gradually increase abduction (■ Fig. 3.163).

We shift the pulleys so as to achieve an abduction of around 70° after 8–10 days. By this time spontaneous reduction has occurred in most cases, and this can be checked by arthrography. If the traction were increased

to 90° abduction, there would be an increased risk of femoral head necrosis. Reduction with overhead traction must be followed by immobilization, for which we use the Fettweis spica cast (■ Fig. 3.164). Traction improves the chances of a successful closed reduction and reduces the risk of avascular necrosis of the femoral head[94].

### Immobilization

The following can be used for immobilization:

- plaster casts,
- splints,
- braces,
- abduction pants.



■ Fig. 3.163. Infant in *overhead traction*. The pulleys are shifted laterally to increase hip abduction



■ Fig. 3.164. Child in a *Fettweis cast*. This hip spica holds the hips in over 90° flexion and approx. 60° abduction

### Plaster casts

**Hip spica in the Lorenz position:** This oldest known immobilization treatment described by Lorenz in 1895 [56] fixed the hips in an abduction position of 90° (also known as the »frog position«). We know from large-scale statistical analyses [81] that very many cases of avascular necrosis of the femoral head have occurred as a complication of immobilization in this position. While it was once assumed that this complication was caused by compression of the medial circumflex femoral artery by the posterior acetabular rim during the right-angled abduction, more recent studies have shown that the intra-articular pressure produced by pronounced abduction and internal rotation is excessive and causes constriction of the intra-epiphyseal vessels in the soft cartilage [92]. This also explains why femoral head necroses are less frequent after reductions if the ossification center of the head is present [73]. Immobilization in the Lorenz position is therefore no longer practiced.

» *...Medical specialists also primarily objected to this method because of the need to keep a child in a plaster cast in such a barbaric position for months on end ...* «

*(Albert Lorenz writing about the bloodless reduction and immobilization method developed by his father Adolf Lorenz).*

**Immobilization in the Lange position:** In 1898 Lange [53] proposed immobilization in a position of maximum internal rotation and pronounced abduction. This has likewise become obsolete.

**Immobilization in a squatting position according to Fettweis:** In 1968 Fettweis [27] proposed a treatment of reduction and immobilization in a hip spica in the squatting position, in which the hips are flexed by up to 110–120°, but limiting the abduction to approx. 50°–60° (■ Fig. 3.164). Various statistical analyses have shown that the rate of avascular necrosis is much lower, at around 5%, with the squatting position than with the Lorenz position at approx. 15%. The long-term treatment with the Fettweis cast is also very well tolerated by the children. Age is not a relevant factor for this treatment.

Another major advantage of cast treatment is the optimal compliance, which avoids the risk of the child being moved out of the ideal position for prolonged periods. After a reduction we accordingly always use the Fettweis cast for at least 8 weeks for immobilization purposes. The cast must be changed after 4 weeks. The cast can be changed under light sedation and does not usually require general anesthesia. The feet do not need to be included in the cast but can be allowed to move freely. The cast need not necessarily be prepared from white plaster and

we often use Softcast instead. A sufficiently wide section is cut out of the cast around the buttocks. Self-adhesive plastic inserts that prevent soiling of the cast are available on the market.

### Splint treatment

Various abduction splints are used for immobilization purposes. These are particularly suitable as follow-up treatment after immobilization in a Fettweis hip spica. The Denis Browne splint, introduced in 1948 [10], used to be very popular since it was very easy to manage. However, since it suffers from the drawback of having been designed for an abduction position of 90° this splint should no longer be used.

Numerous modifications of the Denis Browne splint, with the aim of producing a better position, have been proposed. A well-known example is the Tuebingen splint (■ Fig. 3.161), which we tend to use. After a congenital dislocation of the hip, we follow 3 months of permanent immobilization in the squatting cast with a further 3 months of splint treatment. We consider the abduction pants to be inadequate as a maturation treatment after dislocation. The abduction pants are worn over the clothing, while the splint is worn under the clothing. We do not usually administer a maturation treatment exclusively during the night.

The Pavlik harness (■ Fig. 3.162) is also suitable for immobilization purposes, although it is not particularly appropriate for use in infants older than 9 months. Since the Pavlik harness is not very practical for the mother, we only use it occasionally. Various reports in the literature have described failed reduction or subsequent dislocation in the caudal direction after the use of the Pavlik harness [69]. The treatment is only suitable if the parents are cooperative and intelligent. The Pavlik harness is very popular in English-speaking countries.

### Complications after conservative treatment

#### Avascular necrosis of the Femoral head

The commonest and most serious complication of treatment of congenital dislocation of the hip is avascular necrosis of the femoral head. Although it can also occur in untreated hip dislocation, it is very rare in this context. In most cases, the necrosis is a consequence of treatment and does not result from the dislocation itself. The necrosis can occur in the epiphyseal plate either laterally, centrally or medially (■ Fig. 3.165) [83], but most often laterally (■ Fig. 3.166). This results in shortening of the femoral neck, or »head in neck position«, and overgrowth of the greater trochanter. The same shortening of the femoral neck and overgrowth of the greater trochanter is also seen with central necrosis, whereas medial necrosis results in a coxa vara. But the necrosis can also affect the acetabulum.

According to Salter the following 5 factors are important for the *diagnosis of femoral head necrosis*:

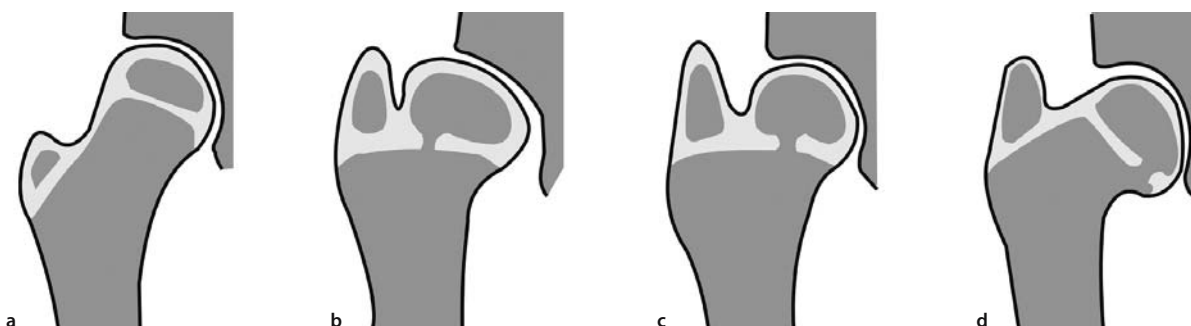
1. Absence of ossification of the femoral head center for more than 1 year after the reduction.
2. Absence of growth of an existing femoral head center for at least 1 year after the reduction.
3. Widening of the femoral neck during the year following the reduction.
4. Increased bone structure of the femoral head center on the x-ray, possibly with subsequent fragmentation.
5. Presence of a deformity of the femoral head and neck after the end of the recovery phase (coxa magna, coxa plana, coxa vara, short femoral neck).

A *classification* for the severity of the necrosis, presented in **Table 3.16**, was proposed by *Toennis* [81].

The necrosis rate depends partly on the type of reduction and partly on the immobilization method. **Table 3.17** shows this correlation on the basis of statistical data collated by the Hip Dysplasia Study Group of the German Orthopaedics and Traumatology Association. As regards the type of reduction, the overhead method appears to be associated with the lowest rate of necrosis, while the Hoffmann-Daimler brace caused the most cir-



**Fig. 3.166.** X-ray of a 4-year old girl after congenital hip dislocation and lateral femoral head necrosis with lateral epiphyseal closure, head-in-neck position and shortening and valgus displacement of the femoral neck



**Fig. 3.165a–d.** Avascular necrosis. of the epiphyseal plate after (conservative or surgical) treatment of hip dysplasia or dislocation (*light gray cartilage, dark gray bone*). **a** Normal situation. **b** Lateral epiphyseal closure (most common); growth in the head-in-neck position, femoral neck shortening, overgrowth of the greater trochanter, coxa plana.

**c** Central epiphyseal closure (rare); femoral neck shortening, slight overgrowth of the greater trochanter. **d** Medial epiphyseal closure (very rare); femoral neck shortening, tilting of the femoral head medially, slight elevation of the greater trochanter

**Table 3.16.** Classification of the severity of avascular necrosis. (After [81])

Grade	Radiographic finding	Prognosis
Grade 1	Slightest degree of changes, femoral head ossification center with slightly blurred margins, slightly granular and irregular in structure	Generally regresses over time
Grade 2	More blurred margins of the ossification center with irregular structure, more granular than for Grade 1; poss. also cyst formation or punched out partial defects	Changes usually regress, poss. slight flattening of the head
Grade 3	Whole femoral head ossification center disintegrated or only visible as individual fragments or a flat strip. Very small head nucleus, poss. completely broken up or only visible after many months	Frequently: deformations of the femoral neck initially, which regress at a later date
Grade 4	Additional involvement of the epiphyseal plate, irregularities are also apparent on the margins of the epiphyseal plate at the femoral neck	Serious consequences for growth

## 3.2.4 · Developmental dysplasia and congenital dislocation of the hip

culatory problems. As regards the immobilization on the other hand, the Fettweis squatting position was by far the most favorable method with just 2% of necroses. Necrosis rates of 16% and 27%, respectively, were recorded for the Lange and Lorenz positions. The Pavlik harness was also associated with a fairly low necrosis rate, at 7%. Naturally, the necrosis rate after surgical treatment cannot be compared with the conservative methods since this involves a different population.

» *...The improvement of the Lorenz reduction method did not simply spring from a single individual, like armed Athene from the head of Zeus, but emerged gradually from the cooperation of many scientists... «*  
(Albert Lorenz)

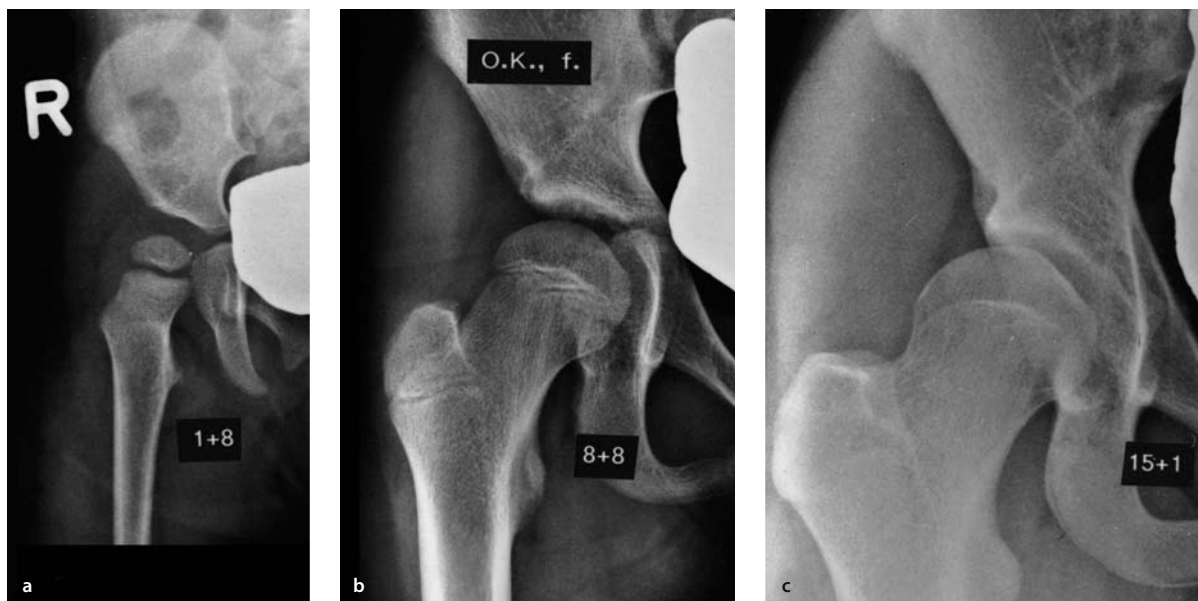
**Secondary deterioration**

For a long time, doctors assumed that once a hip had returned to normal after treatment it could no longer deteriorate. But this assumption now needs to be revised. In recent years we have observed several cases in which a normal hip during childhood has deteriorated into a distinctly dysplastic hip during puberty (■ Fig. 3.167). Evidently, premature closure of the triadiate cartilage can occur during puberty so that the acetabulum no longer adequately matches the growth in size of the femoral head.

! **Every treated hip must be monitored radiographically until adulthood. X-rays (AP) should – as a minimum requirement, i.e. if no special features are present – be recorded after the start of walking, at the age of 8–10 years and on completion of growth.**

■ **Table 3.17.** Overall percentage of head necroses in various fixation positions, classified according to the reduction methods [(-) insufficient number for statistical evaluation]. Pooled statistics from 3,137 treated hip dislocations. (After [81])

Reduction method	Fixation position according to			
	Lorenz (%)	Pavlik (%)	Fettweis (%)	Lange (%)
Overhead traction	15	(-)	(-)	(-)
Manual	25	(-)	(-)	(-)
Surgical	28	(-)	(-)	(-)
Hoffmann-Daimler	32	(-)	(-)	(-)
Total	27	7	2	16
n	1843	369	855	70



■ **Fig. 3.167a–c.** AP x-ray of the right hip in a 20-month old girl after splint treatment. The lateral acetabular epiphysis is still fairly flat and the cup itself fairly steep. No operation was performed (a). By the age of almost 9 years the hip is normal with good acetabular coverage

(b). By completion of growth, the hip has deteriorated significantly. It is clearly dysplastic, and acetabular coverage is inadequate. The CE angle is less than 10°



### Surgical treatment

The surgical treatment of congenital dislocation of the hip serves the following purposes:

- open reduction,
- joint-correcting measures.

### Open reduction

An open reduction (see below) is needed if the hip cannot be reduced in the closed procedure. In the young infant this almost always applies only in cases of teratological dislocation. The longer a hip is dislocated, the more likely it is that secondary changes aggravating any reduction of the head into the acetabulum and impairing the stability of the joint will develop. The femoral head becomes displaced cranially and the capsule is pulled out. The primary acetabulum does not develop correctly and becomes dysfunctional.

If the femoral head strikes the acetabular rim, the cartilaginous epiphysis becomes deformed, possibly resulting in the formation of a cranially-extending channel. Fatty and connective tissue accumulate in the unused hollow space. As the femoral head is displaced, the iliopsoas muscle is pulled upwards and shortened, potentially constricting the capsule tube. The transverse ligament can also protrude like a crescent and thus hinder reduction.

The open reduction can be performed via a medial [57] anterior, lateral or dorsal approach. We prefer the anterior approach. The incision in this case is cranial to the inguinal ligament, subsequently resulting in a very satisfactory cosmetic result. We approach the hip both medially and laterally to the psoas muscle to produce a very good overview. The following factors must be borne in mind during open reduction:

- the ligament of the femoral head usually has to be resected,
- the acetabulum must be completely cleared out and freed of soft tissues,
- the transverse acetabular ligament must be indented,
- aponeurotic lengthening of the psoas muscle is often required,
- if the femoral head is in a high position, a shortening osteotomy may also be needed,
- the widened joint capsule must be sutured and drawn tight.

**! Two points are crucially important for the subsequent recovery:**

- An abnormally high pressure must not develop in the joint.
- The femoral head must be deeply centered.

Studies have shown that the deep centering is by far the most important prognostic factor for the subsequent development of the hip, including in respect of the risk of renewed dislocation [12, 29]. More recent MRI studies, however, indicate that the centering is not usually ideal

even after a good operation and only returns to normal after 1 year [23]. Since the incidence of femoral head necrosis increases with age we no longer attempt a closed reduction of a high dislocation in children after the first year of life, but proceed directly to an open reduction. In children aged 2 and over an additional shortening osteotomy is usually required, as it is for a high dislocation in children from 1 year of age.

Open reduction is *indicated*:

- in the first year only if closed reduction proves unsuccessful (particularly with a teratological dislocation; ► chapter 3.2.7); as an alternative an attempt can be made to cut the psoas tendon and the transverse ligament arthroscopically and then retry closed reduction.
- in the second year primarily for a high dislocation, i.e. if the femoral head center is higher than the triradiate cartilage or if the closed reduction proves unsuccessful;
- from the third year we no longer attempt closed reduction, but proceed directly to open reduction;
- from the fifth year we perform an open reduction only for a unilateral dislocation. The situation should be left as is for a bilateral dislocation (unless a neoacetabulum has formed). The suffering after a reduction attempt is probably greater than if the dislocation is left as is (■ Fig. 3.168).

After performing an open reduction we immobilize the hip in a *hip spica* in the squatting position [27] for at least 3 months. Splinting is then required for at least a further 3 months. The older the child, the longer the follow-up treatment lasts.

Even an experienced orthopaedic surgeon is not always able to reduce the hip in a primarily stable position



■ Fig. 3.168. AP x-ray of the pelvis of a 19-year old female patient with bilateral, high, untreated hip dislocation. The girl walks with a definite »waddle«, but does not have any symptoms apart from occasional lumbar pain; she also takes part in sport (ice skating, skiing)



with open reduction. The anteromedial capsule, transverse ligament, psoas muscle [14] or a constricted, hour-glass-shaped capsule are often responsible for preventing a proper reduction. Sometimes the acetabulum is too small in relation to the femoral head. If the first reduction fails, we generally wait until the child reaches the age of 18 months before making a second attempt. The reduction can then be supported with joint-correcting measures on the acetabulum and femur (see below). Aseptic necrosis occurs as a complication of open reduction in up to 27% of cases [1].

**! Every experienced pediatric orthopaedic surgeon has a list of failures that has caused many a sleepless night. Dislocations – and not just teratological ones – can sometimes show anatomical features that prevent the stable centering of the hip, particularly in small children.**

#### Joint-correcting measures (■ Table 3.18)

Joint-correcting measures are usually performed secondarily, i.e. not at the same time as the reduction, whether closed or open. Technically correct osteotomies on the pelvis are only feasible from the age of 18 months. Joint-correcting measures can be performed essentially at the following sites:

- the thigh,
- the pelvis.

#### Femoral osteotomies as joint-correcting measures

Operations on the femur can be performed at the following sites:

- intertrochanteric,
- subtrochanteric,
- on the greater trochanter (trochanteric transfer).

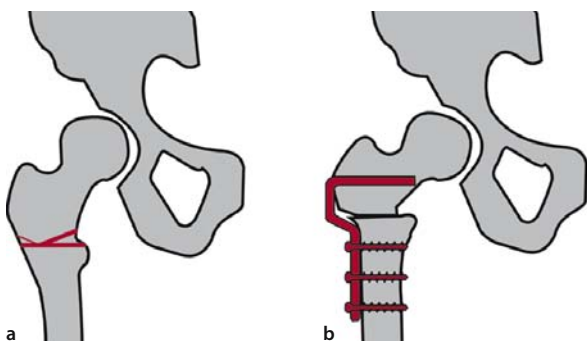
**Intertrochanteric shortening osteotomy:** This operation is frequently required for infants with a high dislocation of the femoral head simply in order to move it to a lower position. The femur can be shortened at inter- or subtrochanteric level. The disadvantage of the intertrochanteric osteotomy is the need to chisel the attachment of the psoas muscle off the lesser trochanter. The disadvantage of subtrochanteric shortening, on the other hand, is the substantial tension arising at the shortened psoas tendon, although this can sometimes be offset by aponeurotic lengthening of the tendon. We tend to shorten the femur with the intertrochanteric procedure. We do not use a step-cut osteotomy for shortening in infants but simply divide the bone smoothly and remove a bone fragment of the desired length. The result is fixed with an infant's angled plate. Further details of the shortening osteotomy with a step cut are provided in ► chapter 4.2.2.

**Intertrochanteric varus/derotation osteotomy:** Increased anteversion of the femoral neck is frequently seen in connection with hip dysplasia or dislocation. This is only rarely associated with a coxa valga. The valgus position of the femoral neck can often be misinterpreted on the AP x-ray because of the increased anteversion. A correction x-ray with internal rotation can provide information about the precise neck-shaft angle configuration (► Chapter 3.2.8, ■ Fig. 3.220).

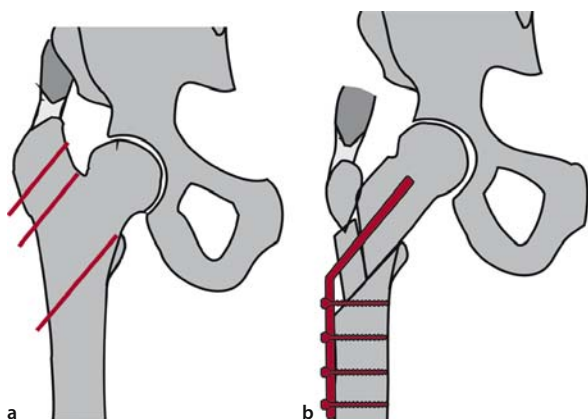
While an anteverted hip in association with hip dysplasia used to be surgically corrected (at least in Europe) up until the 1970's, the value of this correction is now disputed. In the USA, even then, preference tended to be given to acetabular roof reconstruction. In recent years, the belief that acetabular roof reconstruction is better than intertrochanteric osteotomy for improving the biomechan-

■ Table 3.18. Indications for joint-correcting measures

Age	Finding	Operation
<2 years	–	Joint-correcting operations not usually indicated
2–8 years	AC angle >25°, flat lateral epiphysis	Salter pelvic osteotomy, poss. intertrochanteric derotation/varization osteotomy
>8 years	CE angle ≤10°, head and acetabulum spherical and congruent	Triple osteotomy
>8 years, including adults	CE angle ≤10°, head subluxated cranially, acetabular radius too large	Intertrochanteric shortening osteotomy, acetabuloplasty
>10 years, including adults	CE angle ≤10°, head aspherical, acetabulum clearly too small	Poss. Chiari pelvic osteotomy, poss. augmented with shelf
Adults	CE angle ≤10°, head and acetabulum spherical and congruent	Periacetabular osteotomy
Adults	CE angle ≤10°, head and acetabulum aspherical, but congruent	Combination of periacetabular osteotomy with intertrochanteric valgization osteotomy



■ Fig. 3.169a, b. Principle of the *intertrochanteric varus osteotomy* and fixation with 90° angled blade plate; a preoperatively, b postoperatively



■ Fig. 3.170a, b. Principle of the *femoral neck lengthening osteotomy* according to Morscher [40]. Three parallel osteotomies are performed at the same time. The shaft is moved to a more lateral and distal position using a 130° angled blade plate, the middle fragment is inserted between the femoral neck and the bend in the plate while the trochanteric fragment is moved distally; a preoperatively, b postoperatively

ics of the joint has also gained acceptance in Europe. The latter procedure also has the disadvantage that revalgization frequently recurs during the course of subsequent growth. At least the intertrochanteric derotation/varus osteotomy has a secondary effect on the acetabulum, improving the shape of the acetabulum directly by altering the pressure distribution [72]. The principle [57] of the intertrochanteric osteotomy is shown in ■ Fig. 3.169. The result is fixed with an angled plate.

An anteverted hip on its own, without the presence of hip dysplasia, does not constitute an increased risk for osteoarthritis [90]. On the other hand, a retroverted hip is definitely carries a significant risk for early osteoarthritis [84] because of impingement.

**Femoral neck lengthening osteotomy:** A typical consequence of femoral head necrosis is shortening of the femoral neck with concurrent overgrowth of the greater trochanter, since the trochanteric apophyseal plate is not affected by the necrosis. This configuration will result in abductor weakness of varying severity. A femoral neck lengthening osteotomy can be performed to restore the proper biomechanical configuration [41]. ■ Fig. 3.170 shows the principle of this operation, in which

1. the femoral neck length is restored,
2. the lever arm of the abductors is improved by transfer of the greater trochanter and
3. the leg shortening that is usually present is at least partially compensated at the same time.

A lengthening of around 1–1.5 cm can be achieved with this operation (■ Fig. 3.171). The surgeon must be very careful, however, to avoid injury to the vessels that enter



■ Fig. 3.171. X-ray series for a 12-year old boy after a congenital hip dislocation and lateral femoral head necrosis with lateral epiphyseal closure, head-in-neck position and shortening of the femoral neck (a). At 14 years of age on completion of growth (b) pronounced trochan-

teric elevation. A femoral neck lengthening osteotomy was implemented to correct the length of the femoral neck and the lever arm of the abductors (c). Situation 1 year postoperatively (d)

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the joint capsule and supply the femoral head. Since the pressure in the joint is increased as a result of lengthening of the femoral neck, the procedure is indicated only if the joint conditions are good (largely normal).

#### Pelvic procedures

As regards the pelvis, the following basic distinction is made between the following types of operation:

- Salter osteotomy of the innominate bone,
- acetabuloplasty,
- Chiari osteotomy of the ilium,
- triple osteotomies,
- periacetabular osteotomies,
- shelf operations.

All of these operations have their own indications and are still commonly performed.

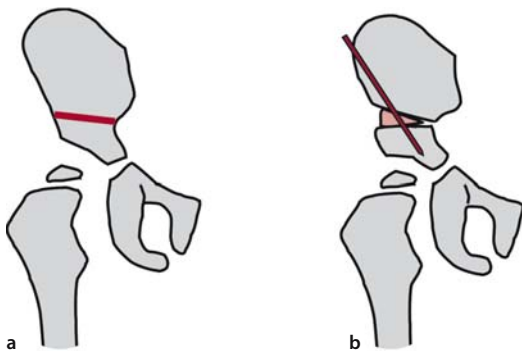
**Salter's osteotomy of the innominate bone (ilium):** In Salter's osteotomy [71], the pelvis is divided above the anterior inferior iliac spine down to the transverse sci-

atic foramen. The acetabulum is pulled ventrally and laterally. A triangular wedge of bone secures the resulting position. The pivot point for the transfer is the symphysis. This operation flattens an excessively steep acetabular roof, improves the roof coverage ventrally and narrows the acetabular angle (see above) (■ Fig. 3.172 and 3.173).

The Salter pelvic osteotomy is indicated for an excessively steep acetabulum in a child aged between 2 and 8 years. We hardly ever perform the Salter osteotomy before the age of 2, preferring to wait and see how the situation develops spontaneously. Many mild cases of hip dysplasia improve over time and do not require treatment [26]. Only if the acetabulum is very small, thus preventing a stable closed reduction, do we follow the Salter osteotomy with an open reduction in the same session. Even in 2-year old patients we frequently await the spontaneous outcome of events despite an acetabular angle of over 30°, since the acetabulum can largely correct itself during this stage of development provided the femoral head is well centered.

Even more important than the acetabular angle for the evaluation is the shape of the lateral acetabular epiphysis and the concavity of the joint surface. If, by the age of 3 years, an acetabular angle of 30°, a flat epiphysis and inadequate concavity of the joint surface are all still present, then the Salter osteotomy is indicated. Since the operation is only feasible while the symphysis remains sufficiently mobile, it is no longer indicated after the age of 8 [38]. A Salter osteotomy can restore the normal hip configuration in small children and even excellent long-term results can be expected. Although one would expect lateralization of the femoral head to occur as a result of the angular movement with the center of rotation in the area of the epiphysis, this does not actually happen in reality [93].

The postoperative management after a Salter osteotomy involves fixation in a hip spica for 6 weeks. The fixation wires are subsequently removed and the child is mobilized. The operation should not be performed on



■ Fig. 3.172a, b. Principle of the *Salter pelvic osteotomy (osteotomy of the innominate bone)*. The ilium is divided above the anterior inferior iliac spine (a), the distal fragment is transposed ventrally/laterally, a triangular bone graft with the base facing laterally is wedged between the fragments and fixed with a Kirschner wire (b)



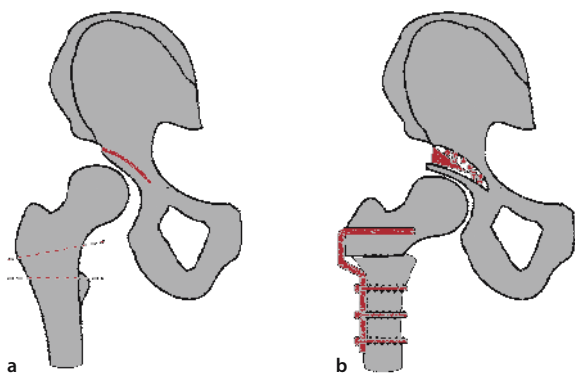
■ Fig. 3.173a–c. X-ray series for a 3-year old child with hip dysplasia involving an excessively steep acetabular roof and slightly off-center hips on both sides (a). An *intertrochanteric derotation varus osteotomy*

(here fixed with an external fixator instead of an angled blade plate) and a *Salter pelvic osteotomy* was performed on both sides with an interval of 4 weeks (b). Situation 1 year postoperatively (c)

both sides at the same time as a counter support is needed on the opposite side for the rotation of the acetabulum. The contralateral side should therefore be operated on at the earliest after 4–6 weeks.

While the Salter osteotomy is a relatively simple and tried-and-tested operation, complications can still occur with this procedure. A lesion of the sciatic nerve can occur when the Gigli saw is used in the greater sciatic foramen. We ourselves have had the misfortune to observe an irreversible partial sciatic nerve lesion (after several operations). Vascular injuries, delayed bone healing and deformation of the iliac crest are other possible complications.

**Acetabuloplasty:** An acetabuloplasty involves a dome-shaped osteotomy approx. 1–1.5 cm above the acetabulum in the direction of the triradiate cartilage. The acetabulum is shifted distally by the insertion of a wedge. The



**Fig. 3.174a, b.** Principle of acetabuloplasty. A curving osteotomy is cut above the acetabulum extending to the (former) Y-line (a), and a robust bone graft is pinned in place (b). This operation is usually combined with an intertrochanteric shortening osteotomy

main indication for an acetabuloplasty is a non-round or excessively flat acetabulum (■ Fig. 3.174 and 3.175). We perform an acetabuloplasty most often for neuromuscular hip dislocations (▶ Chapter 3.2.8). In principle, acetabuloplasty is also a suitable operation for an excessively steep acetabulum in toddlers.

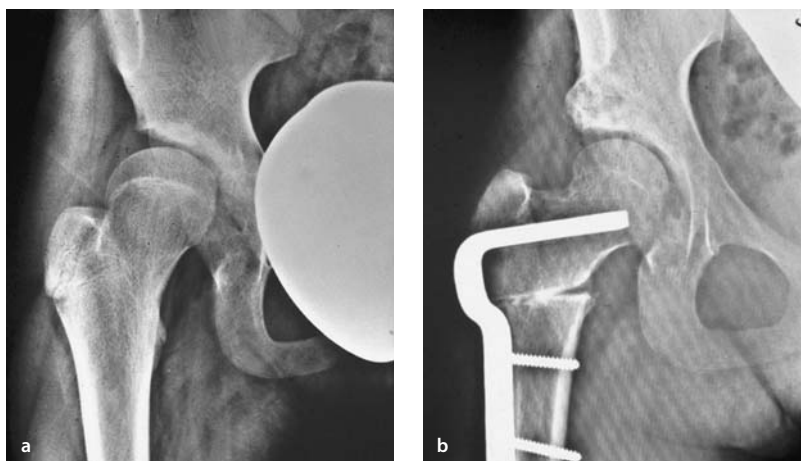
The correction options with acetabuloplasty are better than those with the Salter osteotomy, as the pivot point with the former procedure is nearer the acetabulum (triradiate cartilage compared to the symphysis). However, the risks associated with acetabuloplasty are greater. Growth disorders in the triradiate cartilage, in particular, can occur. Furthermore, the osteotomy is performed closer to the joint and is technically more demanding than Salter's innominate osteotomy. We therefore perform the acetabuloplasty primarily in cases of a non-round acetabulum.

Various techniques have been described for the acetabuloplasty, the first originating from Koenig [52]. Spitzzy described a technique involving the insertion of a tibial bone graft, which then protruded laterally as in the shelf operation (see relevant section) [76]. The standard technique used nowadays derives from Dega. Pemberton [66] modified this technique and shifted the acetabulum not only distally, but anteriorly as well.

Since we perform an acetabuloplasty primarily for non-round acetabula, the technique is adapted to the initial situation in each case. If the lateral part is too steep, for instance, then this section is turned down accordingly. If the deformation is located ventrally, the correction focuses mainly on the anterior section.

**Triple osteotomy:** Le Coeur [54] was the first to describe a triple osteotomy of the pelvis. Modifications were subsequently proposed by Hopf [47], Sutherland [79], Steel [78] and Toennis [82]. In all of these osteotomies the ilium, ischium and pubis are divided. The ilium is divided

**Fig. 3.175a, b.** 13-year old girl with very dysplastic acetabulum with an excessive radius and a subluxating femoral head that is too high. **a** AP x-ray of the right hip. **b** 1 year after intertrochanteric shortening osteotomy of 2 cm and acetabuloplasty. The radius of the acetabulum was normalized by this procedure, and the femoral head is well centered

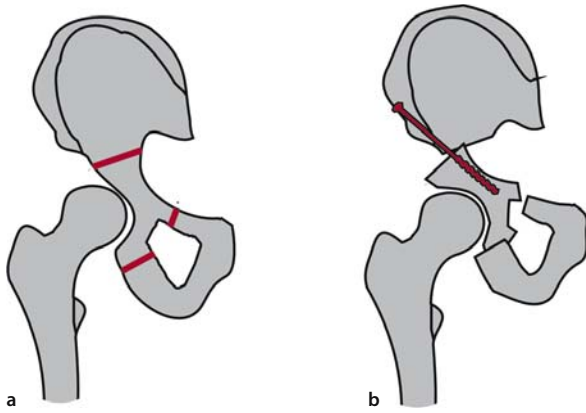




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with an osteotome or saw horizontally above the anterior inferior iliac spine, i.e. roughly at the same level as the Salter osteotomy. The ischium and pubic bone are divided differently in the various methods. Le Coeur [54] and Sutherland [79] osteotomized the two bones close to the symphysis. As a result the pivot point was located relatively far from the hip.

Hopf [47], Steel [78] and Toennis [82] proposed osteotomies close to the acetabulum. For children, we employ a modification of the technique described by Steel



■ Fig. 3.176a, b. Principle of the *triple osteotomy*. The ischium, pubis and ilium are divided close to the acetabulum (a); the acetabular fragment is rotated laterally so that movement centers on the middle of the head rather than the ischium (b). If the latter forms a hinge, the joint has been placed in an excessively lateral position, which produces an adverse effect on the lever arms of the muscle and thus the exerted muscle force

[78]. Through a separate medial Ludloff approach [59], we cut the ischium much closer to the acetabulum than described by Steel. We osteotomize the pubic bone by making the cut above the inguinal ligament, while the ilium is divided with the Gigli saw, as in Salter's osteotomy (■ Fig. 3.176 and 3.177).

The triple osteotomy can increase the loading area in the mechanically important anterior and lateral sections of the hip, although this is achieved at the expense of the biomechanically less important caudal medial sections. The biomechanical efficacy of this principle was presented in a recent study [48]. The acetabulum is rotated in a lateral-anterior direction – or if necessary in the individual situation – in a lateral-posterior direction. Since the acetabulum is then able to swivel over a very wide range, there is also a certain risk of over-correction.

The triple osteotomy is indicated if the acetabular coverage in the lateral or ventral direction is too small. This is expressed in a CE angle of less than  $10^\circ$ . The ventral coverage can be checked using the template for spherical hip measurement [42] (► Chapter 3.2.3) or on a faux-profil x-ray.

! An important precondition for a triple osteotomy is the need for both the acetabulum and femoral head to be roughly spherical. If this is not the case, the femoral head and acetabulum must be swiveled by the same amount at the same time so that the aspherical congruence is maintained. On the other hand, if the head and acetabulum are spherical but with differing radii, acetabuloplasty is usually the better option.



■ Fig. 3.177a, b. 10-year old girl with DDH and inadequate coverage of the femoral head. a AP x-ray of the left hip. b Situation after *triple osteotomy* with outward rotation and lateralization of the acetabular fragment. Note that the caudal section is medialized, thereby shifting the pivot point to the center of the head and away from the ischium



The triple osteotomy can be performed on children from the age of 8 or on adults. The indications are similar to those for the periacetabular osteotomy, the main difference being that the triple osteotomy can also be implemented with an open triradiate cartilage, which is not the case with the periacetabular osteotomy. We only perform this operation when either definite hip-related and load-related symptoms are present or if the CE angle is less than 10°. However, the surgeon must carefully establish whether the symptoms are actually associated with poor acetabular coverage rather than an impingement problem (► Chapters 3.2.3 and 3.2.6). The latter can also occur after an incorrect reorientation of the acetabulum with a reduction in acetabular anteversion.

The specific technique used is of secondary importance. The techniques in which the pubis and ischium are divided close to the symphysis are less suitable these days [54, 79], as the pivot point for the swivel movement in such cases is too far from the hip. We use a modified surgical technique according to Steel [78]. Although the Tönnis technique [82] has the advantage of exposing the sciatic nerve via the dorsal approach to the ischium, the resulting scar over the buttocks is not esthetically appealing. Another drawback is the need to turn the patient during the operation.

The most important *complication* of the triple osteotomy is a sciatic nerve lesion. Fortunately, this is a rare event and the damage is usually transient. The sciatic nerve is at risk during the osteotomies of the ischium and ilium. In over 100 triple and periacetabular osteotomies we have only observed one transient lesion of the sciatic nerve. In theory, the femoral nerve (during the pubic osteotomy) and major vessels are also at risk. A case of premature closure of the triradiate cartilage has also been described [67].

Probably the most common complication is overcorrection or an incorrect orientation of the acetabulum. Thus, an excessive swiveling maneuver can lead to retroversion of the acetabulum instead of anteversion. Another dangerous situation can occur during lateralization of the acetabulum if the caudal part is not medialized. Incorrect positioning of the acetabulum can change the lever arms of the muscles, potentially resulting in permanent weakness of the abductors in particular.

Another possible complication is necrosis of the acetabulum. This risk applies particularly if the pubic osteotomy is performed too far laterally, since the vessels supplying the acetabulum from the obturator artery radiate into the acetabulum at the lateral margin of the pubic bone [4]. We have not observed this complication personally. Another rare event is pseudarthrosis, although no osteotomy is completely free of this risk. Another (rare) complication is the occurrence of periarticular

calcifications, which can obstruct movements at a later stage. The complication risks seem to increase with the age of the patient [37].

**Periacetabular osteotomy:** In the periacetabular osteotomy, the acetabulum is chiseled out without the complete division of all the bones (ilium, pubis, ischium). This method was first described by Blavier [7]. Wagner [89] modified the procedure to produce a »spherical acetabular osteotomy«, in which the acetabulum is chiseled out spherically approx. 1.5 cm above the cup. On the one hand, this operation is technically demanding while, on the other, the risk of avascular necrosis of the acetabular fragment is very great.

Ganz [30] described a periacetabular osteotomy in which the ilium and ischium are not completely divided, but the two cuts are linked by a dorsal osteotomy. This operation can be performed from the ventral side via a single incision. We have accumulated considerable experience with this operation. A precondition is closure of the triradiate cartilage, and the indications are otherwise similar to those for the triple osteotomy.

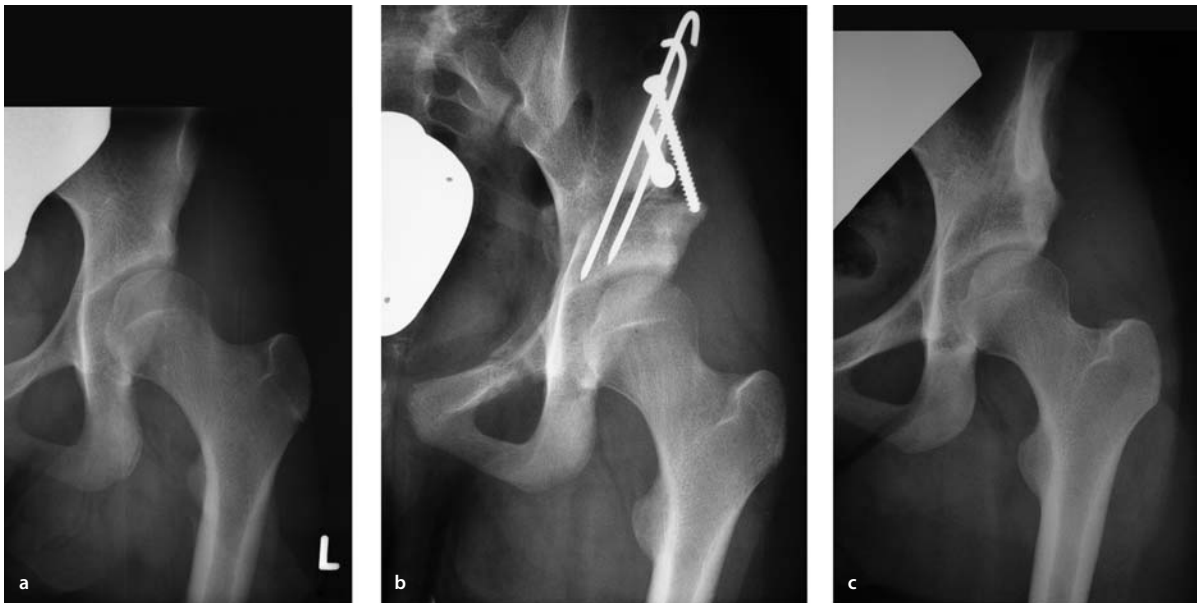
The advantages of the periacetabular osteotomy over the triple osteotomy:

- It can be performed via a single incision.
- Sacrospinal ligament not attached to the acetabular fragment, more options for reorientation.
- Better stability, since the pelvic ring is preserved intact.
- Less fixation required (2 screws), reduced risk of pseudarthrosis
- Risk of sciatic nerve lesion slightly less, since the ischium does not need to be divided completely.

A disadvantage is the slightly greater (theoretical) risk of avascular necrosis of the acetabular fragment, although we have not observed this complication in over 300 periacetabular osteotomies. For adult patients, we tend to perform the periacetabular osteotomy according to Ganz. Since the cut crosses the triadiate cartilage this procedure cannot be performed while the child is still growing. In view of the greater general mobility of the pelvis during adulthood, the sacrospinal ligament does not obstruct reorientation of the acetabular fragment as much. A clinical example is shown in

■ Fig. 3.178.

The *complication risks* associated with a periacetabular osteotomy are similar to those of the triple osteotomy. In 30 patients we measured the relevant loading area before and after periacetabular osteotomy using the template described in ► chapter 3.2. (■ Table 3.19). Preoperatively, the average area was 11.3 cm<sup>2</sup>, and postoperatively 15.6 cm<sup>2</sup>, corresponding to an improvement of 38%. Similar results based on computerized measurements have also been reported in the literature [20].



■ **Fig. 3.178a–c.** 14-year old girl with DDH and inadequate coverage of the femoral head. **a** AP x-ray of the left hip. The femoral head is also slightly lateralized and aspherical. **b** Situation after *periacetabular osteotomy according to Ganz* [30]. In this operation the ilium and ischium

are not completely divided, rather the osteotomy is performed in the bone around the acetabulum and the pelvic ring remains intact. The reorientation must be implemented very gradually while preserving the acetabular anteversion. **c** Two years after the operation

■ **Table 3.19.** Change in the relevant loading area produced by a periacetabular osteotomy in 30 patients. The measurements are recorded using the template for spherical measurement shown in Fig. 3.146. The area that is relevant to hip loading, marked in Fig. 3.149, is calculated

Change in the relevant loading area produced by a periacetabular osteotomy	
Average relevant area preoperatively	11.3 cm <sup>2</sup>
Average relevant area postoperatively	15.6 cm <sup>2</sup>
Improvement (absolute)	4.3 cm <sup>2</sup>
Improvement (percentage)	38%

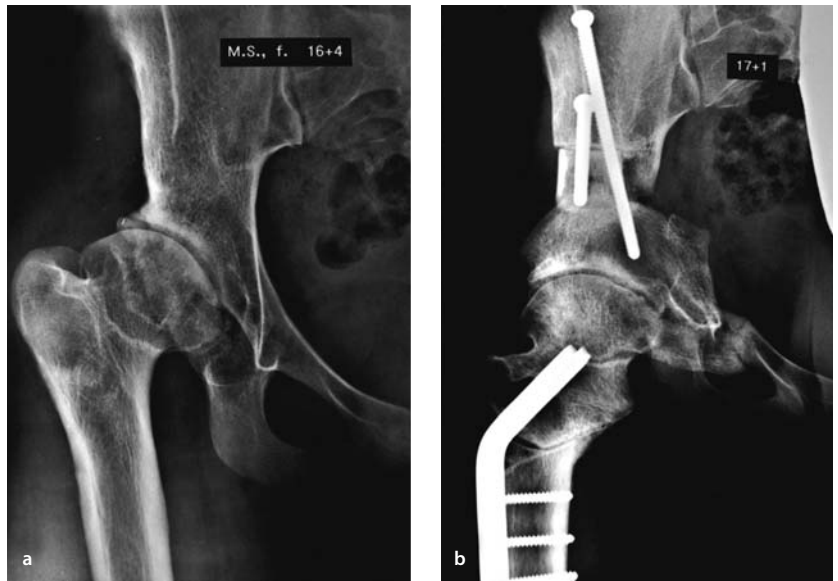
⚠ **The triple and periacetabular osteotomies should only be performed by experienced operators. The most difficult task is to assess the correct orientation of the acetabulum. A common mistake is to reduce the acetabular anteversion, potentially resulting in an impingement problem [61, 75]. Good long-term results have been reported by corresponding centers [36, 59, 74, 87].**

**Pelvic osteotomy according to Chiari:** This osteotomy was described by Chiari in 1955 [13]. The technique

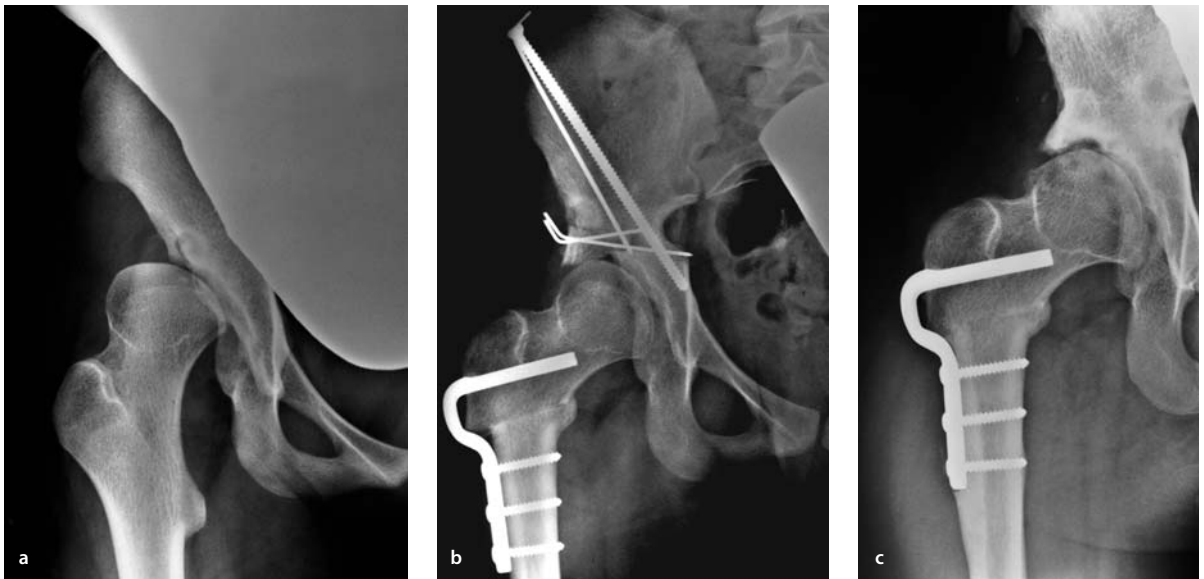
involves an oblique osteotomy of the ilium at the level of the lateral acetabular epiphysis, ascending upwards in the medial direction, and lateral displacement of the proximal section of the ilium over the femoral head. The disadvantage is that the new acetabular roof primarily consists of bone rather than hyaline cartilage. Moreover, the new acetabular roof is relatively small in the ventrodorsal plane. Before the triple and periacetabular osteotomies became popular procedures, the Chiari osteotomy was the only way of improving acetabular coverage in adult hips, particularly in cases where the roof was too short and not too steep (in the latter case acetabuloplasty was also available of course).

We consider that the Chiari osteotomy is almost never indicated nowadays. Even with an aspherical configuration, we prefer the combination of a periacetabular osteotomy with simultaneous intertrochanteric valgization (■ Fig. 3.179). Only for a very small aspherical acetabulum might the Chiari osteotomy still be justified, since it can increase the overall surface area of the acetabulum.

**Shelf operation:** Augmentation of the acetabulum by the insertion of bone grafts, the so-called »shelf operation«, is a common treatment in English-speaking countries [77]. A similar operation was described by Spitzzy as early as 1923 [76]. He wedged tibial grafts in a slot above the lateral acetabular rim. Nowadays, the shelf operation,



**Fig. 3.179a, b.** 16-year old girl with severe hip dysplasia, small acetabulum and oval femoral head after a avascular necrosis of the femoral head. **a** AP x-ray of the left hip. **b** In this situation, a periacetabular osteotomy was combined with an intertrochanteric valgus osteotomy with valgization to match the degree of swiveling of the acetabulum



**Fig. 3.180a–c.** 15 1/2-year old girl with severe hip dysplasia with high-standing, subluxating and slightly aspherical femoral head and very flat, steep acetabulum with a large radius. **a** AP x-ray of the right hip. **b** In this situation a Chiari pelvic osteotomy was combined with

an intertrochanteric varus osteotomy. The bone taken from the femur was used to augment the acetabular roof as a shelf. **c** 3 years later the autologous femoral graft is well integrated and forms a robust lateral epiphysis

like the Chiari osteotomy, is only considered as a stopgap measure when the acetabulum is much too small overall.

**Fig. 3.180** shows a combination of the shelf procedure and Chiari osteotomy, in which the graft taken from the femur was used to augment the acetabular roof, wedged against the laterally displaced upper section of the ilium.

***Our therapeutic strategy for congenital dislocation of the hip (ultrasound types III or IV according to Graf or radiological dislocation)***

Our therapeutic strategy for congenital dislocation of the hip is shown in **Table 3.20**.

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**Table 3.20.** Our therapeutic strategy for congenital dislocation of the hip (ultrasound types III or IV according to Graf or radiological dislocation)

Therapeutic strategy	
Age 1	Overhead traction – closed reduction (open reduction only if closed reduction not possible) – hip spica for 6 weeks – Tübingen splint for 3 months; poss. acetabular roof reconstruction (usually Salter osteotomy) at an older age (min. 15 months)
Age 2	
Femoral epiphyseal plate at the level of, or below, the triradiate cartilage	Overhead traction – closed reduction (open reduction only if closed reduction not possible) – hip spica for 6 weeks – Tübingen splint for 3–6 months; poss. acetabular roof reconstruction (usually Salter osteotomy)
Femoral epiphyseal plate above the triradiate cartilage («high dislocation»)	Longitudinal traction – open reduction – hip spica for 6 weeks – Tübingen splint for 3–6 months; poss. acetabular roof reconstruction (usually Salter osteotomy) and intertrochanteric shortening osteotomy (poss. with additional derotation)
Age 3–5	Longitudinal traction – open reduction – acetabular roof reconstruction (Salter osteotomy or acetabuloplasty), poss. intertrochanteric shortening (and derotation) osteotomy – hip spica for 6 weeks, poss. body cast for 12 weeks
Age 5–8	
Unilateral dislocation	Longitudinal traction – open reduction – acetabular roof reconstruction (Salter osteotomy or acetabuloplasty), intertrochanteric shortening (and derotation) osteotomy
Bilateral dislocation	Attempt reduction only if a secondary acetabulum has formed, in which case pain is highly likely to occur
From the age of 9	No reduction attempt (Fig. 3.167)

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### 3.2.5 Legg-Calvé-Perthes disease

» *The outlook in Perthes can be very grim, for a well-fed child that is far from trim.* «

#### ► Definition

Hip disease occurring during early childhood and caused by impaired circulation in the femoral head. The cause of the ischemia is not known.

■ *Synonyms:* Childhood aseptic necrosis of the femoral head, Perthes disease, osteochondritis deformans juvenilis

#### Historical background

The disease was described almost simultaneously, in 1910, by G. C. Perthes [53] in Germany, J. Calvé [8] in France and A.T. Legg [41] in America. The newly discovered x-ray technique allowed doctors to differentiate it from inflammatory forms of hip disease.

#### Etiology

While the etiology of Legg-Calvé-Perthes disease is not known, numerous studies have highlighted various factors that play a role in the development of the illness.

■ *Vascular supply:* Angiograms and laser Doppler flow measurements in patients with Legg-Calvé-Perthes disease have shown that the medial circumflex artery is missing or obliterated in many cases and that the obturator artery [16] or the lateral epiphyseal artery are also affected in some cases.

■ *Increased intra-articular pressure:* Animal experiments have shown that an ischemia similar to that in Legg-Calvé-Perthes disease can be generated by increasing the intra-articular pressure [72]. However, the condition of *transient synovitis of the hip* does not appear to be a precursor stage of Legg-Calvé-Perthes disease as the increased pressure resulting from the effusion in transient synovitis does not lead to vessel closure [63].

■ *Intraosseous pressure:* The measurement of intraosseous pressure in Perthes patients has shown that the venous drainage in the femoral head is impaired, causing an increase in intraosseous pressure [65]. In animal studies, the intraosseous injection of fluid, and the associated increase in pressure, produced a condition similar to Legg-Calvé-Perthes disease [65].

■ *Coagulation disorder:* One study found a coagulation disorder in 75% of 44 investigated children with Legg-Calvé-Perthes disease. In most cases the disorder was thrombophilia with an absence of factor C or S. Rarely the disorder involved elevated serum levels of lipoprotein, a thrombogenic substance [19]. More recent studies have questioned the significance of clotting factors as an etiological component [18, 22, 34].

■ *Growth hormones:* While earlier studies found reduced levels of the growth hormone somatomedin, recent studies have not shown any difference from control groups in respect of hormone status [34].

- Growth:** Children with Legg-Calvé-Perthes disease are shorter, on average, than their peers of the same age and show a retarded skeletal age (cartilaginous dysplasia) [9]. The maturation disorder occurs between the ages of 3 and 5 years [37]. Both the trunk and extremities lag behind in terms of growth. The shortening of the extremities is also accompanied by small feet [22]. Since this shortening is offset by excessive growth at a later age, patients who suffered from Legg-Calvé-Perthes disease as children are no shorter, as adults, than the population average [9, 73]. More recent experimental studies have shown that the metaphyseal changes are based on a growth disorder [44].
- Social conditions:** Studies in the UK have shown that Legg-Calvé-Perthes disease is more common in the lower social strata [45, 55]. The authors suggest a poorer diet during pregnancy as one possible explanation for this phenomenon. A recent study did not confirm this theory [66].
- Genetic factors:** Genetic studies have shown that first-degree relatives of children with Perthes disease are 35 times more likely to suffer from the condition than the normal population. Even second- and third-degree relatives show a fourfold increased risk [43]. The authors postulate a multifactorial inheritance. Legg-Calvé-Perthes disease also occurs in dogs (Manchester terriers). If two diseased dogs are crossed, all the male descendants will also suffer from the disease [71].

**! To sum up, genetic factors play an important role in the etiology of Legg-Calvé-Perthes disease. The illness develops as a result of impaired circulation in the medial circumflex artery in association with a skeletal maturation disorder with delayed growth in children aged from 3–5 years.**

### Occurrence

The annual incidence of Legg-Calvé-Perthes disease in the white population is 10.8 per 100,000 children and adolescents aged from 0–15 years, compared to 3.8 per 100,000 in Asians [62], 1.7 per 100,000 in mixed-race populations and 0.45 per 100,000 in blacks [57]. The highest reported incidence was for the city of Liverpool (UK) in the early

1980's, with 15.6 cases per 100,000 individuals under 15 years of age [3]. A decline was subsequently observed in the 1990's – possibly as a result of the improved social conditions [45]. A similarly high incidence (15.4 per 100,000) was recently reported in a rural area of Southwest Scotland [55]. In Sweden an annual incidence of 8.6 per 100,000 people under 15 years was determined [50].

**! Boys are four times more likely to be affected than girls.**

### Classification

All known classifications of Legg-Calvé-Perthes disease are based exclusively on the morphological findings on x-rays.

### Morphological classifications of the extent of the lesion

#### Classification according to Catterall

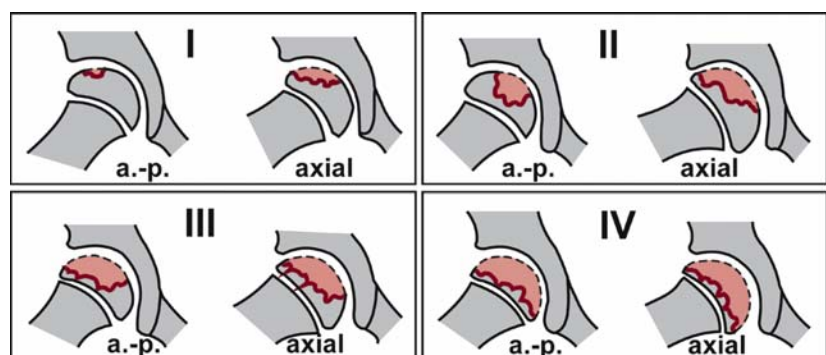
The classification proposed by Catterall in 1971 [10] divides the femoral head into 4 quadrants on AP and axial x-rays. The classification refers to the number of affected quadrants (■ Fig. 3.181, ■ Table 3.21). Catterall supplemented this classification with a number of »head at risk signs«; ■ Table 3.22).

#### Classification according to Salter and Thompson

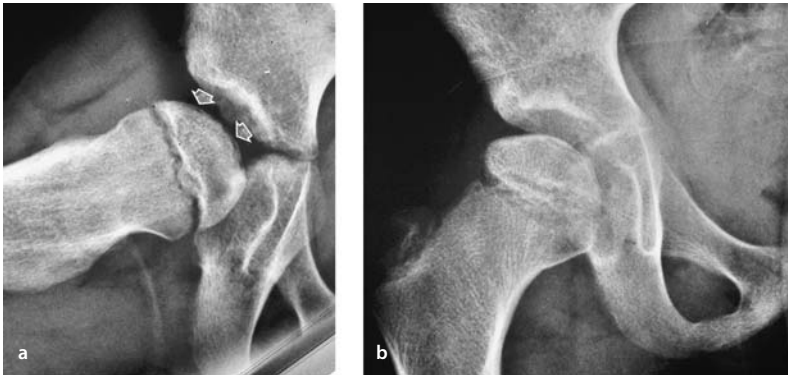
In 1984 Salter and Thompson [64] proposed a new classification that distinguished only two groups. The classification relates to the subchondral fracture that can be seen in the initial stages primarily on axial x-rays (■ Fig. 3.182, ■ Table 3.23).

■ Table 3.21. Classification of Legg-Calvé-Perthes disease according to Catterall [10]

Grade	Characteristics
I	Only anterolateral quadrant affected
II	Anterior third or half of the femoral head
III	Up to 3/4 of the femoral head affected, only the most dorsal section is intact
IV	Whole femoral head affected



■ Fig. 3.181. Classification of Legg-Calvé-Perthes disease according to Catterall: I only anterolateral section affected; II anterior third or half of the femoral head involved; III up to 3/4 of the femoral head affected, only the extreme dorsal part remains intact; IV whole femoral head affected



■ Fig. 3.182a, b. 8-year old boy with *subchondral fracture* and incipient Legg-Calvé-Perthes disease. a Axial x-ray, b AP x-ray

■ Table 3.22. Supplementary classification of Legg-Calvé-Perthes disease according Catterall [10] («head at risk signs»)

Lateral calcification	Appearance of a calcification shadow on the x-ray lateral to the femoral head
Subluxation	Lateral displacement of the head center
Metaphyseal involvement	Osteolytic foci in the area of the metaphysis bordering the epiphyseal plate
»Gage sign«	Triangular section of osteoporosis on the lateral femoral head
Transverse epiphyseal plate	Realignment of the epiphyseal plate along the horizontal plane

■ Table 3.23. Classification of Legg-Calvé-Perthes disease according to Salter & Thompson [56]

Group	Characteristics
A	Subchondral fracture involving <50% of the femoral dome
B	Subchondral fracture involving >50% of the femoral dome

The necrosis spreads out beneath this fracture and the extent of the subchondral osteolysis provides an indication of the subsequent spread of the necrosis (■ Fig. 3.183).

Group A roughly corresponds to the Catterall groups I and II, while group B is identical to Catterall groups III and IV.

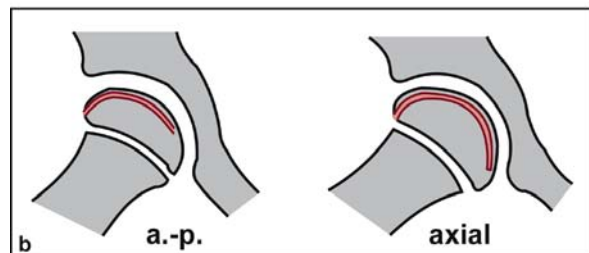
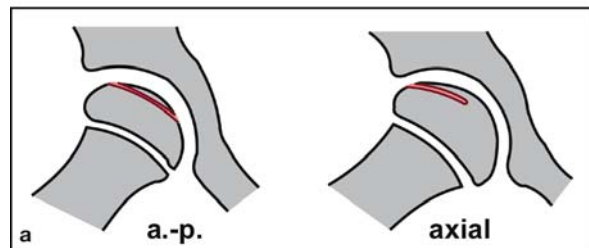
Salter and Thompson [64] believe that the inclusion of the subchondral fracture enables the condition to be classified much sooner than is possible with the Catterall classification. If no subchondral fracture occurs, bone resorption does not take place and the condition heals without any defect.

#### Classification according to Herring («lateral pillar classification»)

In 1992 Herring et al. [26] proposed a classification based on the morphology of the lateral pillar of the femoral head on the AP x-ray (■ Fig. 3.184, ■ Table 3.24).

■ Table 3.24. Classification of Legg-Calvé-Perthes disease according to Herring et al. [23]

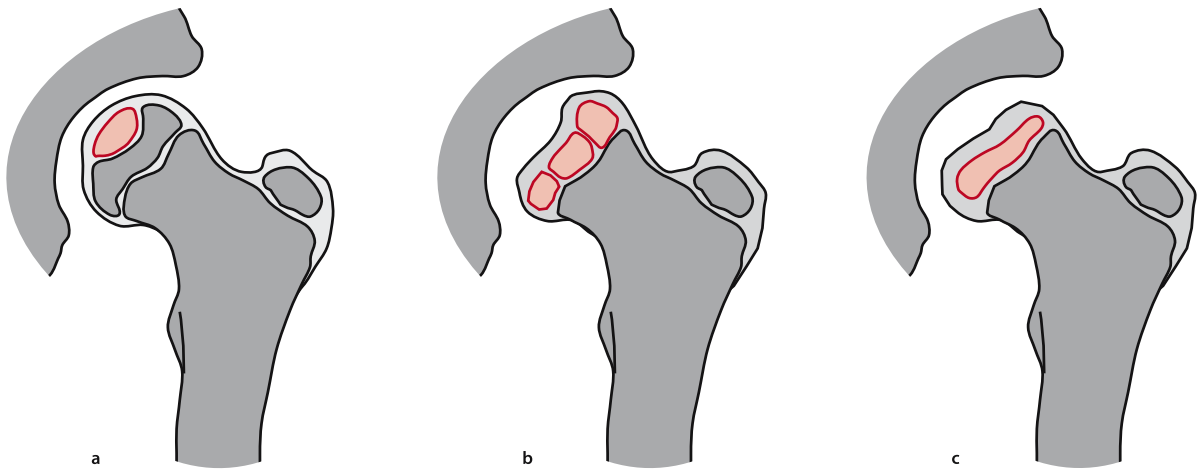
Group	Characteristics
A	Lateral pillar not affected
B	>50% of the height of the lateral pillar preserved
C	<50% of the height of the lateral pillar preserved



■ Fig. 3.183a, b. Classification according to Salter and Thompson. a Subchondral fracture affecting <50% of the femoral dome (corresponding to Catterall groups I and II). b Subchondral fracture affecting >50% of the femoral dome (corresponding to Catterall groups III and IV)

#### Morphological classification of progression

The various *progression stages* of Perthes disease are listed in ■ Table 3.25 (■ Fig. 3.185). The period from the onset of the illness to the end stage invariably lasts several years. The older the child at the onset of the disease, the longer the individual stages will last. The youngest onset age reported is 24 months [19]. The recurrence of the disease in the same hip after several years has been observed in isolated cases [20, 68].



**Fig. 3.184a–c.** Classification of Legg-Calvé-Perthes disease according to Herring (lateral pillar classification). a Lateral pillar not affected. b >50% of the height of the lateral pillar preserved. c <50% of the height of the lateral pillar preserved



**Fig. 3.185a–d.** Radiographic progression stages of Legg-Calvé-Perthes disease. a Boy aged 3 years, femoral head in the condensation stage; b at the age of 5 years in the fragmentation stage; c at the age of 7 years in the repair stage; d at the age of 9 years in the end stage

**Table 3.25. Morphological classification of the progression of Legg-Calvé-Perthes disease**

Stage	Characteristics
Condensation stage	The femoral head appears slightly denser than normal on the x-ray and is slightly flattened, the joint space is widened
Fragmentation stage	Femoral head breaks up into fragments (depending on the extent of the lesion in each case) with osteolytic and sclerotic areas
Repair stage	The femoral head is rebuilt
End stage	End stage with or without defect healing (normal hip, coxa magna, coxa parva, flattened head etc.)

**Morphological classification of the end stage**  
**Classification according to Stulberg**

In 1981 Stulberg et al. [69] suggested a classification of the end stage of Legg-Calvé-Perthes disease that is still widely accepted (Table 3.26). The study by Stulberg et al. [69] is based on 99 hips of individuals with the end stage of Legg-Calvé-Perthes disease. The appearance of the femoral head in the end stage correlated closely with the age of the patient at the onset of the disease. The risk of osteoarthritis increased from groups I through V. Interestingly, it was not the patients in Catterall group IV (whole femoral head affected) who showed the highest risk of osteoarthritis, but the patients in group III. This finding is explained by the fact that the flattening is less marked when the whole head is affected than if a part of



## 3.2.5 · Legg-Calvé-Perthes disease

the head remains intact. The characteristic features of the patient groups for the 99 hips in the study by Stulberg et al. [69] are shown in ■ Table 3.27.

### Other classifications

Various authors have attempted to produce classifications based on MRI [60] or bone scans [12, 70], although none has yet gained widespread acceptance.

### Prognosis and evaluation of classifications in respect of prognosis

Numerous studies [26, 30, 48, 53, 74] have investigated the prognostic significance of the various classifications. Some of these studies are based on observation periods of more than 30 years [30, 48, 74]. The prognostic significance of the individual clinical and radiological parameters is summarized in ■ Table 3.28. All authors agree that *age* is clearly the most important prognostic factor. The older the patient, the worse the prognosis.

A certain »magical limit« applies at 6 years. If the disease starts before this age the prognosis, disregarding all other factors, is fairly good, whereas a poorer end result can be expected if the onset of the disease occurs at a higher age. Since we know that children with Legg-Calvé-Perthes disease show delayed skeletal maturation, one would really need to include the skeletal age in the evaluation, although no reliable data is currently available on this point.

Other findings with substantial prognostic significance include the occurrence of the *lateral calcification shadow* and the degree of *subluxation* (■ Fig. 3.186) or *containment*. These were first identified by Catterall [10], who included these parameters as risk factors. All long-

term studies have since confirmed this assessment [30, 48, 53, 74]. Both factors are evidence of a deterioration in containment. The femoral head starts to develop outside the acetabulum, and the rim of the latter produces a depression in the femoral head, disrupting the normal curvature. A non-spherical femoral head is clearly more at risk of osteoarthritis than a round head [69].

*Mobility* is a very significant factor for the prognosis, although few authors have referred to this fact [64]. In our experience, patients with considerably restricted range of motion, regardless of age, will show much worse progression and a marked tendency toward subluxation in view of the shortening of the adductors.

Legg-Calvé-Perthes disease affects boys four times more frequently than girls. *Sex* also has a relatively high prognostic value since the late results in girls are much worse than in boys [51].

The *extent of necrosis* is only moderately significant in terms of the late prognosis, even though it is the determining parameter in the classifications of Catterall and Salter & Thompson. The only valid conclusion in respect of the classification according to Catterall is that patients in group I (only anterolateral section affected) always

■ Table 3.26. Classification of Legg-Calvé-Perthes disease according to Stulberg et al. [60]

Grade	Characteristics
I	Round head, normal hip
II	Round head, coxa magna
III	Oval or mushroom-shaped head, coxa magna
IV	Flat head, congruent with acetabulum
V	Flat head, incongruent

■ Table 3.28. Prognostic factors for Legg-Calvé-Perthes disease

Prognostic factor	Prognostic significance
Age	+++
Shape of lateral pillar <sup>1</sup>	+++
Lateral calcification <sup>2</sup>	++
Subluxation <sup>2</sup>	++
Mobility	++
Sex	++
Extent of necrosis <sup>3</sup>	+
Metaphyseal involvement <sup>2</sup>	+
Gage sign <sup>2</sup>	-
Transverse epiphyseal plate <sup>2</sup>	-

<sup>1</sup> Risk factor according to Herring classification

<sup>2</sup> Risk factors according to Catterall / »head at risk-signs«

<sup>3</sup> Classification according to Catterall / Salter & Thompson

■ Table 3.27. Characteristics of patient groups for 99 hips in the study by Stulberg et al. [60]

Stulberg group	Age at onset of Legg-Calvé-Perthes disease (years)	Predominant Catterall group	Patients with incipient osteoarthritis (%)
I	6.0	I–II	0
II	6.4	II–IV	16
III	8.2	III–IV	58
IV	8.3	III–IV	75
V	10.4	III	80



have a good prognosis regardless of age. No prognostic statements can be made, however, about groups II–IV [30, 48, 74]. Patients in group IV tend to have a better prognosis than those in group III, since a non-spherical head is less likely to occur if the whole femoral head is necrosed than if one section is preserved [69]. Otherwise, it is extremely difficult to make any distinction in terms of prognosis between groups II and III.

The classification according to Salter & Thompson merges Catterall groups I and II (A) and III and IV (B). Although the creators of this classification state that groups A and B differ distinctly in terms of prognosis [64], it may be concluded from the above statements that the boundary should rather be drawn between the Catterall groups I and II, rather than II and III. Furthermore, since the subchondral fracture, which must be detected for the classification according to Salter & Thompson, is only visible on the x-ray for a relatively short period, this classification cannot even be used for many patients (since no corresponding x-ray exists). If such images are available, however, this classification is much easier to use than that of Catterall [16, 19, 61, 64].

The classification according to Herring does not take into account the overall extent of the necrosis, but only the involvement of the lateral pillar. Since this particular section has a substantial influence on subsequent containment, the prognostic value of this classification is rather better than that of the other two [16, 61], even if the classification requires further correction during the first 6

months of the disease in a third of cases [40]. A recently published prospective multicenter study with 438 patients with 451 affected hips made clear statements concerning the outcome of treatment. The lateral pillar classification and age at the time of onset of the disease strongly correlated with outcome. Patients who were over the age of 8 years at the time of onset and had a hip in the lateral pillar B group or B/C border group had a better outcome with surgical treatment than they did with nonoperative treatment. Group-B hips in children who were less than 8 years of age at the time of onset had very favorable outcomes unrelated to treatment, whereas group C hips in children of all ages frequently had poor outcomes, which also appear to be unrelated to treatment [27].

*Metaphyseal involvement* has only slight prognostic significance [51, 74]. Moreover, the extent of epiphyseal involvement is extremely difficult to assess on conventional x-rays since the peripheral sections of the widened femoral head project over the metaphysis on both the AP and axial views, potentially causing what are in fact epiphyseal processes to be misinterpreted as changes in the metaphysis [28].

Long-term studies [30, 51, 74] have shown that the »head at risk signs« cited by Catterall in 1971 as risk factors, i.e. the »*gage sign*« and a *transverse epiphyseal plate* do not have any prognostic significance.

**! Factors with the greatest prognostic significance are age, subluxation, lateral calcification, mobility and sex. Although it has not been studied much, overweight does have a strongly negative effect on progression, particularly in elderly children. Unfortunately, all these factors are ignored in the standard classifications. Since the classifications do not provide information that is relevant either to prognosis or the treatment to be administered, their usefulness in everyday clinical practice is restricted. They all have a relatively poor inter- and intra-observer reliability [33]. They are helpful, however, for an understanding of the disease. Herring's classification probably provides the best prognostic information [27].**



**Fig. 3.186.** *Head-at-risk signs* in Legg-Calvé-Perthes disease: lateral calcification, metaphyseal involvement, subluxation

### Diagnosis

Children with Legg-Calvé-Perthes disease limp and complain of mild to moderate hip pain. This situation can persist for several weeks. Clinical examination usually reveals a slight stiff, protective limp. The range of motion of the affected hip is usually significantly restricted, in particular with reduced abduction and internal rotation.

### Radiographic findings

The initial signs on a *plain x-ray* are slight flattening of the femoral head and slight condensation of the bone structure« and widening of the joint space (Fig. 3.187). The latter can best be evaluated on the basis of the dis-

### 3.2.5 · Legg-Calvé-Perthes disease

tance of the femoral head from Koehler's radiographic teardrop. These changes generally occur four weeks, at the earliest, after the start of the history. At this stage, an axial view (Lauenstein view; ► chapter 3.2.2) should always be recorded in addition to the AP x-ray as the subchondral fracture is almost always visible on this view at this phase of the illness. The extent of this osteolysis has prognostic significance (see above).

**! We do not consider other imaging procedures to be useful in the initial stages.**

A case of Legg-Calvé-Perthes disease can be diagnosed several weeks earlier with bone or MRI scans than with conventional radiographs (the latter especially in combination with gadolinium [19]). However, since the early diagnosis does not have any therapeutic consequences the costs of these imaging procedures can safely be avoided.

*Ultrasound examination* is useful for follow-up checks as they can show the protrusion of the (cartilaginous/bony) femoral head laterally and/or ventrally (■ Fig. 3.188).



■ Fig. 3.187. X-ray of 5-year old boy in the *early stage* of Legg-Calvé-Perthes disease: Note the widening of the joint space (visible particularly over Koehler's radiographic teardrop) and the slight flattening and condensation of the femoral head compared to the healthy side

### Differential diagnosis

Circulatory problems in the femoral head can occur in a range of illnesses, or else other changes with a similar radiological appearance may be present. These lesions will need to be differentiated from Legg-Calvé-Perthes disease as some of them may have a different prognosis and thus require different treatment.

### Epiphyseal dysplasia

Epiphyseal dysplasia is a disorder of the bony structure of the epiphysis. It is a hereditary condition that occurs in two forms: as multiple epiphyseal dysplasia and as spondyloepiphyseal dysplasia (with involvement of the spine; ► chapter 3.2.7). The femoral head findings can appear very similar to those in Legg-Calvé-Perthes disease.

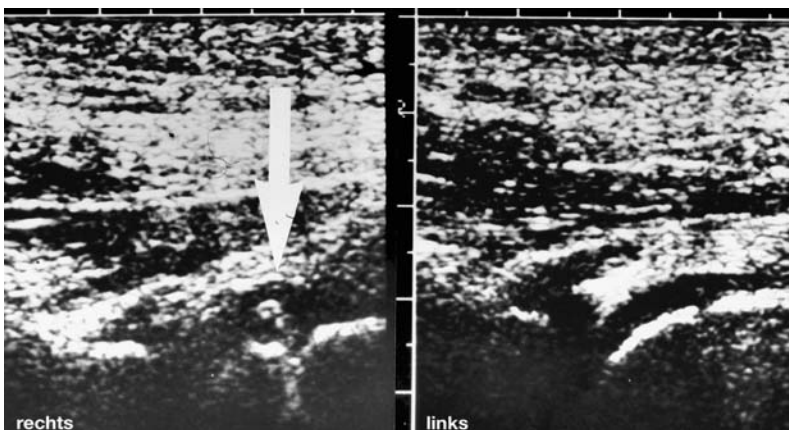
*The following are diagnostic indications of epiphyseal dysplasia:*

- bilateral involvement,
- largely symmetrical findings,
- poss. involvement of other joints or the spine (in this case platyspondylia),
- poss. involvement of the acetabulum,
- few sclerotic or cystic changes in the femoral head,
- little tendency toward lateral calcification or subluxation.

*The following are more typical of Legg-Calvé-Perthes disease:*

- unilateral involvement,
- if bilateral involvement is present: pronounced asymmetry, disease in differing stages, poss. also of differing severity,
- no involvement of other joints or the spine,
- no involvement of the acetabulum,
- sclerotic and cystic changes in the femoral head,
- cystic changes in the metaphysis,
- tendency toward lateral calcification and subluxation.

Sometimes it is not possible to make a clear distinction between the two diseases. Epiphyseal dysplasia likewise



■ Fig. 3.188. *Ultrasound scan* showing protrusion of the right femoral head in Legg-Calvé-Perthes disease (compared to the left side)

affects boys four times more frequently than girls [59]. The distinction is important as the mid-term prognosis for epiphyseal dysplasia is much better than for Legg-Calvé-Perthes disease. The orthopaedist therefore has to be more reserved in deciding whether therapeutic measures are indicated. Treatment is almost *never* required for epiphyseal dysplasia.

### Other disorders associated with Legg-Calvé-Perthes disease

Avascular necrosis of the femoral head similar to that in Legg-Calvé-Perthes disease occurs with increased frequency in the following illnesses:

- sickle cell anemia [1],
- thalassemia is associated with an extremely high incidence (25%) of avascular femoral head necrosis [1],
- trichorhinophalangeal syndrome [29],
- Klinefelter syndrome [23],
- Morquio syndrome [23],
- Down syndrome (trisomy 21) [23],
- achondroplasia [23],
- Gaucher disease [23],
- myelomeningocele [23],
- hemophilia [23] (the incidence in hemophilia is 7% [54]),
- congenital tibial pseudarthrosis [23].

Since the prognosis for the avascular necrosis that occurs in the above disorders is no different from that of Legg-Calvé-Perthes disease, it is treated according to the same guidelines.

### Osteochondrosis dissecans

This condition is extremely rare in the hip (in contrast with the knee). The osteochondrotic focus is always located in the femoral head (never in the acetabulum). As a rule, it is readily viewed on standard imaging procedures.

### Tumors

An important differential diagnosis in relation to Legg-Calvé-Perthes disease is a *chondroblastoma* (▶ Chapter 3.2.13, ■ Fig. 3.246).

This tumor occurs predominantly in children. It is usually located exclusively in the epiphysis and can induce changes in the femoral head that can be confused with Legg-Calvé-Perthes disease. In a case of chondroblastoma, however, the height of the femoral head is not initially reduced, nor is the cartilage thickened. Moreover, the presence of non-load-related pain should indicate the possibility of a tumor.

*Metaphyseal tumors* or *cysts* can occur secondarily to a vascular occlusion and cause femoral head necrosis. Depending on the age at onset, the course of the disease can closely resemble that of Legg-Calvé-Perthes disease. Lymphomas can also cause femoral head necroses.

### Inflammatory disorders

❗ **Septic arthritis of the hip almost always results in femoral head necrosis if it is not treated adequately at a sufficiently early stage (i.e. within 4 days). In contrast with Legg-Calvé-Perthes disease, however, cartilaginous damage with narrowing of the joint space is invariably present as well.**

### Trauma

Posttraumatic femoral head necrosis can occur at any age, and particularly after femoral neck fractures, although we have also experienced cases in which this complication has occurred after an avulsion fracture of the greater trochanter (as a result of tearing of the circumflex femoral artery). The unambiguous history makes the differential diagnosis very easy in such cases.

### Drug-induced femoral head necrosis

Children and adolescents undergoing chemotherapy or who receive long-term cortisone treatment for malignant tumors or severe juvenile rheumatoid arthritis are at risk of suffering a femoral head necrosis. This can affect up to 5% of patients with leukemia [58]. Fortunately, we have not encountered the alcohol-induced form, which frequently occurs in adults, in our adolescent patients.

### Treatment

The therapeutic measures for Legg-Calvé-Perthes disease are primarily designed to pursue *three objectives*:

- *improved mobility,*
- *weight relief,*
- *improved containment.*

No reliable findings exist on the options for the *drug-induced improvement of the circulation* in the femoral head.

### Improved mobility

The efficacy of measures for improving the mobility of the hip in Legg-Calvé-Perthes disease is undisputed. Restricted movement is one of the important risk factors for the subsequent course of the disease and the end result. The primarily restricted movement is *abduction*, while *internal rotation* may also be diminished. The ability to abduct is particularly impaired in the presence of lateral calcification and increasing subluxation. If the femoral head loses the ability to slide smoothly in the acetabulum, a hinge abduction can develop. This is especially the case if the acetabulum has gouged an indentation in the femoral head, causing the head to protrude beyond the acetabulum as a result of the lateral calcification.

Mobility can be improved by the following measures:

- physical therapy,
- injection of botulinum toxin in the adductors,
- (hydraulic) mobilization under anesthesia,
- extension of the adductors,
- creation of an extension in increasing abduction.

Preserving mobility is an extremely important therapeutic objective in Legg-Calvé-Perthes disease. As soon as a restriction occurs, regular *physiotherapy* should be initiated as this measure is generally sufficient for preserving adequate mobility. In some cases, the physiotherapy will need to be continued for several years.

The preservation of mobility is also the basic requirement for measures designed to improve containment. If a hinge joint has already formed, then an intertrochanteric varization or pelvic osteotomy is not indicated. In many cases, however, appropriate mobilizing measures can restore joint mobility to some extent.

We inject *botulinum toxin* primarily into the *adductors*. In most patients this produces significant pain reduction and improved abduction performance [4], which indicates that muscle spasms of the adductors are mainly responsible for the pain. The methods of administering the botulinum toxin (Botox) injection are described in ► chapter 4.7. This drug has primarily been used in the past in spastic patients with infantile cerebral palsy. If correctly administered, the risks associated with this treatment are extremely low.

If this measure on its own does not achieve the objective, then we will attempt a *hydraulic mobilization under anesthesia*. For this purpose we use an arthroscope that is inserted with the hip in traction on the traction table. The joint is filled with Ringer lactate solution under great pressure with the aim of stretching the shrunken joint capsule. If the arthroscope cannot be inserted into the narrow joint, the hydraulic mobilization can alternatively be performed with a wide-bore cannula. The correct intra-articular position should be checked under the image intensifier using a contrast medium (Jopamiro). Joint mobility can also subsequently be checked by this method.

If it has proved possible to restore genuine abduction, we apply a *Petrie cast* in a position of maximum abduction (■ Fig. 3.189). Effective *epidural anesthesia* administered for several days via a catheter provides adequate analgesia, thus ensuring that the mobility achieved can be maintained with physical therapy. Only when the mobility has been maintained for a fairly prolonged period may containment-improving measures, e.g. intertrochanteric varization or pelvic osteotomy, be implemented.

If no genuine abduction is achieved, i.e. if the hinge abduction remains, then the usual surgical procedures for improving containment are no longer appropriate.



■ Fig. 3.189. Pretreatment prior to osteotomy in cases of poor mobility: Hydraulic mobilization of the hips under anesthesia, Petrie cast in maximum abduction, physical therapy under epidural anesthesia

However, the surgeon can attempt to improve the situation with a resection of the laterally developing protrusion (»bump resection«) or with a valgization osteotomy.

### Weight relief

The concept of weight relief in the treatment of Legg-Calvé-Perthes disease is highly controversial. The load on the hip can basically be relieved by the following methods:

- bedrest,
- wheelchair,
- walking with crutches,
- bracing devices (Thomas splint, Mainz orthosis, etc.).

The principle of weight relief is based on the idea that the necrotic femoral head is soft during the regrowth phase and therefore must be protected from collapsing by taking the weight off the hip. Considerable doubt exists, however, as to whether the femoral head really is soft. Whether the head is widened in Legg-Calvé-Perthes disease depends not on the fact that the »soft« head expands widthways, but that the necrotic bone must be replaced by new bone and that this regrowth occurs concurrently with the breakdown of the necrotic tissue. The bone therefore has to be replaced outside the necrotic area [64]. Necrotic bone is not soft, but the replacement bone has to form outside the existing (necrotic) femoral head and this is what gives the impression of the femoral head »flowing outwards« on the x-ray.

The regrowth of the femoral head takes years, with a minimum of 2 years. If the concept of weight relief is to be observed, then this must be continued consistently for at least 2 years.

*Bedrest* is certainly an effective way of taking the weight off the hip. Until recently, and in Eastern Europe in particular, Legg-Calvé-Perthes disease had been treated by consistent bedrest, with the affected children having to spend several years in specialist hospitals. While there is no scientific confirmation of the efficacy of this treat-



ment, anyone can readily imagine the adverse psychological effects of such a treatment. A *wheelchair* has similar effects and side-effects.

Doubt also exists about the relieving effect of *weight-bearing braces* [36]. At best these act as a hindrance to children and thus help prevent load peaks. But a brace treatment also has to be continued consistently for at least 2 years. Although children under 10 appear to tolerate a treatment with a brace better than during puberty, negative psychological effects can still be expected, as has been demonstrated in one psychological study [56]. Since considerable doubt also exists about its efficacy, we consider that such a treatment is *not indicated*. Even when used to supplement surgical treatment, no efficacy was observed for the weight-bearing brace [6].

Long-term *weight-bearing with crutches* is also not without its problems and, in addition to the psychological side effects, can also have adverse effects on the upper extremities. We therefore only consider this measure to be indicated as a stopgap solution for a limited period, during phases with severe pain and restricted mobility. It should be borne in mind, however, that this can promote an adduction contracture of the affected hip, particularly if the footwear is not adequately elevated on the other (healthy) side. One comparative study [13] has also shown that the use of weight-bearing crutches as a treatment for Legg-Calvé-Perthes disease is not sufficiently effective.

Since we do not prescribe weight-relieving treatment, many parents ask whether the child is allowed to take part in *sports*. In most cases the patients are, after all, small children who are not yet involved in competitive sports. There is no objection to gentle sporting activity in which movement, rather than weight-bearing, predominates (romping around, swimming, cycling). Load peaks should obviously be avoided (no jumping exercises, no apparatus-based gymnastics or contact sport). We believe that a complete ban on sport for several years is absurd since, on the one hand, movement is good for the joint and, on the other, the child should not be kept in social isolation.

### Improving containment

In addition to the preservation of mobility, the maintenance or restoration of joint containment is the most important therapeutic principle in Legg-Calvé-Perthes disease. Containment simply refers to the fact that the acetabulum »contains« the femoral head, i.e. the femoral head should be located completely within, and not lie partly outside, the acetabulum.

The following options are available for *improving containment*:

- Conservative: *abducting braces*
- Surgical: *intertrochanteric osteotomies*
- Surgical: *pelvic osteotomies*

Abduction causes the lateral part of the femoral head to rotate under the acetabulum. The same effect is produced with an intertrochanteric varus osteotomy or, by the reverse principle, with a pelvic osteotomy according to Salter or a triple osteotomy: In the latter case the acetabulum is rotated over the lateral section of the femoral head. In addition, the acetabulum can also be rotated ventrally to improve coverage of the ventral sections of the head. The intertrochanteric osteotomy must be accompanied by extension to achieve the same effect.

The containment treatment must be continued until the femoral head has regrown, i.e. for a minimum of 2 years.

### Conservative treatment: abducting braces

Abducting braces can be designed according to the following principles:

- braces with a hip strap and thigh sections held in an abducted position by hinges (e.g. »Atlanta brace«),
- braces with knee sections linked together and held apart by a bar (e.g. »Berne brace«),
- shoes permanently fixed to a bar and held apart at a fixed distance (e.g. »Ponseti Foot Abduction Bar«).

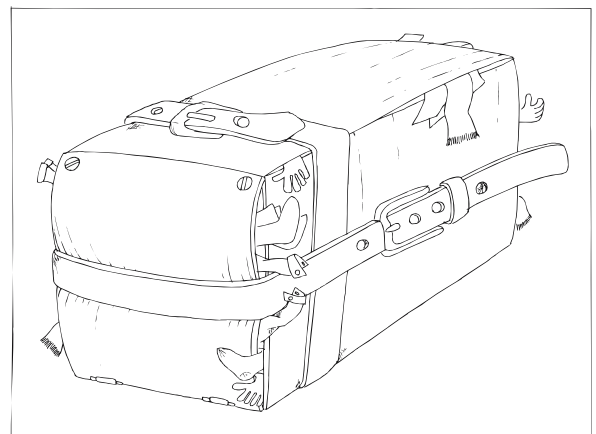
All these braces hinder children considerably. While the abducting effect is achieved with the first two types, walking with these braces is only possible with flexed knees. With the Ponseti Foot Abduction Bar, on the other hand, walking is almost impossible. Moreover, the child can avoid the abducting effect by sitting with flexed knees.

### Surgical treatment: femoral or pelvic osteotomy

The result produced with the abduction brace, namely improved containment, can essentially be achieved by surgical means.

### Advantages of conservative treatment

- no hospitalization required
- no operation risks



Good containment is present if nothing spills out of the container



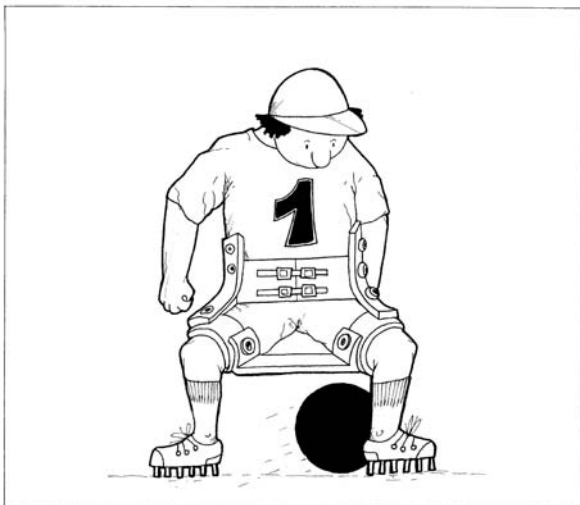
**Disadvantages of conservative treatment**

- much longer duration of impairment (2 years instead of 6 weeks)
- negative psychological effect

**! Even though one automatically tends to think that surgical treatment is more aggressive than conservative treatment, we do not share this view in this particular case. The negative psychological impact on a child resulting from a substantially handicapping 2-year brace treatment can, in our view, be much worse than that associated with a surgical treatment involving a 5-day hospital stay followed by 6 weeks of using crutches.**

Containment can be improved by surgery to the femur or the pelvis. In certain cases (particularly older children) an operation involving both the femur and pelvis may be advisable [14].

The intertrochanteric varus osteotomy (■ Fig. 3.190) produces the same effect as permanent abduction of the leg, while the Salter osteotomy produces an effect that would be achieved by inclining the upper body, and thus



Abducting braces hinder children considerably...

the pelvis, towards the affected side. In both cases the lateral section of the femoral head is contained in the acetabulum, while the Salter pelvic osteotomy additionally provides better coverage for the anterior section of the femoral head. This effect can be achieved on the femoral side by adding an extending component to the varization. The triple osteotomy (■ Fig. 3.191 and 3.192) produces the same effect as the Salter pelvic osteotomy. The former is indicated if the pelvis is not elastic enough to allow sufficient movement of the acetabulum. This generally applies from the age of eight.

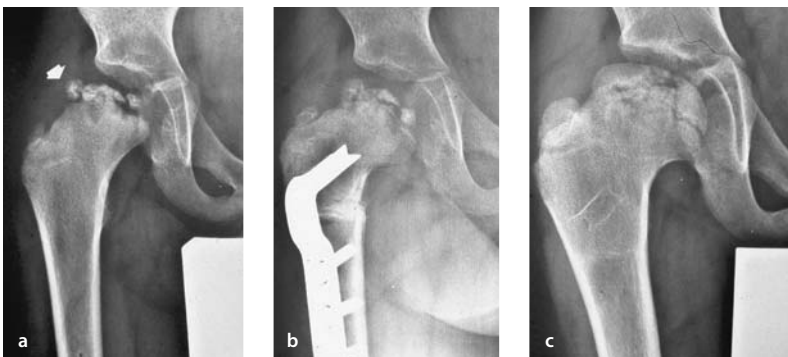
**Advantages of intertrochanteric osteotomy compared to pelvic osteotomy**

- Reduced surgical risk (compared to the risk of sciatic nerve injury, particularly with the triple osteotomy)
- No intra-articular increase in pressure (in contrast with the Salter osteotomy, but not with the triple osteotomy)

**Advantages of the pelvic osteotomy over the intertrochanteric osteotomy**

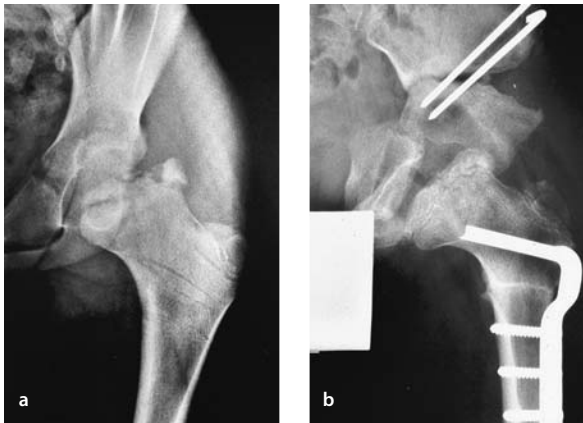
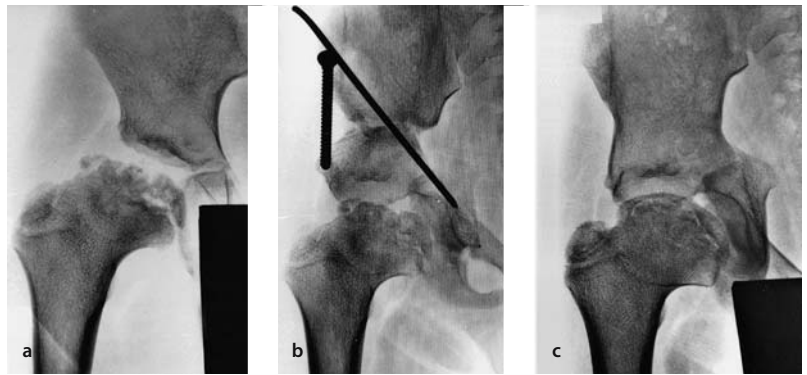
- No shortening of the leg
- No change in the lever arm of the abductors
- No verticalization of the epiphyseal plate
- Reduced risk of an adduction contracture

The Salter pelvic osteotomy results in increased pressure in the joint, since the acetabular roof is transferred not just laterally and anteriorly, but distally as well. The psoas muscle is primarily responsible for this increase in pressure, and this effect can be reduced by aponeurotic lengthening of the psoas tendon. We achieve the best outcome with the triple osteotomy. It avoids the problems inherent in the intertrochanteric varus osteotomy, does not increase the intraarticular pressure (in contrast with the Salter osteotomy), but is technically more demanding.



■ Fig. 3.190a–c. Improving containment by intertrochanteric osteotomy in a 6-year old boy with lateral calcification and subluxation. a Preoperatively, b postoperatively, c 2 years after operation

**Fig. 3.191a–c.** Improving containment by triple osteotomy in a 9-year old boy with lateral calcification and subluxation. **a** Preoperatively, **b** postoperatively, **c** 2 years after operation



**Fig. 3.192a, b.** Improving containment in a case of pronounced subluxation and deformation of the femoral head by concurrent triple osteotomy and intertrochanteric varization osteotomy in an 8-year old boy. **a** Preoperatively, **b** postoperatively

### Does the containment treatment improve the prognosis compared to spontaneous progression?

This question is answered differently in the literature. Two particular studies on the abduction brace that should be taken very seriously produce a negative answer to this question [46, 49]. Appropriately indicated surgical treatment, on the other hand, does appear to improve the child's prognosis significantly, compared to the untreated state, as has been demonstrated by studies involving age-matched groups of patients with conditions of comparable severity [11, 31, 32, 52]. However, some studies have also found that abducting braces can be just as effective as surgical treatment [7, 13, 17]. The results were better than spontaneous progression primarily in children over 5 years, and only the anterolateral section of the femoral head was affected (Catterall group I) [11]. The additional relief provided after surgical treatment does not provide any further improvement in the result [6]. In the recently published, aforementioned prospective study, based on the lateral pillar classification only those patients who were over the age of 8 years at the time of onset with a

hip in the lateral pillar B group or B/C border group had a better outcome with containment treatment than they did without it. Children who were less than 8 years of age at the time of onset and were not group C had favorable outcomes unrelated to treatment, whereas group C hips in children of all ages frequently had poor outcomes, whether or not they were treated [27].

No consensus exists about the type of surgical treatment. As already mentioned, one can achieve essentially same result produced by the intertrochanteric osteotomy, in terms of containment, with the pelvic osteotomy according to Salter and the triple osteotomy. In recent years, because of the aforementioned disadvantages, there has been a clear trend away from the intertrochanteric osteotomy towards the triple osteotomy [38]. We ourselves use the latter as the standard containment procedure in children over 7 years of age. For younger patients we perform the intertrochanteric varus osteotomy or the Salter pelvic osteotomy. The latter is particularly useful if the femoral head epiphyseal plate is rather steep or if leg shortening is already present. The overgrowth of the greater trochanter and a Trendelenburg limp that persists for a prolonged period are observed more frequently after an intertrochanteric osteotomy than after pelvic osteotomies [42, 67].

The *overall mobility* of the hip prior to an osteotomy must be good since it is not improved as a result of the operation and because the femoral head will not be centered properly during the procedure if the hip is not sufficiently mobile. Nowadays, botulinum toxin injection and/or postoperative epidural anesthesia left in place after mobilization for several days are two very efficient ways of improving the range of motion. The inability to abduct properly, particularly after a varus osteotomy, involves the risk of a postoperative adduction contracture with further decentering of the hip. Consequently, the mobility should not be allowed to fall below a critical limit before the operation.

### Follow-up controls

Irrespective of the treatment:

- Clinical check-ups every 3 months (particularly to examine mobility), possibly also ultrasound examination

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- Control AP and axial x-rays every 6 months for 2 years
- Subsequently (depending on the findings in each case), annual AP x-rays until the condition has returned to normal (less frequently thereafter). A further AP x-ray on completion of growth.

#### Drug-induced improvement of femoral head circulation

The drug iloprost appears to produce a positive effect on bone marrow edema and the vascularization of bone [2], although no controlled studies have yet demonstrated its efficacy in Legg-Calvé-Perthes disease.

#### Treatment of the defect

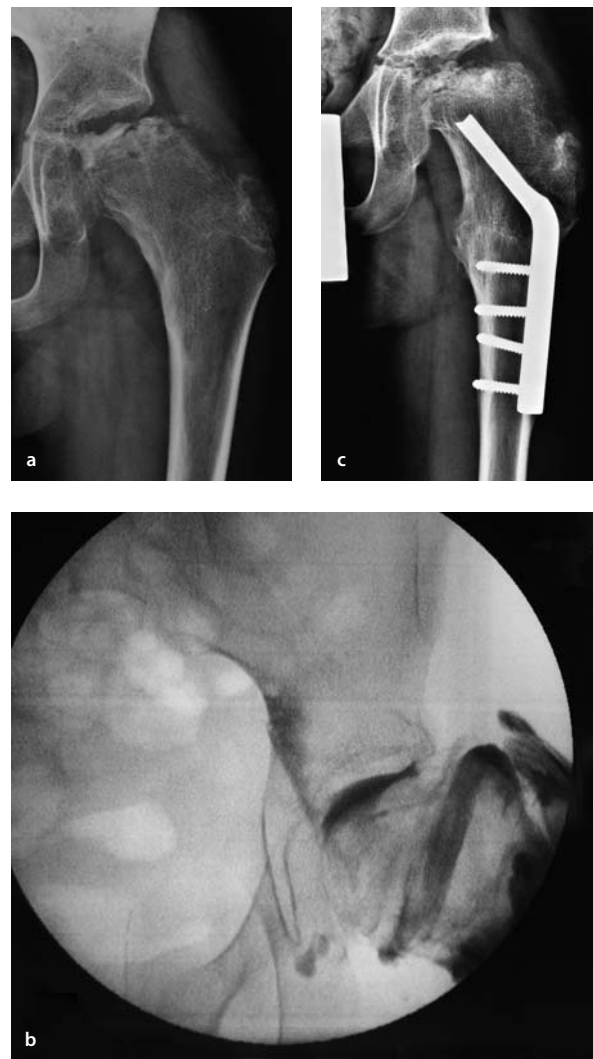
If the femoral head is deformed with a laterally protruding bump and hinge abduction in the older child (generally over 7 years), and if containment of the head is no longer possible, there are two treatment options:

- resection of the protruding bump,
- arthrodiastasis,
- intertrochanteric valgus osteotomy.

We have had little – and not particularly positive – experience of the bump resection. Since the joint cartilage inevitably has to be resected along with the bump, it is not surprising that the condition of the joint is frequently not improved by this procedure. In *arthrodiastasis* an external fixator is used between the pelvis and the femur. Distraction is applied and the hinge of the distractor is periodically opened to allow movement of the joint [47]. We have no experience with this method. The *intertrochanteric valgus osteotomy* on the other hand is often an effective procedure [60] (■ Fig. 3.193), allowing the surgeon to integrate the medial, better preserved, section of the femoral head in the weight-bearing zone, reduce the adduction contracture, distalize the greater trochanter and functionally lengthen the femoral neck. Our experience with this procedure has been very positive.

In a recent multicentric investigation among the members of the »European Pediatric Orthopedic Society« (EPOS) we tried to find out what principles of treatment are used in Europe [25]. A questionnaire was sent to all 297 members of the society describing four cases of Legg-Calvé-Perthes disease with 2 x-rays each and a short description of the clinical situation. Two of the patients were younger and two were older than 6 years of age. In both age groups there was one patient with a good range of motion and an x-ray classified Herring A or B, while the other patient had a poor range of motion and an x-ray classified as Herring C. The members were asked to choose from various treatment options or to describe any other therapy that they would advise in the clinical scenarios. 151 members answered the questionnaire. The participants had an average of 20 years of experience in pediatric orthopaedics. There was a consensus, that no

operation should be done in a young patient with a good range of motion, and that there should be no weight relief in older patients with a good range of motion. Conservative containment treatment (abduction splint, Petrie cast) and arthrodiastasis was suggested in only very few centers. There was a tendency operate on older patients with a poor range of motion, to provide surgical treatment only when there were subluxation or head at risk signs, and to perform pelvic osteotomies or a combination of pelvic and femoral osteotomies rather than femoral osteotomies alone. Age did not determine the indication for treatment and there was no agreement on the indications for physiotherapy. There was also no consensus on the type of pelvic osteotomy to be used. The study showed that indications



■ Fig. 3.193a–c. Treatment in the defect stage with intertrochanteric valgization osteotomy for severe hinge-abduction. a X-ray of the left hip of an 11-year old boy with pronounced lateral calcification and poor mobility. b Intraoperative arthrogram shows the hinge-abduction. c Situation 1 year after intertrochanteric valgization osteotomy. The »bump« has been rotated out of the weight-bearing zone

■ Table 3.29. Our therapeutic strategy for Legg-Calvé-Perthes disease

Under 5 years	If mobility is restricted: <i>physical therapy</i> Check ultrasound scan every 3 months X-rays (AP and axial) every 6 months up to 2 years after the diagnosis, and then annually
5 to 7 years	If mobility is restricted: <i>physical therapy</i> If mobility is very restricted: <i>botulinum toxin injection in the adductors</i> Check ultrasound scan every 3 months X-rays (AP and axial) every 6 months up to 2 years after the diagnosis, and then annually If decentering is present: <i>operation</i> Generally <i>intertrochanteric osteotomy</i> (precondition: epiphyseal plate not too steep, no major leg shortening and only slight restriction of abduction) If the preconditions for an intertrochanteric osteotomy are not satisfied, then <i>pelvic osteotomy according to Salter</i>
Over 7 years	<i>Triple osteotomy</i> of the pelvis (if mobility is greatly restricted poss. with pretreatment, see below)
Very pronounced decentering	Combined intertrochanteric varization osteotomy and triple osteotomy of the pelvis (if mobility is greatly restricted poss. with pretreatment, see below)
Pronounced restriction of abduction, but centering still possible under anesthesia	(usually as pretreatment prior to a triple osteotomy or intertrochanteric osteotomy) <i>Hydraulic mobilization</i> of the hip under anesthesia, Petrie cast in maximum abduction, intensive physical therapy under epidural anesthesia
Adduction contracture, centering not possible under anesthesia	Intertrochanteric valgus osteotomy

for treatment of Legg-Calvé-Perthes disease are based more on the personal experience of the surgeon rather than on scientific data.

### Our therapeutic strategy for Legg-Calvé-Perthes disease

Our therapeutic strategy for Legg-Calvé-Perthes disease is shown in ■ Table 3.29.

❗ **No weight-relieving devices, no abduction splints, crutches only for very severe pain, no total ban on sport!**

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## 3.2.5 · Legg-Calvé-Perthes disease

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### 3.2.6 Slipped capital femoral epiphysis

» When knee pain lets rip, always think of the hip. «

#### Definition

*Slipped capital femoral epiphysis refers to the atraumatic separation of the epiphysis in the epiphyseal plate of the femoral neck with dislocation of the femoral head, usually in a medial and dorsal direction, during the pubertal growth spurt.*

#### Historical background

E. Müller was the first to describe this condition, in 1888, in his paper entitled »On abnormal curvatures of the femoral neck during growth« [41].

#### Occurrence

An epidemiological study in the USA showed an incidence of approx. 2/100,000 adolescents under 20 years of age [25]. The male:female ratio is approx. 1.5 : 1, while that for unilateral to bilateral involvement is 4 : 1 [33]. An increase in the incidence of slipped capital femoral epiphysis has also been reported in the spring and summer months from April to August [29], while a more recent study has observed a concentration of cases in the Fall [36]. Its occurrence is also related to race: Slipped capital femoral epiphysis occurs more frequently in the black population than in whites [2, 31]. Our own study (► Chapter 1.1) based on the annual reports of the pediatric orthopaedic institutions of Switzerland found a marked decline in the incidence of this condition in Switzerland since 1960, even though the risk factors (overweight, sporting activity) have increased (also ► chapter 1.3).

#### Classification

Slipped capital femoral epiphysis can be classified according to the *duration of symptoms* [30]). Another classification [3, 30] makes a distinction based on *walking ability* (► Table 3.30).

#### Etiology

Experimental investigations with animals have shown that the mechanical strength of the epiphyseal plates is reduced by the influence of *hormones* during the pubertal growth spurt [40]. Testosterones promote growth, while estrogens tend to accelerate the maturation process. Testosterones and estrogens occur in boys and girls at the same time, but in differing concentrations.

Because of its anatomical situation, the proximal femoral epiphyseal plate is subjected to very high shear forces. The growth plate is protected from these forces by the perichondrial ring of fibrous ligaments. This zone is thicker in small children than in adolescents during the pubertal growth phase. If this zone is chronically overloaded, as is often the case with obese adolescents or



Fat children and sporting children are particularly at risk of suffering a slipped capital femoral epiphysis...

► **Table 3.30.** Classification of epiphyseal separation according to the duration of symptoms [28] and walking ability [3, 28]

– According to duration of symptoms	
Acute	Duration of symptoms <2 weeks
Chronic	Duration of symptoms >2 weeks
Acute on chronic	Duration of symptoms >2 weeks, but with sudden deterioration of symptoms, inability to walk
– According to walking ability	
Stable	Patient able to walk
Unstable	Patient not able to walk

those who are very keen on sports, the perichondrial ring can tear, causing the epiphysis to separate. The hormonal weakening of the epiphyseal plate is a physiological condition rather than a pathological process (► Chapter 2.2.3). Overweight, however, plays a major role in the etiology of slipped capital femoral epiphysis. In a recent study patients with this disease had significantly higher body mass index than the control group [37].

One study investigated the hormone levels in adolescents with slipped capital femoral epiphysis and found that these were normal in all cases, but that over half of the children with this condition were clearly overweight, with a *weight* above the 90th percentile [7]. If the hormone status is disrupted, however, a slipped capital femoral epiphysis can also occur in children of normal weight. This particularly applies in cases of hypothyroidism, growth hormone deficiency, panhypopituitarism and hypogonadism [32]. Maturation is impaired generally by hypopituitarism, causing the pubertal growth spurt to take place at an abnormally late stage, hence the possibil-

### 3.2.6 · Slipped capital femoral epiphysis

ity of slipped capital femoral epiphysis occurring at a later age (■ Fig. 3.194). Adolescents with adiposogenital dystrophy (Fröhlich syndrome) are at particular risk.

Slipped capital femoral epiphysis is observed over a wide range of *chronological ages*. We ourselves have observed the condition in 7- and 18-year olds. However, these are all chronological ages. If *bone age* is considered, there is a relatively short period of 4 years during which slipped capital femoral epiphysis can occur. In girls this lasts from 11–15 years, and in boys from 13–17 years [31]. A recent study [43] found that 90% of cases occurred in the accelerating phase of the pubertal growth spurt (► Chapter 2.2.2, ■ Fig. 2.8 and 2.9). Two-thirds occur even before the closure of the triadial cartilage. A recent study on a population of very young patients (below age 10, n=36) showed that a third of them had a previous pathology (Legg-Calvé-Perthes disease, leukemia, precocious puberty, DDH etc.) [16]. Finally, *genetic components* may also play a role, as has been demonstrated in a recent

study among the Amish people [37]. The genetic influence is also reflected by the fact, that in central Europe the incidence of the disease has diminished significantly since 1960, although obesity has become much more frequent among adolescents (see also ► chapter 1.3).

#### Pathophysiology

The slippage process is usually described as follows: The femoral head slips in a medial and dorsal direction. But this description is not wholly correct, since it is the femoral neck rather than the head that actually slips. The femoral head remains centered in the acetabulum, while the femur slips in a lateral and ventral direction. Rather than a pure translation, this movement involves rotation about an eccentric axis [4].

There have also been individual reports of slippage in the opposite direction, i.e. shifting of the femoral neck in a medial and ventral direction, or of the femoral head in a lateral and dorsal direction [46, 49]. Up to a certain extent, the slippage of the femoral head in a medial dorsal direction is a physiological process. In children with coxa valga or an anteverted hip, this process takes place during growth and leads to physiological derotation. As well as weight, sporting activity also places a particular stress on the hip. A comparative investigation with two groups of adolescents showed that a tilt deformity occurred much more frequently in adolescents who actively participated in sports than in those who were less active [42] (■ Fig. 3.195).

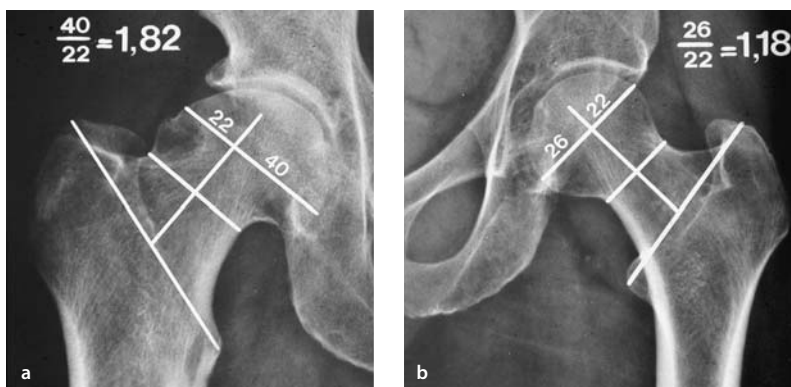
#### Diagnosis

A slipped capital femoral epiphysis is usually diagnosed on the basis of the pain reported by the patient. The pain is often stated to be in the thigh, or even in the knee, rather than the hip.

**!** The hip must therefore always be examined in adolescents who complain of pain in the knee or thigh area. If pain is reported during examination of the hip or restriction of hip movement, an x-ray must always be recorded.



■ Fig. 3.194a, b. AP (a) and axial (b) x-rays of the hips with *epiphyseal separation* in an 18-year old patient with *hypothyroidism* and whose epiphyseal plates are still open



■ Fig. 3.195a, b. Measurement of the *tilt deformity*: A perpendicular line dividing the femoral neck should pass roughly through the center of the widest part of the head. A ratio of 1.35 is normal. **a** Right hip: tilt deformity; **b** left hip: normal

Various attempts have been made to detect the slipped capital femoral epiphysis *sonographically* [23]. While an acute slippage of 15° or more can readily be diagnosed with the ultrasound method, we have not managed to detect mild forms of slipped capital femoral epiphysis on an ultrasound scan, although this would be particularly useful for detecting latent forms that are not yet clinically manifest.

*Examination* usually reveals a restricted internal rotation of the affected hip, and occasionally diminished abduction as well. Forced internal rotation often proves painful.

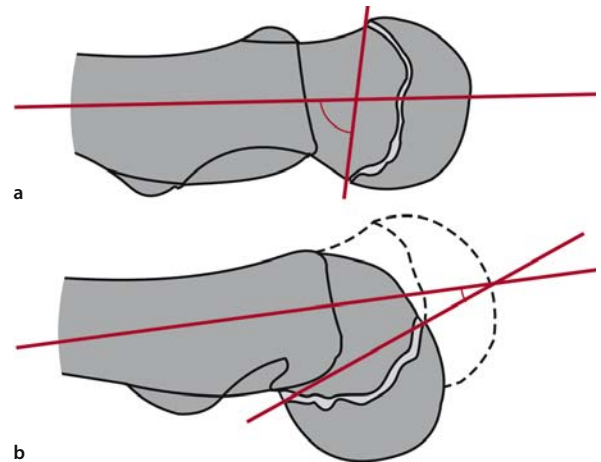
### Radiographic diagnosis

If a slipped capital femoral epiphysis is clinically suspected, both hips must always be x-rayed, with AP and axial views according to Lauenstein (for details of the x-ray technique for the Lauenstein view see ► chapter 3.2.2). One subtle radiographic indication is a widening of the epiphyseal plate on the AP view. In addition, the femoral head no longer projects as much laterally beyond the femoral neck as is normally the case. The offset is missing. It can often prove difficult to diagnose a slipped capital femoral epiphysis correctly on the AP view (► Fig. 3.196a top), hence the importance of the axial view, since the posterior slippage of the head or anterior slippage of the neck is usually clearly visible on this view.

► Fig. 3.197 shows the calculation of the tilt angle. Since the axial Lauenstein view is not effectively standardized, the measurement of the tilt angle is not particularly accu-

rate. More precise are the measurements on x-rays in the »frog position« (► Chapter 3.2.2) [34], although this is not always feasible in an acute case of slipped capital femoral epiphysis if the flexion position of 90° proves too painful.

We look for signs of remodeling on the x-ray, e.g. the rounding of the metaphyses or the filling of defects in the area of the epiphyseal plate. Ossification of the fibrocartilaginous ligament indicates that the slipped capital femoral epiphysis is not acute, but rather a chronic form or – if the pain is of recent onset – an acute flare-up of a pre-existing chronic slippage.



► Fig. 3.197a, b. Measurement of the slip angle in slipped capital femoral epiphysis



► Fig. 3.196a, b. 12 1/2-year old patient with acute epiphyseal separation. a AP (top) and axial (bottom) x-rays of the hips. b AP (top) and axial views (bottom) after bilateral nailing with Steinmann pins

### 3.2.6 · Slipped capital femoral epiphysis

Additional diagnostic imaging procedures are not usually required. If doubt exists about the openness of the epiphyseal plate (particularly on the opposite side), a CT scan may clarify the situation, although this investigation should not be performed routinely. An MRI scan can provide clarity about the circulation in the femoral head, and this information will prove useful prior to a scheduled open reduction of a severely dislocated head. Interpretation of the bone scan in respect of femoral head circulation is difficult, since the avascular zone is obscured by the surrounding hyperemia.

#### Spontaneous progression

In describing the spontaneous progression, we must further differentiate between acute and chronic slipped capital femoral epiphysis.

In a case of *acute slipped capital femoral epiphysis* the slippage is not spontaneously limited, but can continue arbitrarily. Nor are any remodeling processes observed. In a *chronic slipped capital femoral epiphysis* on the other hand, the process becomes self-limiting at some stage as a result of the following mechanisms:

- ossification of the plate,
- docking of the femoral dome dorsally against the femoral neck,
- ossification of the fibrocartilaginous ring on the femoral neck [13].

Premature physeal closure generally occurs some 8–9 months after a slipped capital femoral epiphysis [13].

Further remodeling occurs *after the completion of growth*. The following specific processes take place [13]:

- flattening of the metaphyseal hump,
- the exposed metaphysis is rounded off,
- filling of defects in the epiphyseal plate area,
- correction of epiphyseal abnormal positions.

In around 40% [13, 22] to 66% [27] of cases, slippage also occurs on the opposite side over the course of time, particularly if the plate is relatively steep.

#### Calculation of the osteoarthritis risk in long-term studies [12, 21]

- After 30 years
  - with a slip angle of 40°: Risk 15%,
  - with a slip angle of 60°: Risk 25%.
- After 50 years
  - with a slip angle of 40°: Risk 50%,
  - with a slip angle of 60°: Risk 70%.

Even with a relatively small slip angle, loss of the offset of the femoral head can occur on the anterolateral side, leading to impingement against the anterior rim of the

acetabulum. This can create a labrum lesion and subsequent damage to the cartilage as a result of the shear effect [28]. This impingement is probably the main factor responsible for the subsequent osteoarthritis. Although the impingement is regularly observed even with a small slip angle, the resulting osteoarthritis risk is still very unclear. According to long-term studies, clinically relevant osteoarthritis occurs in only half of the cases after 50 years with slip angles of up to 40°.

The risk of *femoral head necrosis* depends on the extent of the slippage and the speed of the slipping process. A probability of 16–18% has been stated for acute slipped capital femoral epiphysis [44]. The risk appears to be particularly high if separation of the anterior lip of the femoral head from the metaphysis is visible in the axial view [5]. But necrosis more often occurs iatrogenically as a result of a forced closed reduction or an incorrectly performed open reduction of a chronically slipped femoral head.

A special problem is posed by *clinically non-manifest slipped capital femoral epiphysis*. A fairly old study found that tilt deformity occurred more frequently in athletically very active children and adolescents than in those who were less athletic [42]. AP x-rays of the hips were measured in this study. ■ Fig. 3.195 shows an x-ray with the calculation of the tilt deformity, which doubtless represents a pre-arthritis condition. There are also isolated reports of *elite athletes* with a history of slipped capital femoral epiphysis suffering from osteoarthritis more frequently than the average citizen [20].

#### Treatment

Treatment of slipped capital femoral epiphysis in its florid phase is always surgical, whether the condition is unstable (acute) or stable (chronic). Attempts to reduce acute cases



Impingement in the area of the hip...



by longitudinal traction and internal rotation have not been convincing [12, 38].

The following surgical options are available:

- closed reduction and nailing,
- closed reduction and screw fixation,
- open reduction and screw fixation,
- subcapital wedge osteotomy and screw fixation,
- intertrochanteric flexion-valgization-rotation osteotomy.

In a case of unstable slipped capital femoral epiphysis, reduction and fixation must be performed as an emergency measure.

**! Unstable slipped capital femoral epiphysis is one of the few emergency situations in pediatric orthopaedics that are not caused by trauma.**

After the patient has been admitted to the hospital as an emergency, we confirm the diagnosis and assess the extent of the slippage by means of AP and frog-position x-rays (or a Lauenstein x-ray if the latter is not possible). We also determine the skeletal age on the basis of a radiographic hand plate. If the slip angle is 40° or less we perform (where possible) closed reduction and fix the result with nails or screws. If the angle is over 40° we record an MRI scan. If the circulation is intact, we perform a subcapital wedge osteotomy or open reduction and fixation. If the circulation is impaired, we fix the slippage in situ and may possibly implement a correction osteotomy at a later date (depending on the recovery of the femoral necrosis after the avascular crisis).

### Closed reduction

An attempt is made to reduce the femoral head as far as possible on the anesthetized patient with the hip in 90° flexion, abduction and careful internal rotation. The result must be fixed in this position with nails or screws under image-intensifier control.

### Fixation with nails or screws

#### Steinmann pins, hook-pin

The leg is placed in a flexed position with maximum internal rotation and the first Steinmann nail is inserted anteriorly through the femoral neck into the head under image-intensifier control through a lateral incision. The second Steinmann pin is then inserted above the first, in a posterior-anterior direction, and a third pin distal to the first in an anterior-posterior direction. The Steinmann pins now appear parallel to each other on the AP view and fanned out on the axial view (■ Fig. 3.196).

An elegant method involves the use of the Hansson hook-pin [18]. A cannulated nail contains a threaded pin



■ Fig. 3.198. AP x-ray of the hips of a 10-year old girl with slipped capital femoral epiphysis and fixation with Hansson hook-pin

that emerges from the nail tip and can be screwed into the femoral head (■ Fig. 3.198). The screw length is selected so as to achieve a lateral protrusion out of the femur by 1.5 cm. The femoral neck growth is preserved, both implantation and metal removal can be performed by minimally invasive techniques. Subsequent nailing as a result of lengthening of the femoral neck is not required.

#### Cannulated screw

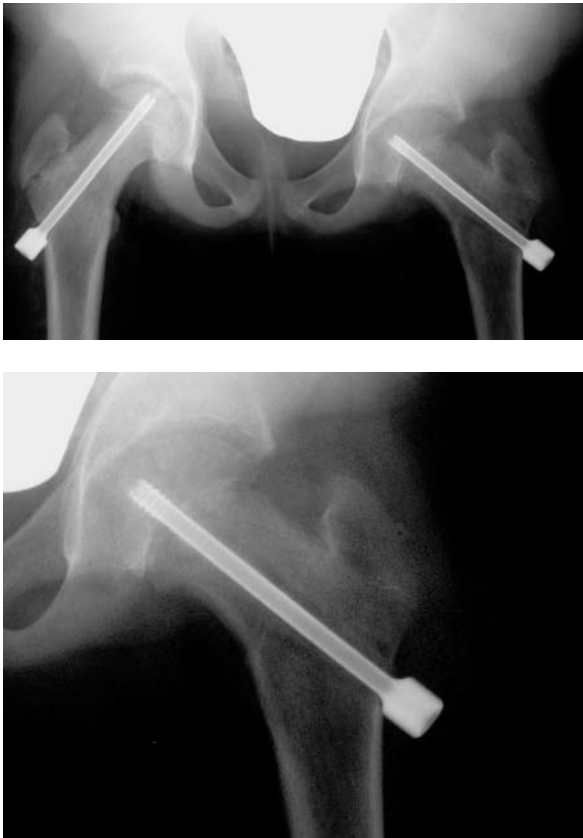
With the leg in maximum internal rotation and flexion, a guide wire with a fine thread at the tip is inserted centrally through the femoral neck and into the head via a stab incision. Under the image intensifier, the position of the wire is checked in two planes. If it is correctly positioned, a cannulated screw is screwed into the femoral head from the lateral side after length measurement and pre-drilling [17, 19]. A useful modification was recently proposed involving a reverse-cutting, cannulated screw. Selecting a screw that is 1 cm too long causes the screw to protrude laterally out of the femur by this length, thus ensuring that length growth of the femoral neck is preserved (■ Fig. 3.199) [26]. A recent survey among the members of the Pediatric Orthopedic Society of North America showed, that in North America pins are rarely used. 57% recommended the use of one single threaded screw for fixation of an unstable hip, 40.3% recommended two screws [39].

#### Open reduction

This technique has been developed in Berne [28]. A trochanteric flip osteotomy is first performed with the patient in the lateral position. The gluteal and vastus lateralis muscles are left in place on the trochanteric fragment as this facilitates subsequent healing of the fragment. The joint capsule is opened with a ventral Z-shaped incision,



### 3.2.6 · Slipped capital femoral epiphysis



■ **Fig. 3.199.** AP x-ray of the hips of a 10-year old girl with slipped capital femoral epiphysis and fixation with cannulated screw

the dorsal callus is removed while preserving the dorsal vessels, and the femoral head is reduced until an adequate offset is restored. Sufficient space must be created to ensure that the head can be reduced without causing any traction to be exerted on the dorsal vessels. The head is fixed with two screws, and the trochanter is also refixed with screws (■ Fig. 3.200).

#### Subcapital wedge osteotomy

We perform the wedge osteotomy (in contrast with open reduction) according to Smith-Petersen via an anterior approach. After exposing the hip, we perform the osteotomy at subcapital level, i.e. slightly proximal to the epiphyseal plate. In contrast with the open reduction, no translational movement is performed on the head, which is shifted dorsally and rotated, although the tilt is cancelled. This measure also corrects the loss of offset. The femoral neck is narrowed slightly depending on the anatomical configuration in each case. The advantage of the wedge osteotomy is the much reduced risk of femoral head necrosis compared that associated with open reduction. If a rotation deformity remains, this can, if necessary, be corrected secondarily by an intertrochanteric osteotomy.

#### Treatment of the opposite side

As a rule, we also perform pinning or screw fixation on the opposite, non-slipped side. While the *need for prophylactic pinning* of the opposite side is a matter of dispute, even the authors of one study who consider pinning of the other side to be unnecessary report the occurrence of slippage on the contralateral side in 40% of cases [22]. Other authors state a frequency of 66% [27]. Since the load must be taken off the affected side after a slipped capital femoral epiphysis, the load on the contralateral side is increased, resulting in a relatively high risk of slippage on that side as well. However, the risk becomes very much smaller after physeal closure [43]. In a recent study morphometric parameters were measured to assess the risk of slip of the contralateral hip. 36 patients and 23 healthy controls were evaluated. In the axial view, control patients had a mean value for the posterior sloping angle of the capital femoral physis of 5 degrees, compared with 12 degrees in unilateral and 18 degrees in patients developing bilateral slipped capital femoral epiphysis. The authors postulated that the risk of developing bilateral disease can be predicted by measuring the posterior sloping angle of the capital femoral physis in the axial view. Prophylactic pinning of the healthy contralateral side should be recommended only in patients showing an axial posterior sloping angle of the physis of over 12 degrees [6].

#### Complications

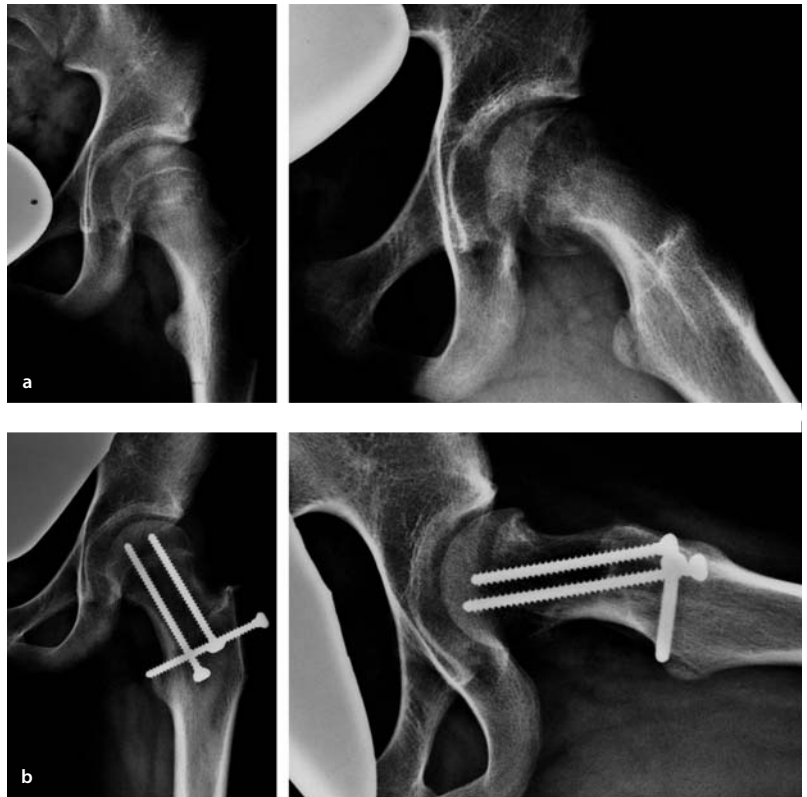
The following complications are known to occur after reduction, nailing or screw fixation [3, 10, 45, 48]:

- chondrolysis,
- femoral head necrosis,
- premature physeal closure,
- infection,
- screw/nail breakage.

*Chondrolysis* can occur in two ways:

- As a result of *perforation by a nail or screw*. This problem is a question of surgical technique. The position of the nail or screw must be checked under the image intensifier in all planes, since it is only visible if the femoral head is struck tangentially by the perforation. An AP and axial x-ray must also be recorded postoperatively.
- *Idiopathically*, i.e. as a complication of the condition itself. This cause is probably less common than the iatrogenic cause.

*Femoral head necrosis* is a serious complication of slipped capital femoral epiphysis and can occur, for example, if force is used to reduce a chronic case. An incorrectly performed open reduction can also cause this complication. When faced with evidence of a chronic process on the x-ray, the surgeon therefore has to decide whether a reduction can be performed at all, or should only be fixed in situ. Various authors recommend in-situ pinning, stat-



**Fig. 3.200a, b.** 12-year old girl with acute on chronic epiphyseal separation with a slip angle of  $>40^\circ$ . **a** AP (left) and axial (right) x-rays of left hip. **b** AP and axial views after open reduction with trochanteric flip osteotomy and screw fixation

ing that partial reduction can occur spontaneously as a result of the remodeling process [9, 45].

We do not share this view, as remodeling was observed in only a third of cases. On the other hand, a closed reduction attempt does not have any disadvantages provided moderate force only is used for the internal rotation. This method is only appropriate for reducing an acutely slipped head. Of course, a chronically dislocated femoral head should not be returned to its original position with force, otherwise the risk of femoral head necrosis becomes much greater.

If the history is not clear, we also always attempt a (gentle) reduction since, in the majority of diagnosed cases, an acute slippage has already been complicated by a chronically dislocated position. A correct technical procedure is particularly important with open reductions. The head circulation can be assessed during the operation. In two cases we have found that the head was avascular even before the reduction. Consequently, we now record an MRI scan preoperatively. However, we also have to report two cases of iatrogenic femoral head necrosis in our own hospital.

The surgical technique described by Leunig et al. [14] must be followed meticulously. Although the wedge osteotomy may not be as effective at restoring the anatomical configuration, the risk of a femoral head necrosis is much less with this procedure since no translation movement is involved and thus no traction is exerted on the posterior vessels. The open reduction should only be performed

by surgeons who are sufficiently familiar with the Berne technique, although even then it remains a risky procedure. Otherwise, the subcapital osteotomy represents a less hazardous alternative.

*Premature physeal closure* occurs more frequently after the use of screws compared to nails, although it has also certainly been observed after nailing as well [45]. The *infection rate* in slipped capital femoral epiphysis is no higher than for other interventions involving metal implants. *Metal breakage* only occurs if excessively thin nails or screws are used.

#### Metal removal

On completion of growth we generally remove nails and screws. Normal screws are difficult to remove as they can break. This risk is reduced with reverse-cutting screws.

#### Correction of the deformity (after completion of growth)

##### Subcapital wedge osteotomy

If the femoral head has slipped by more than  $40^\circ$ , a correction must be performed. The most efficient solution is to implement this as close as possible to the deformity, i.e. ideally at subcapital level. The technique is described above.

While the risk of a femoral head necrosis is slightly higher for this procedure than for the intertrochanteric osteotomy [1, 11, 14], it is low if the surgical technique is meticulous.

### 3.2.6 · Slipped capital femoral epiphysis

#### Intertrochanteric flexion-valgization-rotation osteotomy

In 1954, Imhaeuser described [21] the intertrochanteric flexion-valgus osteotomy for the correction of the deformity after severe slippage in a case of slipped capital femoral epiphysis. After Southwick described the same operation in the English literature in 1967 [47], the technique is more commonly associated with the name »Southwick« in the English-speaking world than with the name »Imhäuser« (■ Fig. 3.201). This operation represents an alternative to the subcapital wedge osteotomy, and the risk of femoral head necrosis is slightly reduced. As a rule, it can also be performed during puberty, even in the florid stage with concurrent nailing.

However, since the correction does not take place at the site of the deformity, the abnormal shape of the femoral neck is not rectified. Flexion of the femoral neck can reduce the anterior impingement, but only to a limited extent. The ventrally protruding femoral neck strikes the anterior acetabular rim not only during sitting, but also during walking, and can be avoided by external rotation of the leg. Consequently, the osteotomy aims to increase not only flexion and valgization, but anteversion as well. Although no correction is performed at the site of the

deformity, the results are perfectly acceptable in terms of the development of osteoarthritis over a follow-up period of over 20 years [24]. Nevertheless, we currently prefer to implement corrections close to the joint.

#### Contouring of the femoral neck

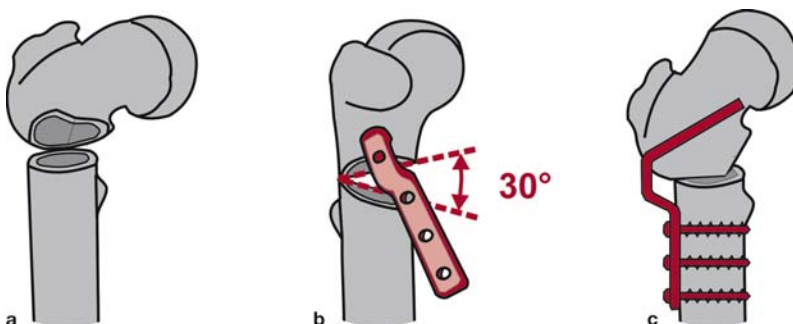
If ventral protrusion of the femoral neck and resulting impingement has occurred after only slight slippage, the impingement can be eliminated by appropriate contouring of the femoral neck (bump resection«). If a relevant labrum lesion is already present (which can be visualized by arthro-MRI), the contouring can be performed in connection with a surgical hip dislocation [15]. This procedure gives the surgeon a complete overview of the joint so that any additional measures for reducing the pain associated with the labrum lesion can be implemented. If no labrum lesion is present, the contouring can also be performed via a ventral approach without surgical hip dislocation.

#### Our therapeutic strategy for slipped capital femoral epiphysis

Our therapeutic strategy for epiphyseal separation is shown in ■ Table 3.31.

■ Table 3.31. Our therapeutic strategy for epiphyseal separation

Acute and acute on chronic epiphyseal separation (unstable hip)		
Slip angle <40°		Careful closed reduction of the slipped side and nailing with hook-pin or screw fixation of both the affected and unaffected sides
Slip angle >40°		Subcapital wedge osteotomy or open reduction with trochanteric flip osteotomy, screw fixation
Chronic epiphyseal separation (stable hip)		
Slip angle <40°		Nailing with hook-pin or screw fixation of both the affected and unaffected sides
Slip angle >40°		Subcapital wedge osteotomy
Deformity after completion of growth		
Slip angle <40°		Shaping of the femoral neck by bump resection with surgical hip dislocation, poss. also by an anterior approach without hip dislocation
Slip angle >40°	Only slight reduction of the offset (i.e. good remodeling)	Intertrochanteric flexion valgus-internal rotation osteotomy (Imhaeuser-Southwick)
	Distinct reduction of the offset	Subcapital wedge osteotomy



■ Fig. 3.201a–c. The Imhaeuser-Southwick osteotomy is an intertrochanteric osteotomy involving concurrent valgization and flexion. **a** osteotomy, anterior view; **b** osteotomy and insertion of angulated blade plate, lateral view; **c** postoperative situation, anterior view

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### 3.2.7 Congenital malformations of the pelvis, hip and thigh

#### Classification

We distinguish between localized disorders and those associated with a systemic illness.

The localized disorders include:

- teratological dislocation of the hip,
- proximal femoral focal deficiency,
- coxa vara and femoral neck pseudarthrosis.

Typical changes in this area are found in association with the following systemic illnesses:

- multiple epiphyseal dysplasia,
- spondyloepiphyseal dysplasia,
- dysplasia epiphysealis hemimelia,
- Down syndrome (trisomy 21),
- osteogenesis imperfecta,
- fibrous dysplasia,
- trichorhinophalangeal syndrome,
- osteopetrosis,
- metachondromatosis,
- pseudoachondroplasia,
- Stickler syndrome.

These diseases are discussed in ► chapter 4.6. At this point we shall restrict ourselves to the specific changes in those forms of multiple epiphyseal dysplasia that are manifested in the hip only.

#### 3.2.7.1 Localized disorders

##### Teratological hip dislocation

#### ► Definition

Dislocation of one, or usually both, hips at birth as a result of malformations rather than immaturity of the joints, and associated with other deformities.

#### Occurrence

Since teratological hip dislocation is not a systemic illness it is included in this section on localized disorders, even though teratological hip dislocations are generally associated with other deformities. Typical concurrently occurring deformities are:

Torticollis, plagiocephaly (32%) [34], arthrogryposis, Larsen syndrome, general ligament laxity [34], flat feet, club feet, proximal femoral focal deficiency, congenital knee dislocation, pyloric stenosis, renal agenesis and orchidocele. Compared to dysplasia-related hip dislocation, teratological dislocation of the hip is extremely rare.

#### Diagnosis

- ! If an abnormality of any kind exists at birth, an ultrasound scan of the hips is invariably indicated.

Sonography at birth shows a Graf hip type III or IV [12] (► Chapter 3.2.4). If a teratological dislocation is suspected, an x-ray and MRI scan are indicated as deformities of the femoral head (e.g. double formation) and/or the acetabulum can occur, which is relevant for the treatment.

#### Treatment

The treatment of teratological dislocations is essentially no different from that for dysplasia-related hip dislocation, although the prognosis is much worse. An open reduction is usually unavoidable, and deformities of the soft tissues and the bony and cartilaginous skeleton also have to be taken into account (see ► chapter 3.2.4 for treatment of hip dislocation). The risk of redislocation is much greater than with dysplasia-related dislocation.

#### Longitudinal femoral malformations (proximal femoral focal deficiency, femoral hypoplasia, congenital femoral deficiency)

#### ► Definition

The spectrum of malformations of the femur range from a slight shortening to complete absence of the whole femur. If a deformity or defect of the femur exists, the proximal part is always affected as well, hence the description of — proximal femoral focal deficiency (abbreviated to PFFD) or congenital femoral deficiency (CFD)

#### Classification

Various classifications have been proposed for proximal femoral focal deficiency [1, 8, 11, 19]. The classification most commonly used is that of *Aitken* [1] (► Fig. 3.202). This is a purely radiological classification and thus incomplete [11]. The condition frequently has to be reclassified during the course of growth. A comprehensive classification of congenital anomalies of the femur has been proposed by *Pappas* [23] (► Fig. 3.203), which divides the condition into 9 classes (► Table 3.32a). More recently *Paley* [25] proposed a classification with 3 types (► Table 3.32b).

In the early stages the non-ossified structures can be visualized sonographically [13], by MRI or by arthrography (► Fig. 3.204, 3.205). In particular, these techniques can show whether a femoral head is present or not, a finding that is important for correct classification.

#### Occurrence

The incidence of proximal femoral focal deficiency calculated in an epidemiological study was found to be 2 per 100,000 neonates [28]. If *all* femoral anomalies are taken into account, the frequency is undoubtedly much greater since mild forms of femoral hypoplasia in particular are very numerous and usually not yet diagnosed at birth.



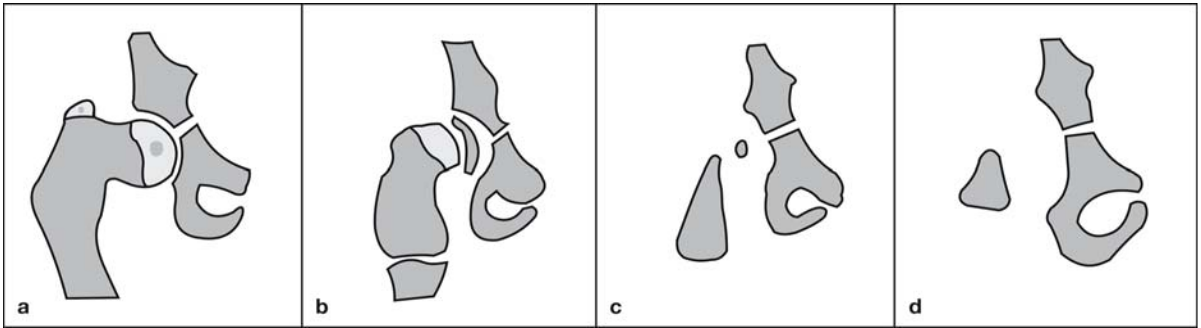


Fig. 3.202a–d. Classification of the proximal focal femoral deficiency (PFFD) according to Aitken [1]. a Bone connection between the femoral head and shaft. b Femoral head present, but no bone bridge with

the shaft. c No femoral head (or just a remnant) present. d Only the distal portion of the femur present

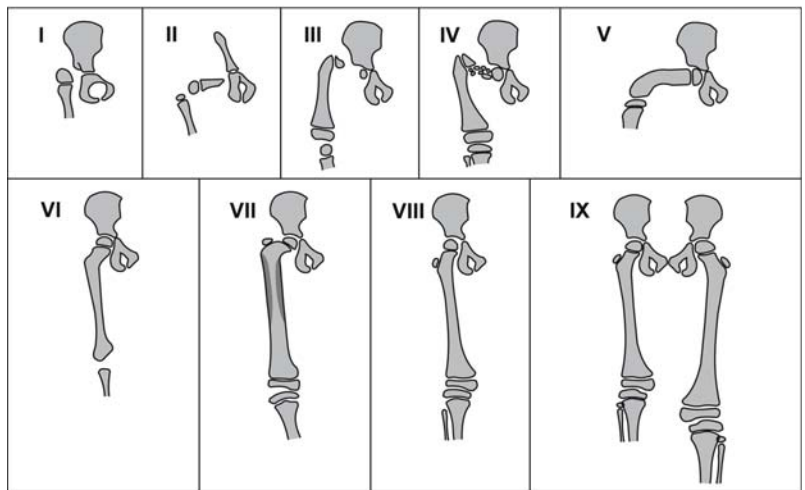


Fig. 3.203. Classification of a proximal focal femoral deficiency (PFFD) (I–IX) according to Pappas [23] (see text)

Table 3.32a. Classification of the congenital anomalies of the femur after Pappas [23]

Class	Characteristics
I	Complete absence of the femur
II	Proximal femoral deficiency combined with lesion of the pelvis
III	Proximal femoral deficiency without bone connection between the femoral shaft and head
IV	Proximal femoral deficiency with poorly organized fibro-osseous connection between the femoral shaft and head
V	Femoral deficiency in the middle of the shaft with hypoplastic proximal or distal bony development
VI	Distal femoral deficiency
VII	Hypoplastic femur with coxa vara and sclerosis of diaphysis
VIII	Hypoplastic femur with coxa valga
IX	Hypoplastic femur with normal proportions

Table 3.32b. Classification of congenital femoral deficiency (CFD) after Paley [22]

Type	Characteristics
1	<b>Intact femur</b> with mobile hip and knee a) normal ossification of proximal femur b) delayed ossification of proximal femur
2	<b>Mobile pseudarthrosis</b> (hip not fully formed, a false joint) with mobile knee a) femoral head mobile in acetabulum b) femoral head absent or stiff in acetabulum
3	<b>Diaphyseal deficiency</b> of femur (femur does not reach the acetabulum) a) knee motion > 45° b) knee motion < 45°



■ **Fig. 3.204.** X-ray of the pelvis and thigh shortly after birth. The right femur is completely missing



■ **Fig. 3.205.** Arthrography of the hip of the same patient shown in Fig. 3.204 3 months later shows, despite the apparent absence on the x-ray, a femoral head is present

### Etiology

Proximal femoral focal deficiency occurs as a result of a noxious event (viral infection, drug, radiation, mechanical factor, etc.) between the 4th and 9th week of pregnancy. The duration of action and aggressivity of the noxious event determine only the extent of the lesion rather than its location. A recent report [2] has provided evidence of a possible hereditary variant.

### Associated anomalies

The incidence of associated anomalies is very high, with figures of up to 70% [1]. A longitudinal defect of the fibula, often with shortening of the tibia, is also present in over fifty percent of cases (► Chapter 3.3.6). The patella is frequently dysplastic and occasionally lateralized as well. The knee is usually in an abnormal valgus position. The foot generally lacks one or more rays while, at the back of the foot, instability of the talus is frequently present because of the dysplastic fibula. This is often compounded by vertical positioning of the talus or talocalcaneal coalition. More rarely, the contralateral side or one of the upper extremities is also affected.

### Clinical features, diagnosis

Since the shortening in femoral hypoplasia is often not very pronounced at birth, the condition is sometimes not diagnosed until early childhood. In the case of a congenital femoral deficiency, on the other hand, the deformity is already clearly visible at birth. The thigh is

large, ungainly and shortened. Sometimes the shortening of the extremity in the infant is so severe that the foot is at the level of the knee on the opposite side. MRI investigations have shown that, although the muscles can also be hypoplastic, all muscles are nevertheless formed even in the more severe forms [25]. Careful clinical examination is always required at birth since, as has already been mentioned, associated anomalies are extremely common.

### Treatment

The treatment of congenital anomalies of the femur is very time-consuming and requires a lot of experience. The following *treatment options* are available:

- shoe elevation
- orthotic provision
- prosthetic provision
- realignment osteotomies
- implantation of the femoral stump in the femoral head
- arthrodesis of the stump with the acetabulum in combination with a Chiari pelvic osteotomy
- arthrodesis of the knee with preservation of the growth plates
- surgical leg lengthening
- rotationplasty
- amputation.

❗ **Any treatment of these patients, who often have highly visible handicaps, should be accompanied by good psychological support.**

The therapeutic strategy should be discussed as soon as possible with the parents and definitively established during the course of early childhood. The most important issue is deciding whether complete preservation of the limb should be attempted with surgical leg lengthening until the conclusion of growth, or whether some other solution should be chosen. In cases of very severe deformities, leaving the leg the same length and amputation of the forefoot with a prosthesis or a rotationplasty with a lower leg prosthesis is often the better solution in functional respects than surgical leg lengthening, although this may be psychologically more difficult for the parents and patients to accept. Apart from the condition of the femur, that of the lower leg and foot must also be considered (► Chapter 3.3.6).

The number of rays on the foot gives a good indication of the prognosis and severity of the deformity. If there are just two toe rays, the chances of achieving a properly functioning lower extremity with leg lengthening are very low. In such cases, the parents and the child should be carefully guided towards other options and helped to accept the handicap.

### Therapeutic strategies

#### No femoral head present (Aitken type D, Pappas types I and II, Paley type 3)

Some authors recommend *arthrodesis between the femoral stump and pelvis* (in combination with a Chiari pelvic osteotomy). The knee then serves as a »hinge hip«. The femoral stump is directed ventrally during the fusion so that flexion of the knee then corresponds to extension of the »hip«. Although this procedure stabilizes the hip and reduces limping slightly, the prosthetic provision is difficult because of the reduced mobility of the joint.

An alternative procedure is *rotationplasty*, in which the femoral stump is turned through 180° and fused with the pelvis. The knee then functions as the hip and the ankle as the knee. This is a good solution in functional terms, but can be rather problematic from the acceptance standpoint because of the rear-facing foot.

Patients without a femoral head can simply be treated conservatively and fitted with a prosthesis. In this case no decision to operate need be taken in early childhood.

#### Major deficiency, but femoral head present (Aitken type C, Pappas type III, Paley type 2b):

In these cases an attempt should be made to create a link between the femoral stump and head (■ Fig. 3.206), although this is not always successful initially. The presence of the head can be confirmed even at birth with modern imaging techniques. However, we prefer to wait until the head center starts to ossify before operating. If there is no femoral neck at all, fixation is very difficult, but more readily achievable in the slightly larger child than in the infant.

If this link can be made the problem of the excessively short thigh must then be resolved. The knee, which is positioned far higher than normal, occasionally has to be fused (with preservation of the growth plates). The forward-projecting foot hampers the prosthetic provision. The two following solutions can produce a functional benefit:

- amputation of the foot,
- rotationplasty (rotation of the foot through 180° so that it functions as a »knee«; ► chapter 4.5.5).

Amputation of the projecting foot facilitates prosthetic management and enhances the cosmetic appearance. Rotationplasty causes the ankle to serve as a knee, producing a substantial functional benefit [9, 15]. However, neither operation is readily accepted from the psychological standpoint. Amputation also carries the risk of phantom pain, which is not the case with a rotationplasty.

#### Small or moderately large deficiency (Aitken type B, Pappas type IV, Paley type 2a):

An attempt should always be made to rectify the defect. A (substantial) varus deformity is often present and will need to be corrected. The treatment of the – usually substantially – shortened leg is addressed in the following section.

#### No deficiency, but abnormal curvature and/or severe shortening (»femoral hypoplasia«; Aitken type A, Pappas types V and VI, Paley type 1b):

The therapeutic strategy is based not only on the deformity but also on the age of the patient.

- *Preschool age (up to 6 years)*: Depending on the extent of the shortening in each case, the leg-length discrepancy can be offset with a sole wedge in the shoe or a lower leg orthosis. If possible, the foot is placed in a plantigrade position in the orthosis. Severe curvatures at this age should be corrected by an osteotomy.
- *School age (6–10 years)*: If the leg is shortened by more than 10 cm at this age, the orthopedist has to decide whether complete preservation of the extremity should be attempted with surgical leg-length matching until the conclusion of growth, or whether some other solution should be chosen. An initial lengthening operation is generally performed at this age (up to a maximum of 8 cm; ► chapter 4.2.2).
- *Adolescence*: A second and third lengthening of a maximum of 8 cm in each case can be performed until the conclusion of growth. In total, elongations of more than 100% of the original length are possible [3, 10, 29].

#### No defect, moderate curvature and/or shortening (»femoral hypoplasia«; Pappas types VII–IX, Paley type 1a):

In these cases the question of whether to lengthen or not is almost irrelevant. The operations for axial correction and lengthening listed above should be performed.



■ **Fig. 3.206a–e.** X-ray of the right hip of a girl with *proximal focal femoral deficiency (Pappas type III)*. **a** At the age of 2 weeks. **b** At 4 years of age a femoral head is visible. **c** The connective tissue defect is resected and the femur positioned beneath the head. **d** A pseudarthrosis

occurred between the femoral neck and shaft as a result of the excessive varus position. **e** A valgization osteotomy and stabilization with angled blade plate was performed at the age of 5 years, producing a stable situation in the proximal femur

### **Congenital coxa vara, femoral neck pseudarthrosis**

#### ➤ **Definition**

Probable hereditary disorder with abnormal varus position of the femoral neck with or without pseudarthrosis.

#### **Occurrence**

This is a very rare disorder. An incidence of 1:25,000 birth has been calculated for Scandinavia [18], where this deformity is more common than in Central Europe [5]. Both sides are affected in around 30% of cases. It is also associated with an increased familial occurrence.



### Etiology

In contrast with a proximal femoral focal deficiency, this is very probably a hereditary disorder rather than the result of damage during the pregnancy. Endochondral ossification of the femoral neck is impaired in this condition. Pseudarthrosis is not generally present initially at birth, but rather occurs as a result of abnormal shear forces in the region of the femoral neck, which cause the abnormal varus position to increase gradually until the pseudarthrosis eventually forms. The biomechanical problems of this condition were described by Pauwels as early as the 1930's [24] (▶ Chapter 3.2.3).

### Clinical features, diagnosis

The disorder is not usually yet apparent at birth. After the infant starts walking a leg-length discrepancy becomes increasingly evident (with a unilateral condition) and, over time, is also accompanied (as a result of insufficiency of the abductors) by a Duchenne-Trendelenburg limp. A flexion contracture of the hip and lumbar hyperlordosis can also be expected. The coxa vara are often compensated for by genua valga, which is not always easy to detect clinically [30].

A narrowed CCD angle is apparent on the AP *x-ray* of the hip (▶ Chapter 3.2.3). In the region of the femoral neck, and laterally to the epiphyseal plate, a section of osteolysis that gradually deviates laterally in the caudal direction is apparent and forms an inverse »V« shape together with the growth plate. This osteolysis is not yet visible as a rule at the onset of walking and only manifests itself in the older toddler (■ Fig. 3.207). No further imaging investigations are required.

The *differential diagnosis* should take into account those diseases in which the bone quality is diminished,

potentially resulting in varus curvature of the femoral neck. One such condition in which this typically occurs is *fibrous dysplasia* (»shepherd's crook deformity«; ▶ chapter 4.6.2.31), while curvature (usually of the whole femur) in a varus direction can also occur in *osteogenesis imperfecta* (▶ Chapter 4.6.2.24). A similar condition can occasionally also be seen in rickets (see ▶ chapter 4.6.2.25). *Epiphyseal dysplasia* can also be associated with a coxa vara (see below).

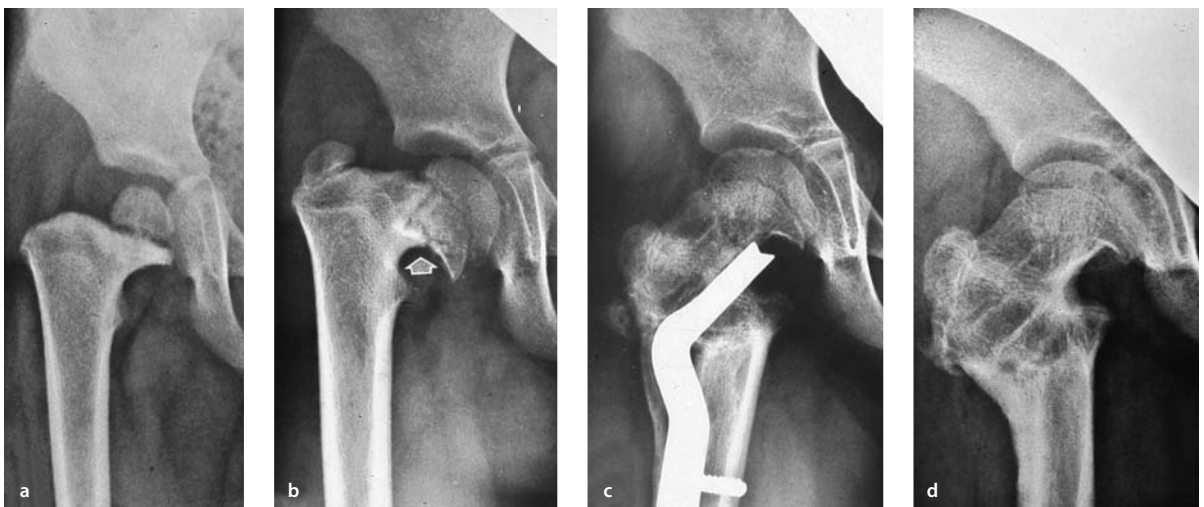
### Treatment

The treatment of choice is a valgus osteotomy, e.g. in the form of the Y-osteotomy proposed by Pauwels[24] (■ Fig. 3.208, also ■ Fig. 3.207), which normalizes the biomechanical situation in the femoral neck area. The pseudarthrosis then heals spontaneously since it is, after all, merely the consequence of abnormal shear forces (■ Fig. 3.207). If the bone has been shifted sufficiently in the valgus direction, recurrence will not occur during the course of subsequent growth [6, 14]. If the correction osteotomy is performed before the age of 10, the acetabulum will develop normally [4]. A conservative treatment offers no prospect of success.

### Pelvic changes in bladder exstrophy

#### Definition

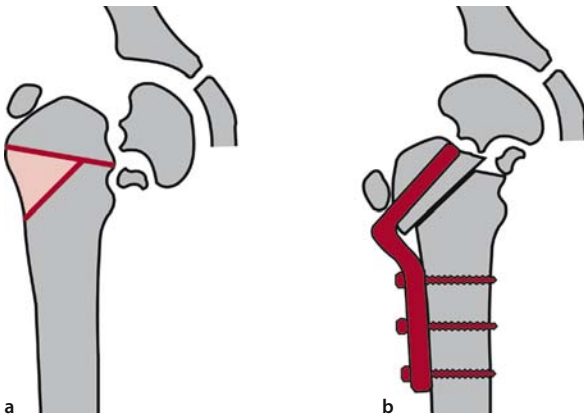
Congenital defect of the anterior bladder and abdominal wall below the navel producing exposure of the posterior wall of the bladder, trigone and urethral wall as a result of failed mesodermal differentiation. An anomaly of the pelvis is always associated with an anterior opening in the area of the symphysis and a sagittal orientation of the acetabulum.



■ Fig. 3.207a–d. 2-year old child with congenital coxa vara. a AP *x-ray* of the right hip. b A femoral neck pseudarthrosis has formed by the

age of 6 years. c A Pauwels Y-osteotomy was subsequently performed. d 1 year postoperatively the pseudarthrosis has healed





■ Fig. 3.208a, b. Principle of the *Pauwels Y-osteotomy* in femoral neck pseudarthrosis. This essentially involves a very pronounced valgization. a Osteotomy, b postoperative situation

### Clinical features, diagnosis

The pelvic anomaly is important particularly in connection with the bladder anomaly. The pelvis is opened at the front like a book. Accordingly, the ilium and acetabulum are oriented in a much more sagittal direction than normal. This incorrect orientation results in an externally rotated basic position of the legs. A myelomeningocele is also occasionally present.

### Treatment

If the bladder is reconstructed, a bilateral osteotomy of the ileum with adaptation of the symphysis will be required. The osteotomy is performed at the same level as the Salter osteotomy (▶ Chapter 3.2.4) [31]. We secure the result with an external fixator (Compact Hoffmann II) and the symphysis is adapted with non-absorbable sutures (■ Fig. 3.209). The long-term prognosis for patients treated in this way is very good, despite the occasional persistence of an abnormally positioned acetabulum [17].

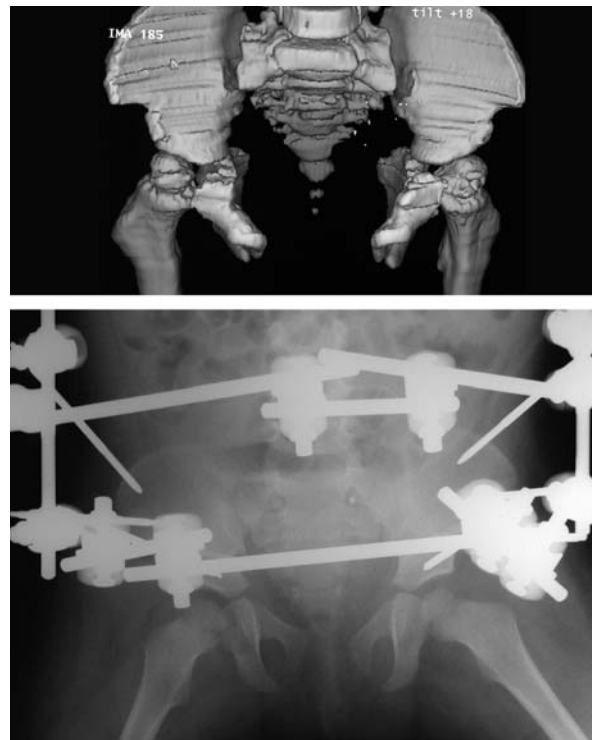
### 3.2.7.2 Changes in the pelvis and hips in systemic illnesses

#### Multiple epiphyseal dysplasia

This illness is discussed in detail in ▶ chapter 4.6.2.11. However, since the hips are generally most affected, and even sometimes exclusively affected, by multiple epiphyseal dysplasia, it will be discussed in some detail here as well.

#### ➤ Definition

Autosomal-dominant hereditary disorder of widely varying severity. The femoral head epiphysis is by far the most commonly and most severely affected.



■ Fig. 3.209. 2-year old girl with *bladder exstrophy* and typical anterior opening of the pelvis. a CT with 3D reconstruction. b Situation after reconstruction of the bladder, bilateral pelvic osteotomy, adaptation of the symphysis and fixation with external fixator (Compact Hoffmann II)

### Classification

We distinguish four forms:

- *Severe form* according to *Fairbank* with delayed appearance of the ossification centers of most epiphyses, with ungainly fingers and toes and moderately stunted growth [7] (▶ Chapter 4.6.2.11).
- *Milder form* according to *Ribbing* with minimal involvement of the fingers and toes, in most cases only the femoral heads are significantly affected [27].
- *Localized mild form* according to *Meyer* with exclusive involvement of the femoral heads (dysplasia epiphysealis capitis femoris) [20, 21].
- *Acetabular form*. This hitherto undescribed form involves irregular ossification of the acetabulum with no involvement of the femoral head (■ Fig. 3.210).

### Occurrence

While the Fairbank type is rare, the milder forms are relatively common (40 cases in 1 million inhabitants).

### Pathogenesis

This condition involves an impairment in the endochondral ossification of the epiphyses. At hip level this can affect the femoral head and/or the acetabulum.

### Clinical features, diagnosis

The condition is not usually diagnosed until early childhood, when the occurrence of hip symptoms prompts the recording of x-rays. The hip symptoms are usually load-related and not particularly serious. Pain can also occur, however, in the milder forms (Ribbing, Meyer) or in the previously undescribed exclusively acetabular form. This latter form is not all that rare in our experience, as we have observed a total of 18 cases over the past 10 years.

The AP x-ray shows delayed, irregular ossification of the femoral head center (■ Fig. 3.211 and 3.212), which is usually widened, although the joint cartilage is not thickened. Coxa vara is also occasionally present. In the purely acetabular form, the femoral head is normal, while the acetabulum is irregular with individual osteolyses (■ Fig. 3.210). The rest of the skeleton is normal in the Ribbing and Meyer types discussed here.

### Differential diagnosis

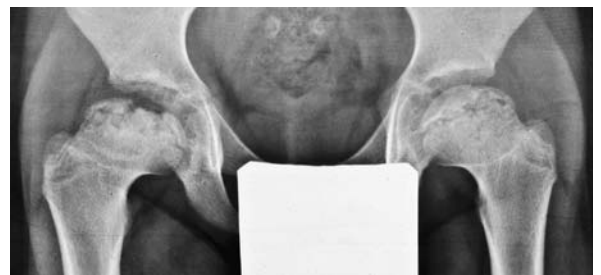
The most important differential diagnoses are Legg-Calvé-Perthes disease and spondyloepiphyseal dysplasia. Differentiating between Legg-Calvé-Perthes disease and multiple epiphyseal dysplasia can sometimes be difficult. As a rule, the involvement of both femoral heads

should always arouse suspicions of epiphyseal dysplasia. Thickening of the joint cartilage is not a feature of this condition, nor do lateral calcification and subluxation occur. In very general terms, the course (at least in childhood and adolescence) is much more benign and the metaphyses are not usually involved. The acetabulum, on the other hand, is usually much more severely affected than in Legg-Calvé-Perthes disease. Certain mixed forms of the condition also evidently occur [26]. The long-term prognosis, i.e. the risk of early osteoarthritis, on the other hand, seems to be more pronounced in multiple epiphyseal dysplasia than in Legg-Calvé-Perthes disease. This is possibly due to the fact that the joint cartilage, in contrast to Legg-Calvé-Perthes disease, is affected by the disease.

Spondyloepiphyseal dysplasia can be ruled out if no vertebral body changes are apparent.



■ Fig. 3.210. Multiple epiphyseal dysplasia of the acetabulum in a 12-year old boy with bilateral hip symptoms. **a** AP x-ray; **b** CT of both hips. Note the irregularities in both acetabula with cyst-like changes



■ Fig. 3.211. AP x-ray of both hips in an 8-year old boy with multiple epiphyseal dysplasia of both hips (Ribbing type)



■ Fig. 3.212. AP x-ray of the right hip of a 3-year old boy with the mild form of multiple epiphyseal dysplasia (Meyer type)

### Treatment

Multiple epiphyseal dysplasia generally follows a benign course. Since it is not significantly affected by therapeutic measures, the orthopaedist is advised not to overtreat the condition. In particular, he should not fall into the trap of misdiagnosing it as Legg-Calvé-Perthes disease and attempting to improve the containment with braces or intertrochanteric varus osteotomies. In fact, the varus osteotomy would be a hazardous procedure since epiphyseal dysplasia often involves a tendency toward coxa vara. By contrast, an intertrochanteric valgus osteotomy is occasionally indicated.

### Prognosis

The long-term prognosis for multiple epiphyseal dysplasia of the Ribbing type is not too good if a flattened, widened femoral head with acetabular changes is present on completion of growth. In such cases, the first signs of osteoarthritis of the hip can be expected even at the age of 30 [32]. On the other hand, the prognosis for the Meyer type is very good since a slight widening of the epiphysis only is observed on completion of growth. Premature osteoarthritis is unlikely to occur [20].

### Other typical malformations of the pelvis and proximal femur in various systemic illnesses

Typical changes in the pelvis and hip are found in a series of congenital systemic disorders (■ Table 3.33):

- Trisomy 21 (■ Fig. 3.213),
- Achondroplasia (■ Fig. 3.214),
- Hypochondroplasia,
- Pseudoachondroplasia,
- Spondyloepiphyseal dysplasia (■ Fig. 3.215),
- Mucopolysaccharidosis (Morquio-Brailsford type; ■ Fig. 3.216),
- Mucopolysaccharidosis (Pfaundler-Hurler type).



■ Fig. 3.214. AP x-ray of the pelvis in a 6-month old boy with *achondroplasia*. Very typical findings are the largely horizontal, irregularly shaped acetabula and the broad iliac wings



■ Fig. 3.215. AP x-ray of the pelvis of a 3-year old boy with *spondyloepiphyseal dysplasia*. The acetabulum is steep and irregular, the bony nucleus of the femoral head is of normal size whereas the cartilaginous portion is enlarged



■ Fig. 3.213. AP x-ray of the pelvis of a 2-year old child with *Down syndrome*. Note the overhanging iliac wings



■ Fig. 3.216. AP x-rays of the hip of a 12-year old boy with *mucopolysaccharidosis of the Morquio type*. While the pelvis is largely normal, the epiphyses of the proximal femur show very irregular ossification

■ Table 3.33. Specific changes on the AP x-ray

Syndrome	Pelvic change	Proximal femur
Trisomy 21	Ilium broader and more overhanging than normal, its base at the acetabulum is much flatter than normal on the lateral side, producing the appearance of »elephant ears«, also known as a »cordate pelvis«. The whole iliac wing appears to be more rotated towards the frontal plane than normal	
Achondroplasia	Acetabulum broad and flat, reduced sagittal diameter of the pelvis, reduced height of iliac wings, narrow sacrum and sciatic notch.	
Hypochondroplasia	The acetabular roof is frequently wider and more horizontal than normal. The greater sciatic foramen is slightly smaller than normal. Reduced sagittal diameter of the pelvis. Changes less pronounced than in achondroplasia	
Pseudoachondroplasia	Dysplasia of the acetabular cups, the Y-lines are wide and ossify at a late stage. The greater sciatic foramina are normal, in contrast with those in achondroplasia. Normal sagittal diameter of the pelvis	Delayed development of the femoral head nuclei with small, fragmented ossification centers. Coxae varae with excessively bulbous metaphyses
Spondyloepiphyseal dysplasia	Few changes at birth. Subsequently irregularities in the area of the acetabulum, poss. also broad and rather steep acetabulum as a result of the large femoral head	<p>Congenital form: Delayed ossification of the femoral heads, which develop in a pronounced abnormal varus position on an excessively short femoral neck. The greater trochanter is displaced in the cranial direction. The femoral heads are normally centered, but flattened with an abnormal pear shape</p> <p>Late form: The changes in the area of the proximal femoral epiphyses resemble those in multiple epiphyseal dysplasia. The cartilaginous part of the femoral head is often enlarged</p>
Mucopolysaccharidosis (Morquio-Brailsford type)	Impaired ossification of the pubis. Ilium broader and lower than normal	Delayed and irregular ossification of the femoral head centers
Mucopolysaccharidosis (Pfaundler-Hurler type)	Ilium normal, acetabula may be rather steep	Possibly delayed ossification of the femoral head centers, epiphysis irregular, femoral neck in valgus position

While very few of these deformities have therapeutic consequences, a knowledge of the malformation can occasionally help in the differential diagnosis of the disorder (details of these disorders are provided in ► chapter 4.6.2).

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## 3.2.8 Neuromuscular hip disorders

*R. Brunner*

### 3.2.8.1 Primarily spastic disorders

#### Functional changes

#### ➤ Definition

Changes in the functions of the hip with no structural deformity and caused by spastic muscle activity.

#### Rotational deformities

##### Internal rotation – Adduction

Functional rotational deviations often exist in primarily spastic locomotor disorders (■ Table 3.34). They occur particularly during standing and walking when muscle tone increases as a result of the upright body position and the requirements for body control. Internal rotation seems to be of multifactorial origin:

- femoral anteversion (▶ Chapters 3.2.3 and 4.2.1),
- spasticity of internal rotators
- indirect effect of triceps surae activity (plantarflexion against the floor resistance produces hip internal rotation, flexion and adduction (unpublished data))
- gait mechanics
- foot instability (valgus foot)

The internal rotation is accompanied by pronounced adduction of the legs. Patients with inadequate postural strength are functionally unstable while standing and press the knees together. Provided the internal rotation and adduction are not excessively pronounced, these patients can derive some benefit from this posture as they are able to maintain a stable upright position without any actual postural motor activity by lowering themselves and pressing the knees against each other. If the knees were in a position of neutral rotation they would simply collapse.

However, the more capable of walking the patients are, the more this inward-turning position proves a hindrance as the knees rub together and catch on each other as the knee or foot swings forward. Furthermore, the joints of the lower limb are often stiff as a result of the spastic muscle contractures and co-contractions and this particularly affects rotation at the hip. The patients therefore have to perform compensatory movements of the spine in order to rotate the pelvis and thus move the legs forward. The malalignment of the lever arms produces an instability in stance which is difficult to cope with for the patients with reduced motor control.

#### Therapeutic options

The correction of internal rotation is part of the correction of all functionally relevant deformities present at the lower extremities. All pathologies must be identified and corrected at the simultaneously. Most patients with spastic pa-



■ Table 3.34. Functional deformities in primarily spastic locomotor disorders

Deformity	Functional benefit	Functional drawbacks	Treatment
Internal rotation/ adduction	Stability while standing	Knees rub together, feet drag behind, instability of the leg due to lever arm dysfunction	Supracondylar derotation, adductor lengthening, Abductor transposition
Abduction/ external rotation	Better hip centering	Loss of ability to walk and stand (risk of dislocation on the other side)	Possibly iliopsoas transposition
Flexion	–	Flexion contracture	Physical therapy



■ Fig. 3.217. AP x-ray of the pelvis in a male patient with severe spastic tetraparesis and a left *abduction contracture*, which has resulted in dislocation of the hip on the other side

ralysis have deformities affecting several joints in the lower extremities, e.g. hip flexion/internal rotation contractures, anteverted hip, knee flexion contractures with contracture of the hamstrings and foot deformities. In auspicious cases, all deformities are corrected simultaneously [1, 18, 33, 42], otherwise any residual deformities will require compensation. Even minor rotational corrections (of 10°–15°) have been found to produce a positive effect.

A gait analysis preoperatively can help disclose any compensatory postural defects, e.g. of the pelvis, and other less obvious deformities. It is not possible to establish the movements of the various body segments at the same time with the naked eye. For example, a pronounced internal rotation may be present on one side with a normal configuration on the other. During walking the patient can turn the pelvis posteriorly towards the side with the internal rotation, thereby causing the defect to appear less pronounced. As a result, the leg without a rotational deformity is likewise internally rotated. In this case, however, only the defective hip should be surgically corrected.

Instrumented gait analysis provides a reliable base for these complex corrections. The multifactorial origin

of exaggerated hip internal rotation is best respected this way, too.

The first step is to correct any bony internal rotation. We prefer today the supracondylar approach, fixed with an AO LCP-plate, as it allows for correction of a knee flexion deformity at the same time, and for immediate weight bearing.

A soft tissue approach involves detachment of the hip abductors (gluteus medius and minimus) at the greater trochanter together with a flake of bone and reattachment with screws to the femur under slight tension and with submaximal external rotation (with the hip flexed at approx. 90°) (Steel operation) [47].

This procedure causes the abductors to produce a concurrent externally rotating effect. The risk of power loss in this operation is low and there have been no reports of the disastrous consequences that can occur after the more extensive adductor procedures. On the other hand, the extent of correction is limited, and we have performed this operation only in a small number of selected cases. Since a major problem is exaggerated tone and spasticity, the therapeutic strategy must focus on *reducing the muscle tone* as much as possible (► Chapter 4.7.2).

In order to correct the additional adduction component, the overactive *muscles*, particularly the adductors, require *lengthening*. In contrast with reports in the literature however [14, 34, 38], these muscles should not be divided as this may result in insufficiency of this muscle group [29] (■ Fig. 3.217). Division of the *intramuscular aponeurosis* is sufficient. The surgeon must also be careful to preserve the obturator nerve in particular. Insufficiency of the adductors manifests itself in a broad-based gait or a broad position in the wheelchair. Both are functionally disruptive. Unfortunately, the adductors cannot be re-stored in the event of postoperative insufficiency.

! **Extensive adductor operations involving a tenotomy and obturator neurectomy involve the risk of uncontrollable and uncorrectable external rotation and abduction deformities (see below) with a loss of the ability to walk and stand.**

A reversible and less invasive procedure is the injection of botulinum toxin. This measure can be repeated and

indicates to the surgeon whether the desired effect can be achieved by weakening the adductors as an alternative to surgery.

One tried-and-tested conservative functional measure designed to preserve abduction is a *nocturnal splint*. In order to preserve abduction during walking, a *hip-guiding orthosis* (e.g. Mancini hip abduction orthosis or SWASH orthosis) can prove successful even without a prior operation.

#### External rotation – Abduction

- External rotational deformities are rare and, in most cases, iatrogenic. Such abnormal positions with 50°–60° or more of external rotation and abduction can sometimes appear grotesque. The deformity is usually caused by the simultaneous division at operation of the adductors, the obturator nerve and the iliopsoas muscle. While this major destruction of important hip stabilizers corrects the functional internal rotation in the immediate postoperative period, the above-mentioned severe deformity, which is almost impossible to correct, can develop in the long term. A corrective femoral osteotomy is a therapeutic option.
- Changes in the frontal plane:  
The adduction contracture is the commonest problem. A distinction must be made between the assessment of abduction with the hips in flexion (adductors) or extension (hamstrings). While abduction of up to 20° is often perfectly possible in a flexion position, and thus poses no impediment to intimate hygiene, abduction with extended hips is much more restricted, frequently with angles of just 10°–15°. If functional

problems exist as a result of this adduction deformity, adductor lengthening and/or lengthening of the hamstrings (by graduated intramuscular division of the aponeurosis without neurectomy) may be indicated.

While recurrences are common during the growth phase (► Chapter 4.7.2), the functional restriction is a good indication for this minor surgical procedure. The injection of botulinum toxin is an alternative to surgery and can both correct the increased adduction during walking (scissor gait) and facilitate intimate nursing care. However, its use to improve hip centering is questionable and probably limited by the intermittent recovery of muscle power.

Destruction of the hip adductors and flexors can often lead not only to an external rotational deformity but also to an abduction contracture. Physical therapy usually proves ineffective in resolving this problem. *Surgical treatment* is indicated if a pronounced functional leg-length discrepancy results from the abduction contracture, if there is a risk of subluxation or dislocation of the contralateral hip or if severe asymmetries exist in the sitting position. The appropriate procedure is the *Campbell operation*, in which the hip abductors are detached from the iliac crest and reattached to the wing of the ilium with the leg in an adducted position.

#### Structural changes

##### ► Definition

Structural deformity of the hip and femur caused by spastic muscle activity (■ Table 3.35).

■ Table 3.35. Structural deformities in primarily spastic locomotor disorders

Deformity	Functional benefit	Functional drawbacks	Treatment
Increased anteversion	Stability while standing	Knees rub together, feet drag behind	Derotation osteotomy
Reduced anteversion	–	–	(Rotation osteotomy)
Flexion contracture	–	Squatting position (walking/standing), hyperlordosis, windswept deformity	Lengthening of hip flexors, posterior placement of the muscles inserting at the iliac anterior superior spine after resection of part of the iliac crest
Extension contracture	–	Sitting restricted	Proximal lengthening of the hamstrings, reconstruction of the dislocated hip
Windswept deformity	–	Instability while sitting, hip dislocation	Splints, correction of bones and soft tissues
Hip dislocation	–	Pain, instability, restricted movement	Joint reconstruction, femoral osteotomy (according to Schanz), head resection, soft tissue release

### Anteverted hip and coxa valga

*Rotational deformities of the femur* are very common in patients with spastic locomotor disorders. The anteversion, which is already more pronounced at birth than in adults, increases still further in such cases. The valgus position of the femur is therefore often overestimated on the AP x-ray as the femoral neck-shaft angle is projected in a very oblique view. The correct view (with just as much internal rotation as anteversion) generally shows an age-matched valgus position of the femoral neck of 135°–140° [26].

Since the growing skeleton is shaped according to the forces acting on it, it must be assumed that the change produced in the daily transfer of forces between the acetabulum and femur as a result of the locomotor disorder is the reason for this rotational deformity. A follow-up study of 102 hips after a derotation varus osteotomy found that the rotational deformity recurred if the remaining growth period was long enough [7]. When the operation was performed on children under 4 years, the surgical correction regressed completely in some patients, whereas hardly any recurrences were recorded after operations on children over 8 years of age [7]. As a rule, however, motor control and gait exert a significant influence on the shape of the proximal femur [27].

Gait analysis studies have shown that the level of the osteotomy, whether intertrochanteric or supracondylar, does not affect the end result [36]. Consequently, we currently prefer the supracondylar osteotomy fixed with aAO low contact plate (LCP). The supracondylar procedure offers the advantage of immediate weight-bearing. As a result, the patient, who is already in a poor training condition, does not lose further power as a result of postoperative immobilization.

The increased inward rotation and abnormal adduction position do not always interfere with function to the same extent. While the knees will knock together in a patient with good walking ability and thus hamper progress, the increased internal rotation may be useful if the patient is only capable of a transfer function or standing. When patients with poor body control and impaired balance reactions try and remain upright but then sink toward the floor, both legs knock against each other and provide mutual stabilization. This just about enables such patients to stand. If the knee faces forward, or even outwards, the patients will fall to the floor without this form of support and thus lose the ability to support the body weight. For these reasons we have ceased our practice of correcting the rotational deformity in the femur to a neutral position, but rather seek to leave a residual postoperative internal rotation position of 5°. Overcorrections must be avoided.

If the legs are in external rotation, the patient must shift his center of gravity in front of the knee in order to

stabilize the leg via external moments. To achieve this he has to incline the trunk forwards and laterally, producing a Duchenne limp that can be misinterpreted as weakness of the abductors.

### Retroverted hip

This occurs in the so-called »wind-swept« deformity, but does not have any functional significance. Treatment: rotation osteotomy if necessary.

### Flexion contracture

The spasticity of the hip flexors together with the frequent flexed posture of the hips not infrequently leads to flexion contractures. The lumbar spine becomes hyperlordosed. Physical therapeutic stretching of the hip flexors may prove successful, although a muscle extension is required in many cases. Botulinum toxin injections can prove helpful temporarily. Although it is generally believed that the iliopsoas muscle is primarily responsible for the flexion contracture, in our experience it is the muscles attached to the anterior superior iliac spine that tend to be contracted. Resection of the iliac crest and dorsal displacement of the spine is an effective way of treating this contracture. Lengthening of the iliopsoas muscle on the other hand is not without its risks, since the muscle loses strength as a result, and the patient suffers a decline in walking speed that cannot subsequently be recovered. Iliopsoas lengthening procedures should therefore be viewed with caution.

### Extension contracture

While this deformity is described in the literature, the cause in our patients has always been a ventral hip subluxation or dislocation.

### Windswept deformity

When hip flexion contractures are present, gravity forces the flexed knees downward on the side on which the muscle tone is strongest. Since the patients often remain fixed in this position asymmetrical contractures can form accordingly: on the one side there is flexion, adduction and internal rotation, while on the other there is flexion, abduction and external rotation. Instability results during sitting, because the patient tends to fall to the side over the adducted and internally rotated hip. This joint is also at great risk of dislocation. Possible treatment includes physical therapy and splints. If functional disability is present, the deformity must be corrected by interventions on the bones and soft tissues.

### Hip dislocation

#### Radiological investigation

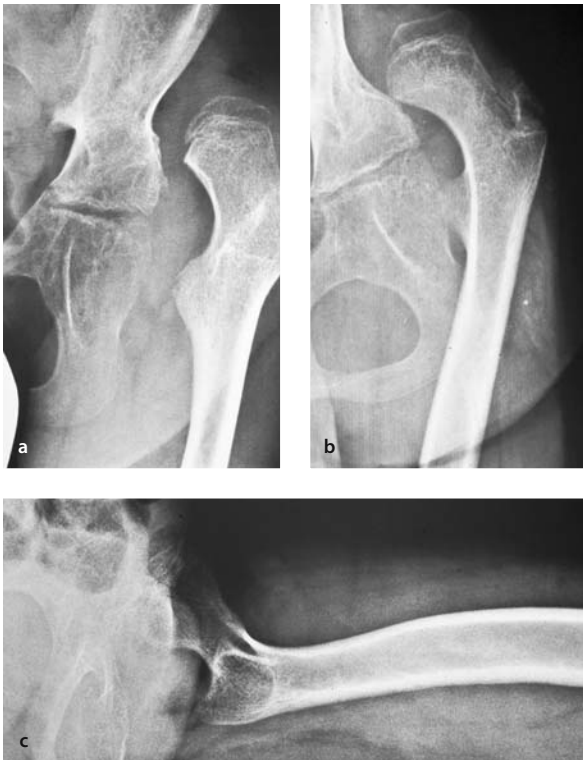
An AP view with suspended lower legs generally permits effective evaluation of the hip situation.



■ **Fig. 3.218.** *Apparent acetabular coverage* on a AP x-ray of the pelvis. The right hip is adducted and appears to have poorer acetabular coverage than the abducted left hip. However, the lateral acetabular epiphysis is pointed on both sides



■ **Fig. 3.220.** In a case of pronounced *anteversion* the rotated position of the hips is important for the centering: In neutral rotation (*top*) both hips appear dislocated, while in internal rotation (*bottom*) they are centered (the two x-rays were recorded immediately after each other)



■ **Fig. 3.219a–c.** Valgus femur (a) on AP view. b Near-normal femoral neck-shaft angle on the *corrected view* (internal rotation according to the angle of the anteversion). c The Dunn view confirms the substantial anteversion

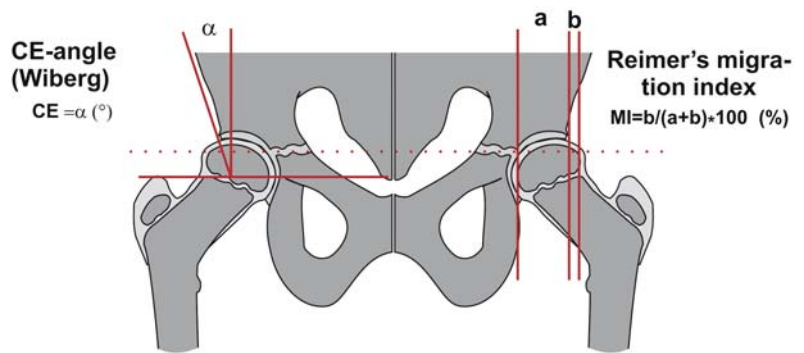
#### Mistakes in the evaluation of the standard AP x-ray

- Adduction of the leg appears to aggravate the centering of the hip (■ Fig. 3.218).
- A purely ventral or dorsal dislocation can be associated with a normal AP x-ray (a three-dimensional CT-scan will show the pathology best).
- A neutral rotation with pronounced anteversion projects the femoral neck in a valgus direction (■ Fig. 3.219).
- A neutral rotation with increased anteversion aggravates hip centering (■ Fig. 3.220).
- A flexed hip with raised knees can be mistaken for a coxa vara.
- A hip flexion contracture without raised knees distorts the acetabula, thereby aggravating with the evaluation of the hips.

#### Other useful views

- The *Dunn-Rippstein view* for the evaluation of *anteversion* (although this method is associated with a wide margin of error, it is effective in clinical practice). Thanks to the flexion of the hips the *acetabula* can readily be evaluated even if flexion contractures are present;
- the *faux-profil view* for anterior dislocations;





■ Fig. 3.221. Measurement of hip centering on the x-ray: center-edge angle according to Wiberg (CE) and migration index according to Reimers (MI)

— CT with three-dimensional reconstruction for unclear dislocation directions or for detecting an anterior or posterior dislocation.

**Hip centering measurements:** The standard measurements are the *center-edge (CE) angle according to Wiberg* (normally over 15°) and the *Reimers migration index according to Reimers* (normally below 22%) [37] (■ Fig. 3.221). Both measurements are based on the AP x-ray and can only record the lateral component of a dislocation. The latter method in particular is generally used nowadays for evaluating hip x-rays.

❗ **Normal results can sometimes be measured for purely anterior or posterior dislocations! Radiographs only show the component of a dislocation that is directed parallel to the x-ray plane.**

The *appearance of the lateral acetabular epiphysis* has also proved an effective indicator: If the lateral acetabular epiphysis is rounded off at the edges the hip must be considered to be at risk despite good centering on the AP view [11]. By contrast, a lateral acetabular epiphysis with sharp edges is an indication of a stable situation even if the joint is slightly off center (■ Fig. 3.218; 3.222).

### Frequency and occurrence

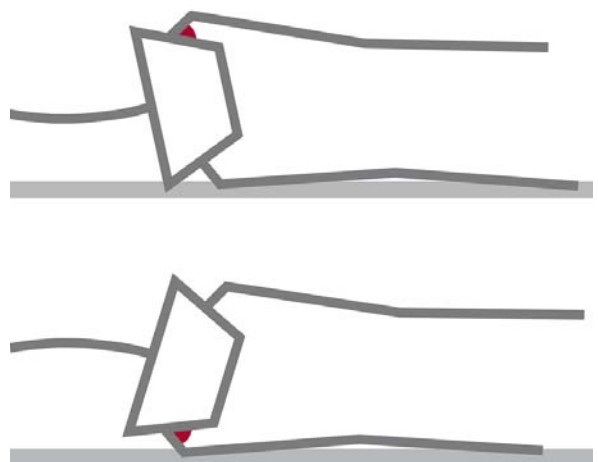
The worse the motor control of a patient's locomotor apparatus, the more likely it is that hip dislocation will occur. In cases of severe spastic tetraparesis the frequency is as high as 60–70%, whereas dislocations are rare in spastic hemiparesis or diparesis (7%) [24].

### Pathogenesis

Dislocation develops as a result of the exertion of constant pressure by the femoral head against the lateral acetabular epiphysis, often as a result of the unfavorable positioning of the patient. If the patient lies on his side, one hip will be constantly adducted, while the other will be abducted (■ Fig. 3.223). Patients with severe cerebral locomotor disorders move very little and tend to lie in the same position for prolonged periods. Therefore, if the patient remains



■ Fig. 3.222. The rounded lateral acetabular epiphysis on the left side (arrow) is an at-risk sign for dislocation, even in a well centered joint



■ Fig. 3.223. Situation of the hips in the lateral position: The patient's legs press against each other under the influence of gravity and tilt the pelvis obliquely. The spasticity determines which hip is primarily held in adduction



in this unfavorable position with adduction, sustained pressure is exerted on the lateral acetabular epiphysis. Another dislocation-promoting factor is the absence of motor control and thus the non-functional dynamic hip stabilizers. Poor proprioception may be an additional factor, resulting in inadequate or lacking motor responses on ligamento-capsular stress.

### Preventive measures

The purpose of preventive measures is consistently to avoid the unfavorable positioning of the patient. This can be achieved by the use of *nocturnal positioning aids*, which keep the patient in a supine or prone position. The legs are held loosely in slight abduction, slight internal rotation and possibly slight flexion. Nocturnal positioning aids are indicated if the x-ray suggests that the hip is at risk. If the hips are already dislocated, such braces can prove painful and are no longer capable of centering the joint. Abduction braces can even be harmful and cause pain in these situations as they increase the intraarticular pressure without reducing the hip joint.

In most cases the hip adductors are considered to be the cause of the dislocation because abduction is already restricted in subluxation and the adductors appear shortened. If the centers of rotation of the head and acetabulum do not match, the gap between the points of muscle insertion and origin will increase. In this position the length of the adductors is too short relatively, thereby preventing the abduction required for centering of the joint. However, no direct evidence can be inferred from this for a causal component for the hip dislocation. Nor should the fact be overlooked that the hip adductors also stabilize the hip and the absence of this muscle group increases joint instability. However, adductor operations are often advocated in order to prevent dislocations [14, 22, 23, 34, 38].

While the prophylactic effect of an adductor tenotomy has been demonstrated in large patient populations, a closer analysis reveals that the indications in these studies varied considerably. They are often based on measurements of the lateralization of the femoral head, in some cases on incorrectly recorded x-rays. It is not certain whether these hips would also dislocate without treatment. On the other hand, our experience has shown that a dislocation can be prevented in at-risk hips only in isolated cases. However, the harm resulting after adductor division or destruction, with external rotation contractures and hip instability, is far greater than that resulting from the actual dislocation of the hip, since the dislocated hip can be reconstructed, whereas the destruction of the adductors is irreversible [29]. Furthermore the abduction deformity of the operated hip forces the contralateral hip into an adduction position, potentially resulting in a dislocation on that side. A constant abduction limits the patient's ability to make motor progress as standing and walking is difficult in this position.

Prophylactic iliopsoas operations can also be helpful. Since such soft tissue procedures are not without their own problems however, we adopt a cautious approach, particularly as regards the adductors, and consider such surgery to be indicated only in patients with radiological changes to the lateral acetabular epiphysis without decentering of the head, those with a pronounced adduction spasticity and those whose abduction is limited to less than 10°. An adductor operation may prove helpful in these cases. We restrict ourselves strictly to the aponeurotic lengthening of the adductor longus and magnus and gracilis muscles. The short adductor muscle should be preserved as a hip stabilizer, and the obturator must be spared. If necessary, the psoas muscle can be lengthened, likewise at the aponeurosis, at the high intrapelvic level.

While the injection of botulinum toxin into these muscle groups does represent an alternative solution, it is less efficient than surgery since the muscle power recovers each time between the injections. Despite the correction of these mechanical factors, it is often not possible to prevent a dislocation for prophylactic reasons. We therefore consider another important factor, in addition to the motor disorder, to be the sensory impairment that is also usually present (► Chapter 4.7). This makes the patient incapable of sensing tension situations in the capsule and thus activating the muscles dynamically to stabilize the joint.

### Course and development of hip dislocation

The consistent application of the same type of pressure and in the same direction by the femoral head on the lateral acetabular epiphysis causes the latter to roll out and be pushed away, producing a *groove-shaped deformity* in the acetabulum (■ Fig. 3.224) [11], known as elongation of the acetabulum.



■ Fig. 3.224. Three-dimensional reconstruction of a dislocated hip in infantile spastic cerebral palsy: The anterior and posterior rims of the acetabulum are clearly visible. The head has slipped out of the cup in a single direction along a groove-shaped spur. The indentation can be seen on the lateral side of the head and is caused by the reflected part of the proximal tendon of the rectus femoris muscle



■ Fig. 3.225. Clinical appearance of a bilateral *anterior hip dislocation* in a female patient with infantile spastic cerebral palsy. Note the skin protrusions over both hips caused by the anteriorly dislocated femoral heads

The femoral head initially moves up and down in this groove until it becomes fixed at its upper end, eventually forming a *secondary acetabulum*. The anterior and posterior edges of this groove, which correspond to the anterior and posterior rims of the acetabulum, are generally intact. This groove most commonly points laterally in a sector between 25° ventrally and 30° dorsally [6].

*Genuine ventral* or *dorsal dislocations* do occur but are rare. *Ventral dislocations* account for approx. 8% of all dislocations in our patients (■ Fig. 3.225), and are more common in patients with muscle hypotonia or flaccid paralysis. *Dorsal dislocations* are even rarer (approx. 1%), and generally occur postoperatively.

### Pathological anatomy

The head is pushed out of the cup as it forms a groove in the acetabulum, resulting in a unidirectional instability along this groove. In younger children the head epiphysis grows increasingly in the lateral direction (»head in neck«), while an indentation forms in older children under the reflected part of the proximal tendon of the rectus femoris muscle. At first the joint cartilage is missing from this section, while a bony furrow forms at a later stage (■ Fig. 3.224).

### Symptoms

The decentering of the hip can result in severe pain, even in cases of mild subluxation [3, 6]. At operation, this pain not infrequently correlates with a substantial effusion and,

occasionally, pronounced synovitis. The instability of the hip leads to a restriction, or even a loss, of the ability to stand and walk, and prevents any further motor progression [3, 6]. The dislocated hip becomes stiff and the long lever arm, together with the reduced weight-bearing as a result of the dislocation, leads to a much greater risk of fractures.

But the dislocated hip also appears to produce general symptoms such as a non-specific malaise or an undefined dull pain in the joint that diverts the children's attention and makes them tired. The general activity level of these children, which is already restricted, thus declines still further, whether in school, family situations, or ultimately even while eating. It is always amazing to see, again and again, how such general and non-specific skills of the patients can be improved by the treatment of the dislocated hip. In a unilateral dislocation, the pelvic obliquity can also result in scoliosis [15].

### Therapeutic options

The conservative treatment of a dislocated hip simply involves acceptance of the dislocation and management of the pain with analgesics. It is important that any seating aids are adapted to the specific movement restriction. This conservative approach may be indicated for severely disable patients who are in a very poor general condition. Abduction should be avoided as it may cause pain by pressing the dislocated femoral head against the pelvis.

Pain usually results from the combination of the dislocation/subluxation and spasticity. Thus either the decentering of the hip can be corrected (see below) or the spasticity can be treated. One possible first-line treatment is Lioresal (baclofen), administered orally or intrathecally (»tone control«; ► chapter 4.7).

Taking the weight off the hip is, in our experience, not beneficial, since this only restricts the training level and mobility of the patient without improving the condition of the hip. We therefore merely recommend that painful situations should be avoided and the patient should be left to continue his rehabilitation program unchanged regardless of the hip. Any hip problems that arise will, of course, have to be resolved accordingly.

Anterior dislocations are particularly awkward. In such cases the femoral head can press directly on the femoral nerve, leading to severe pain. Surgical correction is then unavoidable, even if the prognosis for this particular form of dislocation is poor (see below).

### Surgical approach

A surgical approach is *indicated* if the hip dislocation produces symptoms. Early operation is technically easier since the deformities are less pronounced. The prognosis for redislocations, however, is independent of the grade of dislocation.

The type of *operation* is often selected on the basis of the child's motor skills. However, this approach is highly problematic since the motor skills can be hampered considerably by the actual dislocation. Consequently, such children are often underestimated, and even severely disabled children have at least been able to recover the ability to stand following appropriate surgical procedures. Even this minimal skill can help improve daily nursing care significantly. While the postoperative capability cannot be predicted for severely disabled patients with a dislocated hip, we have not found any disadvantages resulting for our patients as a result of the operation [6]. We therefore consider that surgery is indicated also for severely disabled children.

### Surgical procedures

#### — Soft tissue release:

While the hip can basically be recentered by a soft tissue release of varying degree [45], the division of the soft tissue regularly leaves behind a dynamic instability in the joint and not infrequently results in contractions (often an abduction/external rotation contracture). Furthermore, the acetabulum in older children has little further opportunity of spontaneously correcting its abnormal shape. Children with motor disabilities are unable to compensate for the loss of function of the divided muscles. This procedure will deprive them of the opportunity, possibly even in the short term, to recover the ability to walk or stand.

#### — Resection of the femoral head:

There are various techniques for resecting the femoral head and inserting either the femoral neck, shaft or lesser trochanter into the acetabulum [2]. The best results are achieved with the infracondylar re-

section technique. The interposition of muscle as a buffer has a positive effect [4]. But the disadvantage associated with all these femoral head resection procedures is that they produce significant instability in the hip and leg shortening. Patients with poor coordination and a poor sense of balance will thus be deprived of the ability to maintain a standing position. Moreover, periarticular ossification is common and can itself lead to stiffening of the hip and to pain. In our experience this procedure is indicated only in extreme cases or after other treatments have failed. Actual freedom from pain cannot be guaranteed however. Implanting the humeral part of a shoulder endoprosthesis may improve the outcome as it avoids rubbing of the femoral shaft within the acetabulum.

#### — Angulation osteotomy according to Schanz:

In this operation the proximal section of the femur is placed in a valgus position (usually more than 45°), thereby eliminating the pain associated with painful hips because the head is no longer pressed against the pelvis. Stability while walking is also improved compared to the head resection, although mobility will continue to be restricted. Any subsequent total hip replacement is made more difficult, and freedom from pain is not always guaranteed with this method.

#### — Reconstruction of the hip:

The dislocated can be surgically reconstructed. A femoral derotation varus osteotomy together with soft tissue-balancing measures may suffice. In most cases, however, the acetabulum does not recover sufficiently further dislocations and subluxations are the result [7]. The overall results are better when all existing deformities of the pelvis and femur are corrected [3, 6, 8, 12, 25, 26, 31, 32, 34, 46]. The opera-



■ Fig. 3.226. Bone corrections for the reconstruction of a dislocated hip in infantile spastic cerebral palsy: The femur is shortened, derotated and placed in a varus position. The iliopsoas muscle and lesser trochanter are detached. The surgeon chisels around the acetabular groove and, after open reduction, turns down the acetabulum in this area. The gap is filled with the removed wedge of bone. After fixation of the femur, the lesser trochanter is secured to the planum trochantericum (the iliopsoas transfer is only done in special indications, such as anterior dislocation)

tion consists of a combination of a correcting femoral osteotomy (with shortening, derotation and varization), a modified Dega-type acetabuloplasty (or, in rare cases, a Salter or triple osteotomy), open reduction with resection of the femoral head ligament and dissection of the transverse acetabular ligament and lengthening or transfer of the iliopsoas muscle [7, 8, 17, 43] (■ Fig. 3.226 and 3.227).

Our experience has shown that the transfer of the iliopsoas to the planum trochantericum provides ad-

ditional stability to hip. We regularly perform these steps in a *single session* on patients with poor coordination and severe spasticity. If the adductors are still contracted at the end of the operation, they are lengthened at the aponeurosis.

Lengthening of the hamstrings may also provide additional stability for the hip, although we have not yet had to perform this procedure for this reason. The patient is immobilized postoperatively in a hip spica or an abduction brace for 2–3 weeks in order to alleviate the pain. This treatment usually results in fairly mobile hips (flexion of 100°, extension of up to approx. 5°–10° flexion contracture), slightly increased internal rotation compared to external rotation, and slightly restricted abduction. This restriction increases the further laterally the acetabular roof has been reconstructed.

This is beneficial, however, in severely disabled patients in order to minimize the tendency toward further dislocation. In ventral dislocations, the iliopsoas transfer is particularly important as this muscle is then located over the ventrally dislocating head, pushing it back into the acetabulum when tensed.

Apart from the general surgical *complications* (anesthesia, infections), the possibility of osteoporosis-induced [9] and heterotopic ossification should be borne in mind, although we have only observed one case of the latter in 450 hip reconstructions. The evaluation of the functional results revealed a reduction in pain in all patients as a result of the operation, most of whom were completely pain-free.

A preliminary analysis of our results showed that the postoperative absence of pain was related to the bone age of the patient: If we operated while the triradiate cartilage was still open, hardly ever pain occurred, whereas the pain persisted in 6% of cases if the triradiate cartilage was closed at operation. The number of patients capable of walking was higher postoperatively and sitting problems were less frequent [6]. Redisllocations can be expected in approx. 6–7% of cases. The redisllocation rate is very much higher, at approx. 30%, for anterior dislocations.



■ Fig. 3.227a, b. Female patient with infantile spastic cerebral palsy. **a** Left hip dislocation. **b** Result 5 years after *hip reconstruction* (the patient is able to walk again with an ambulator, which had not been possible before the operation)

■ Table 3.36. Functional deformities in primarily flaccid locomotor disorders

Deformity	Functional benefit	Functional drawbacks	Treatment
Abduction/external rotation	–	Loss of ability to walk and stand, risk of anterior dislocation of the hip	Corrective femoral osteotomy, Campbell operation. Transfer of the external oblique abdominal muscle
Extensor insufficiency	–	Flexion contracture	Posterior iliopsoas transfer (Sharrard)
Flexion	–	Flexion contracture	Physical therapy

### 3.2.8.2 Primarily flaccid locomotor disorders (myelomeningocele, paraplegias)

#### Functional deformities

##### Definition

Changes in the functions of the hip with no structural deformity and caused by reduced or absent muscle activity (■ Table 3.36).

#### Abduction and external rotation contractures

Despite the differing levels at which neurological paralysis occurs, an external rotation and abduction contracture of the hip is common. While the external rotators are innervated only by the low lumbar and upper sacral roots, the nerve supply to the hip internal rotators is distributed across all the lumbar and upper sacral segments.

According to the theory that an active agonist contracts when countered by a flaccid antagonist, the patients would be expected to show an internal rotation and adduction contracture. However, this is almost never the case, probably because the patients generally lie on their back with their legs slightly flexed and spread apart, and the force of gravity pushes these flexed legs down in an external rotation and abduction movement. This produces long-term overstretching of the internal rotators which, though they may be innervated, are not overactive. The inactive external rotators become contracted. However, this functional deformity results in an unfavorable gait pattern with external rotation and abduction at hip level and correspondingly externally rotated knees. This involves the added risk that the hips will dislocate anteriorly as a result of constant overstretching of the anterior structures.

#### Conservative treatment

The sinking of the legs into an external rotation and abduction position can be prevented with positioning aids.

Cushions, splints or wedges can be used for this purpose. As a simple measure the legs can also be fixed in relation to each other at night.

#### Surgical treatment

- *Campbell operation*: The hip abductors are detached from the ilium at their origin and transferred distally. The risk of recurrence is high without consistent follow-up treatment, although the procedure can be repeated if necessary.
- The *external oblique abdominal muscle* can be mobilized, rolled up and attached to the femur in order to stabilize and improve adduction and, if necessary, flexion as well [35]. However, this *muscle transfer* has two major problems: Firstly, the transferred muscle is missing at its origin (making the abdominal press more difficult for these patients), and secondly, the anatomical structure of the muscle differs so much from that of the other flexor muscles that the performance of the muscle is very restricted. For this reason this transfer is only indicated in rare cases. We do not have any personal experience with this procedure.
- If an extensor insufficiency is present, as commonly occurs in association with flexion contractures and a dynamic deficit while walking, the iliopsoas muscle can be transferred dorsally to serve as an extensor. The results of this dorsal transfer are considered to be good, although normal hip function cannot be expected. We ourselves do not have any experience with this transfer procedure.

! As a rule, muscle transfers should be approached with caution.

#### Structural deformities

##### Definition

Structural deformity of hip and femur caused by reduced or absent muscle activity (■ Table 3.37).

■ Table 3.37. Structural deformities in primarily flaccid locomotor disorders

Deformity	Functional benefit	Functional drawbacks	Treatment
Increased anteversion	Stability while standing	Knees rub together, feet drag behind	Derotation osteotomy
Reduced anteversion	–	(Instability in upright position)	(Rotation osteotomy)
Flexion contracture	–	Squatting position (walking/standing), hyperlordosis	Lengthening of hip flexors
Extension contracture	–	Sitting restricted	Proximal lengthening of the hamstrings, reconstruction of the dislocated hip
Hip dislocation	–	Instability, restricted mobility, pelvic obliquity	<2 years: conservative, >3 years: reconstruction of the joint (controversial if both hips are involved)



### Coxa valga and anteverted hip

While rotational deformities such as coxa valga and anteverted hip are rarely a problem in patients with myelomeningocele, the basic approach does not differ significantly from that for spastic locomotor disorders.

### Retroverted hip

A pronounced and fixed external rotation can cause retroversion. If functional impairment is present this can be corrected by a femoral osteotomy.

### Hip dislocation

The dislocation of the hip in myelomeningocele results from the combination of diminished sensation around the joint and the non-functional dynamic stabilizers. The patients want to be able to take part in the same activities enjoyed by children of the same age without neurological disorders. They stress their joints by adopting extreme positions (for example they really throw their legs around during transfer activities) and thereby constantly overstretch the joints. This overstretching is not sensed, and there is no pain reaction to stimulate the already weakened, or even absent, dynamic joint stabilizers into action. As a result, the joint becomes deformed and eventually dislocates.

A neurogenic hip dislocation can even occur inside the uterus. One or both hips are then dislocated at birth, and the sonographic finding is compatible with that in congenital hip dysplasia or teratogenic dislocation. A dislocation groove may even be detected at this stage during arthrography. Dislocations are common in cases of flaccid paresis (up to 80%) [39], while the frequency in myelomeningocele depends on the neurological level involved [5]: L1/L2: 30%, L3: 36%, L4: 22%, L5: 7%, S 1: 1%. Other authors have reported higher rates: L1/L2: 50%, L3 and L4: 75% [21, 41].

### Treatment

#### Conservative treatment

Conservative treatment offers almost no prospect of success since it cannot ensure dynamic stabilization of the hip.

**! Treatment with the Pavlik harness is contraindicated as this treatment relies on the child thrashing about and assumes normal, unimpaired motor function, a requirement that is not satisfied in those with myelomeningocele.**

**Dislocations at 3 years of age or older:** While the inherent ability to walk does not correlate with the centering of the hips [16, 20, 44], patients with successfully operated hips show functional improvement [2]. Unilateral hip dislocations can lead to a pelvic obliquity requiring treatment [13, 20]. On the other hand, we have seen in our own

patients how hips with pathological changes as early as the first year of life are at greater risk of redislocation, compared to dislocations that occur in later life (unpublished data). We therefore recommend reconstruction of the hip only if walking deteriorates as a result of a hip dislocation in initially normal hips.

Astonishingly, dislocated hips in MMC patients hardly show any significant restriction of movement. In fact, we have never had to implement therapeutic measures in our patients for this reason. By contrast, we have observed that centered joints are much more frequently associated with functionally relevant restriction of movement. Reconstruction therefore involves the risk of producing a worse scenario than with the dislocated hip.

### Surgical approach

As with spastic locomotor disorders, the reconstruction of the hip involves the correction of the deformities of the acetabulum and femur and open reduction. An iliopsoas transfer is not performed for lateral and dorsal dislocations. With ventral dislocations, the transferred iliopsoas helps keep the head in the acetabulum. However, an iliopsoas transfer also involves the risk of a subsequent loss of hip flexor power.

Again as with spastic locomotor disorders, the *post-operative management* involves a hip spica, followed by nocturnal positioning orthoses. The reconstruction of the hip can be repeated if necessary. Some of our own patients have required recentering of the hip up to 3 times. It is advisable, therefore, to wait until the skeleton is adequately developed before performing the first procedure to correct the deformities. Recentering on its own, without correction of the acetabulum, has little prospect of long-term success.

The main *complication*, apart from infections and the anesthetic risk, is the occurrence of periarticular ossification. In contrast with other underlying neurological disorders, this is relatively common with myelomeningoceles. We have observed isolated cases although the ossification was never functionally relevant. The *alternative* to hip reconstruction is to leave the hip dislocation and adapt the braces accordingly. This involves stabilization of the hip with an orthosis and, for a unilateral dislocation, offsetting the leg-length discrepancy.

### Muscular dystrophies

#### Functional problems

In muscular dystrophies and spinal muscular atrophies, muscle power is progressively lost. The patients therefore require hyperextension at the hip in order to be able to stand passively in the ligamentous apparatus. Flexion contractures are disabling at this stage as they can lead to the premature loss of the ability to walk and stand. These must be corrected, concurrently with other contractures, in good time (■ Table 3.38, ► chapter 4.7.5).

■ Table 3.38. Functional deformities in muscular dystrophies

Deformity	Functional benefit	Functional drawbacks	Treatment
Abduction/ external rotation	–	Loss of ability to walk and stand	Campbell operation
Flexion	–	Flexion contracture	Physical therapy

■ Table 3.39. Structural deformities in muscular dystrophies

Deformity	Functional benefit	Functional drawbacks	Treatment
Flexion contracture	–	Crouching position (loss of ability to walk and stand), hyperlordosis	Lengthening of hip flexors
Hip dislocation	–	Instability, restricted mobility, pelvic obliquity	Head resection, Schanz osteotomy



■ Fig. 3.228. AP x-ray of the pelvis in spinal muscular atrophy with left hip dislocation

### Structural changes

Hip dislocations occur as a result of the failure of the dynamic stabilizers (■ Table 3.39; ■ Fig. 3.228). A scoliotic spinal deformity is another possible causative factor, hence also the benefit of early correction of the spinal deformity. Since no spasticity is present, pain is rare. By the time of the dislocation, most patients have already lost the ability to walk and stand.

If pain does occur, however, as a result of the increased pressure between the head and pelvis, the Schanz angulation osteotomy or head resection is the method of choice. These procedures can produce freedom from pain with little effort, even in patients who are in a generally poor condition.

Occasionally soft tissue-related symptoms are also caused by the resting of the dislocated head on the supporting surface in a bed or chair without corresponding

padding. In such cases, corresponding bedding will suffice to alleviate the pain.

### Post-polio syndrome

#### Functional deformities

The commonest functional deformities are contractures, particularly flexion contractures. These restrict the patient's ability to walk and increase the energy expended during walking because the knee flexion position requires compensatory postural work by the knee extensors to maintain an upright posture. These contractures usually have to be considered in connection with other problems of the lower extremity (knee flexion) so that a corresponding strategy that deals with all the problems together can be prepared.

#### Structural deformities

As a result of the reduced usage, the skeleton of the affected extremities becomes thinner and smaller. Since it is incredibly robust, however, fractures do not occur with greater frequency. Deformities of the proximal femur are often extreme rotational, varus or valgus deviations. But provided no functional restriction results, such structural deformities are of no concern. Correction is required, however, if the patient's function is impaired.

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## 3.2.9 Fractures of the pelvis, hip and thigh

C. Hasler

### 3.2.9.1 Pelvic fractures

#### Occurrence

5% of all fractures in children occur in the area of the pelvis, hip and thigh.

❗ **The considerable elasticity of the pediatric pelvis, the high proportion of shock-absorbing cartilage and the limited presence of high-energy traumata in children explain the rarity of these injuries and the frequency of concomitant abdominourgenital lesions.**

#### Diagnosis

##### Clinical features

External bruising, pain on pelvic compression, pain and hematomas in the perineal region, leg-length discrepancies, asymmetrical contours, pain on hip movement, and concomitant nerve lesions of the lumbosacral plexus, sciatic, femoral and obturator nerves are signs of pelvic fractures. While life-threatening hemorrhages are rare, it should nevertheless be borne in mind that a child's circulation can compensate for even major blood losses over a prolonged period. In view of the frequent presence of concomitant urogenital and abdominal injuries, which can accompany even relatively minor bone injuries, a multidisciplinary approach is essential. A pelvic fracture must be assumed in a child with polytrauma until evidence to the contrary has been found.

##### Imaging investigations

- **Radiography:**  
*Pelvic AP x-ray.* The ala and obturator view should not be x-rayed routinely in view of the additional radiation exposure. Compared to adults, both the symphysis and the iliosacral joint space are wider in children, which can give rise to misinterpretations. The same applies to the ischiopubic synchondrosis, which can be seen on radiographs in around half of 7-year olds. Correlation with the clinical findings is crucial, following the principle »if all else fails, examine the patient!«
- **Computer tomography** is indicated if doubt exists about the indication for surgery and for surgical planning, e.g. in acetabular fractures. Otherwise the standard x-ray on its own can provide sufficient information [14].

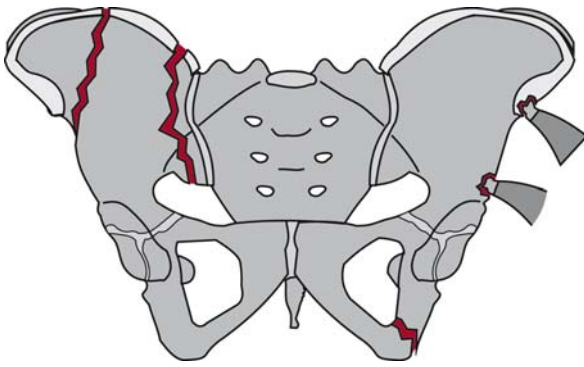
##### Fracture types

- **Type I: Apophyseal avulsion fractures** occur as a result of sudden muscle contractions, rarely as a result of long-term repetitive traction. The growth cartilage

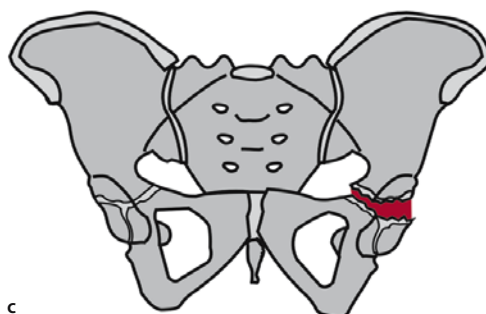
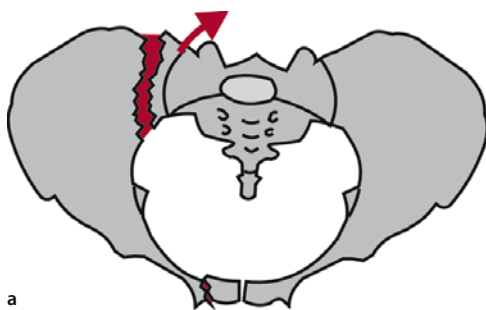
shows reduced mechanical resistance during puberty. As a result, and in combination with the greater willingness to take risks particularly among male adolescents, these avulsion fractures occur primarily during abrupt changes of direction, impact with the ground, etc., classically during a soccer match. The apophysis of the anterior inferior iliac spine (rectus femoris muscle) closes before that of the anterior superior iliac spine (sartorius and tensor fasciae latae muscles) and is therefore affected less often. The apophyses of the anterior superior iliac spine and the ischial tuberosity (biceps femoris, semimembranosus and semitendinosus muscles; ■ Fig. 3.229) are avulsed with roughly equal frequency.

- **Type II: Extra-articular iliac fractures (iliac wing)** are much more common than in adults and explain why pelvic ring fractures occur less frequently as a result of trauma energy acting on the ilium (■ Fig. 3.229).
- **Type III: Stable, single pelvic ring injuries** involve, in ascending order of frequency, isolated iliosacral joint subluxations and fractures, isolated traumatic rupture of the symphysis pubis, ischial fractures and pubic ramus fractures. These can affect one ramus (usually the upper ramus), two contralateral or even two ipsilateral rami. While the latter appear relatively trivial on the x-ray, they are frequently complicated by additional injuries. Even in a case of severe dislocation a second fracture of the pelvic ring is not necessarily present thanks to the intrinsic elasticity of the symphysis and the iliosacral joint. Symphysis injuries do not tear the fibrocartilage, as in adults, but correspond to a cartilaginous avulsion at the transition with the bone and are therefore associated with fewer complications.
- **Type IV: Unstable pelvic ring fractures** are rare. Bilateral, double pubic ramus fractures or rupture of the symphysis pubis with double ipsilateral ramus fractures (»straddle fracture«); rupture of the symphysis combined with fracture/dislocation of the elements behind the ipsi- or contralateral acetabulum (»Malgaigne fracture«; ■ Fig. 3.230).
- **Acetabular fractures** account for less than 10% of all pelvic fractures during the growth phase and are primarily seen in adolescents. Morphologies:
  - Type A: acetabular rim fractures associated with hip dislocation,
  - Type B: linear fractures without instability combined with pelvic fractures,
  - Type C: linear with hip instability,
  - Type D: central fractures with central femoral head dislocation [17].

In types B–D the triradiate cartilage can also be affected with increasing frequency and severity. Types A, C and D represent concomitant lesions accompanying hip dislocations (■ Fig. 3.230).



■ Fig. 3.229. Pelvic injuries without serious late sequelae: Pelvic wing fractures and isolated ilium fractures (right half of the pelvis), isolated pubic ramus fractures and avulsions of the anterior inferior and superior iliac spine (left half of the pelvis)



■ Fig. 3.230a–c. Pelvic injuries with possible serious late sequelae. a Unilateral combination injuries of the posterior and anterior ring. b Bilateral combination injuries of the posterior and anterior ring. c Acetabular fractures)

- Sacral fractures and fractures of the coccyx are probably more common than is generally assumed, since they are very difficult to detect on an x-ray. Clinical diagnosis is therefore particularly important and is based on local tenderness and swelling, usually over the lower part of the sacrum, and pain on rectal palpation.
- Stress fractures of the pelvis are unusual and affect the inferior pubic ramus.

### Treatment

- ❗ **The therapeutic strategy for pediatric pelvic fractures depends to a large extent on the concomitant local and remote injuries. The bone lesions themselves rarely require surgical management.**

### Spontaneous correction

Spontaneous correction can be expected to occur after dislocated pubic ramus fractures and acetabular rim fractures in children under 10 years of age.

### Conservative treatment

Unless concomitant injuries dictate otherwise, a combination of analgesic medication, several days' bedrest in a comfortable position (e.g. lateral position for ruptures of the symphysis pubis), possible traction prior to mobilization with crutches and non-weight-bearing for 3–4 weeks.

- *Type I:* Apophyseal avulsion fractures usually heal with impressive callus formation, which regresses again within a year. Meanwhile the patient should be informed that any palpated lumps are harmless so as to avoid any unnecessary procedures. In very rare cases, however, impingement can subsequently occur during flexion movements between an anterior inferior iliac spine fracture that has consolidated too low down and the femoral neck. Callus formations after avulsion of the ischial tuberosity may be painful when sitting and therefore sometimes warrant surgical removal
- *Type II, ileum:* Thanks to the good circulation and its embedding in the muscles, the ileum consolidates within 3–4 weeks, even with comminuted fractures.
- *Type III, stable, single pelvic ring fractures.*
- *Acetabular fractures: Type A:* the fragments are usually small, but even fairly large rim fractures remodel themselves in children under 10 years.
- *Isolated symphysis and iliosacral joint ruptures.*
- Unstable pelvic ring fractures will heal with bedrest and traction or, alternatively, with external fixation.

### Surgical treatment

- *Apophyseal avulsion fractures:* Possibly appropriate for ambitious athletes in order to shorten the period of rehabilitation, although the



### 3.2.9 · Fractures of the pelvis, hip and thigh

end result is no better. Secondly, if impingement symptoms are present for example.

- **Acetabular fractures:**  
In cases of incongruence/instability.
- **Unstable pelvic ring fractures:**  
Surgical treatment for local concomitant injuries, open fractures, deformity or to facilitate care in poly-traumatized patients [2].

#### Complications

- The vital prognosis depends on the concomitant abdominal and cerebrocranial injuries. The mortality rate is approx. 5% [13].
- Visible *pelvic deformities* are the potential consequences of iliac fractures with apophyseal involvement. Changes in the shape of the pelvic openings after ruptures of the symphysis pubis can represent an obstruction during labor.
- *Leg-length discrepancies* occur as a result of concurrent injury of the proximal femoral growth plate or displacement of one half of the pelvis in unstable fractures (Malgaigne fractures).
- *Hip dysplasia, hip subluxation* after co-involvement of the triradiate cartilage in acetabular fractures in children under 10 years [4, 7].
- The probability of *femoral head necrosis* after acetabular fractures associated with hip dislocation is approx. 10% [9].
- *Coxarthrosis* in incongruence after acetabular fractures and femoral head necroses [7].

#### 3.2.9.2 Proximal femoral fractures

##### Occurrence

Fewer than 0.5% of all fractures in children and adolescents are proximal femoral fractures.

- ❗ **Fracture of the proximal femur during the growth phase is roughly 100 times less common than during adulthood and has correspondingly fewer social and economic implications. Despite their rarity, however, these fractures are very important because they can be associated with serious complications, including femoral head necroses, coxa vara or valga and pseudarthroses.**

Proximal femoral fractures are usually the result of major traumas, particularly traffic accidents. If there is a history of relatively minor traumas, other bone disorders such as a unicameral bone cyst or fibrous dysplasia should be considered.

##### Diagnosis

##### Clinical features

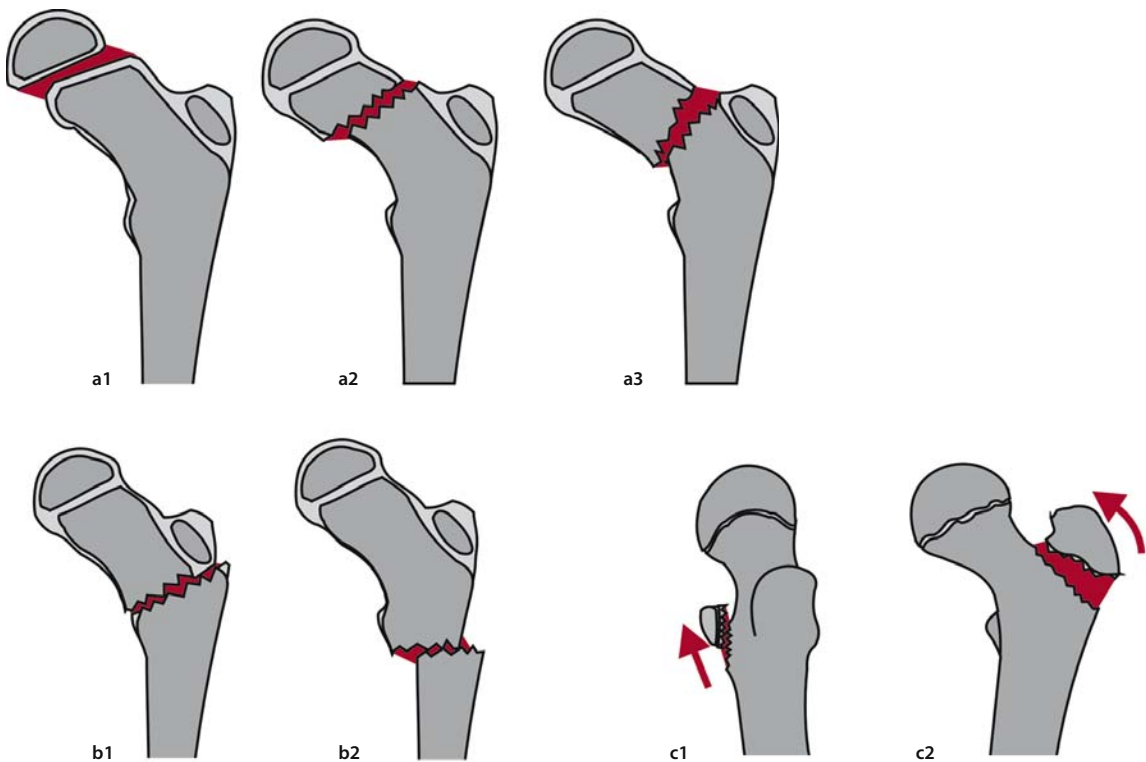
Inguinal pain, also leg shortening and external rotation with dislocated fractures.

#### Imaging investigations

If the AP x-ray of the pelvis does not show any clear fracture or dislocation, an axial view is arranged. Bone scan: if a stress fracture or pathological fracture is suspected, particularly in fibrous dysplasia.

#### Fracture types

- While the classification of *femoral neck fractures* (■ Fig. 3.231a1–a3) does not have any therapeutic consequences, it does have prognostic significance when considered in combination with the patient's age and the degree of dislocation.
- *Epiphyseal separations (type I)* represent the most medial, and also the rarest, femoral neck fractures. The younger the child, the more violent the trauma must be, e.g. in connection with child abuse. They are very rarely connected with birth trauma (differential diagnosis: septic arthritis, congenital hip dislocation). During adolescence the growth plates are loosened under the influence of the sex hormones. Accordingly, the separations are generally not induced by trauma in this age group but are usually associated with overweight or a high level of sporting activity.
- *Medial femoral neck fractures (transcervical, type II)* are usually dislocated, rarely impacted and, at approx. 50%, account for the largest percentage of proximal femoral fractures.
- *Lateral femoral neck fractures (basocervical, cervicotrochanteric, type III)* occur medially to, or on, the upper intertrochanteric line. They are more likely to end in a femoral head necrosis or coxa vara in children than in adults.
- *Per- or intertrochanteric fractures (type IV; ■ Fig. 3.231 b1–b2)* are associated with a much lower risk of avascular necrosis compared to types I–III, although they do involve a risk of a growth disorder if the trochanteric physis is also involved; a high proportion of pathological fractures occur particularly with juvenile bone cysts.
- *Subtrochanteric fractures* are rare and usually the result of direct traumata, e.g. in winter sports or when pedestrians/cyclists/motor-cyclists are hit by a car.
- Isolated *greater trochanter fractures* (■ Fig. 3.231c1–c2) are rare. Dislocated fractures involve a risk of impaired circulation in the femoral head.
- *Lesser trochanter fractures* correspond to apophyseal avulsions of the iliopsoas tendon, usually in peripubertal athletes. The avulsions rarely involve a dislocation of more than a few centimeters and heal without complications, often accompanied by substantial callus formation.
- The femoral neck is a rare site for a *stress fracture*. Isolated cases have been described in children and adolescents undertaking high levels of sporting activity [15].



■ Fig. 3.231. Fracture types at the proximal end of the femur: The classical femoral neck fractures – epiphyseal separation (type I), transcervical (type II) and basocervical femoral neck fracture (type III) – should

be differentiated from the rare peri- and subtrochanteric femoral fractures (type IV) and avulsion of the two trochanters

**Treatment**

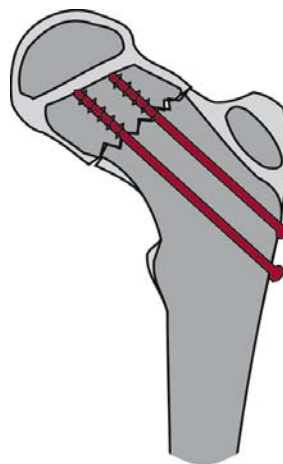
**Conservative treatment**

Aspiration of the hemarthrosis in femoral neck fractures and immobilization in a hip spica extending to just above the knee on the non-fractured side:

- Non-dislocated femoral neck fractures, greater trochanter avulsions, peri- and subtrochanteric fractures (the latter without previous aspiration). Positional check in two planes after 7–10 days. Weight-bearing crutches for 4 weeks.
- Avulsion fractures of the lesser trochanter.

**Surgical treatment**

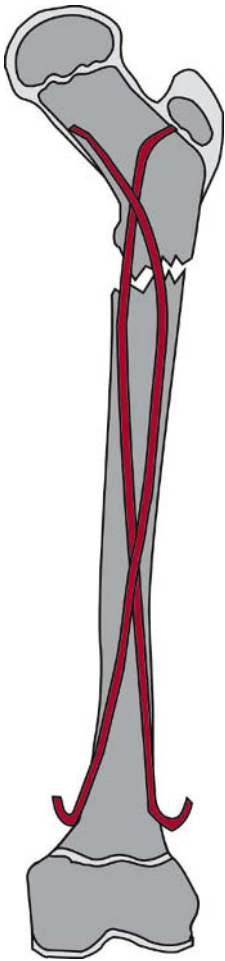
- Epiphyseal detachments: Wiring or nails through the growth plate or a screw.
- Dislocated femoral neck fractures: Open reduction via a standard anterolateral approach, capsule windowing and fixation, but not through the growth plate, with Kirschner wires, Steinmann pins or lag screws (■ Fig. 3.232).
- Dislocated inter- and peritrochanteric fractures: Open reduction and fixation with an angled blade plate.
- Subtrochanteric fractures: For transverse or short oblique fractures: ascending intramedullary flexible nailing from the lateral side



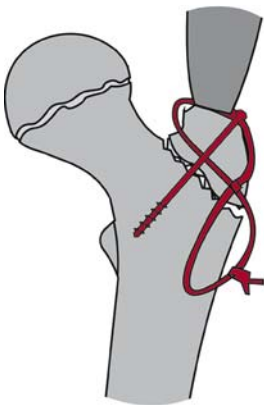
■ Fig. 3.232. Treatment of femoral neck fracture: The classical dislocated transcervical femoral neck fracture is openly reduced and fixed with two cancellous bone screws. If epiphyseal separation is present, Kirschner wires are used and inserted through the epiphyseal plate to secure the top of the head

(■ Fig. 3.233). For spiral and multifragmented fractures: angled blade plate osteosynthesis.

- Greater trochanter avulsions: Tension-band wiring (■ Fig. 3.234).



■ Fig. 3.233. Treatment of subtrochanteric femoral fractures: Non-dislocated fractures are treated conservatively. Dislocated fractures can be stabilized effectively in children with intramedullary nails, although this stabilization may not be sufficient for older children and adolescents. In such cases the surgeon must switch, during the same operation, to internal fixation with angulated blade plate



■ Fig. 3.234. Treatment of the dislocated greater trochanter: Non-dislocated fractures of the greater trochanter are treated conservatively. Dislocated fractures are openly reduced and secured with tension-band wiring to produce a functionally stable result

### Immobilization period

4–6 weeks in a hip spica for non-dislocated fractures or surgically managed fractures in under 6-year olds. Mobilization on crutches after internal fixation in over 6-year olds with partial weight-bearing for 4–6 weeks.

### Follow-up management and complications

- Until completion of consolidation and full mobility: Lesser trochanter avulsion fractures.
- Every 6 months until at least 2 years after the trauma: All other fractures, since they all involve the possibility of an aseptic head necrosis and growth disorders.

### Complications

- ❗ Whereas deformities resulting from a growth disorders can be rectified by corrective procedures, a femoral head necrosis can only be influenced to a limited extent by treatment in terms of its severity, course over time and consequences.

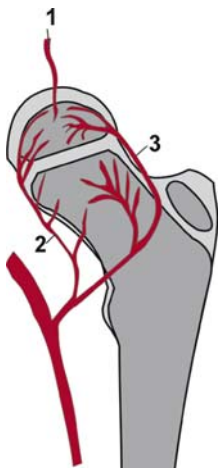
The risk of an *avascular femoral head necrosis* can be as high as 40%, i.e. much higher than in adults. Type I fractures are more at risk of necrosis than types II and III, principally because of the age-dependent vascular supply. Around the age of 4 the anatomy of the arterial supply changes, thereby also affecting the risk of a post-traumatic circulatory problem of the growing femoral head [11, 16].

Three vascular systems are relevant in this context (■ Fig. 3.235):

1. The direct epiphyseal supply from the pole of the head via the foveolar artery in the ligament of the head of femur contributes very little to the arterial circulation up to the age of 8, but accounts for approx. 20% of the arterial circulation in adulthood.
2. Up until the age of 4, metaphyseal side branches of the medial and lateral circumflex femoral arteries that cross the growth plate extend to the femoral head. Thereafter, the plate acts as a vascular barrier and the head is supplied only from the epiphyseal side.
3. The primary supply is then ensured almost exclusively via the epiphyseal posterosuperior and posteroinferior branches of the medial circumflex femoral artery, which then re-anastomose with the metaphyseal network only after physeal closure.

The importance of immediate evacuation of the hematoma is disputed. The extent of the dislocation at the time of the trauma, together with the age of the patient, is far more important than the type of fracture, the time and type of treatment and the duration of protected weight-bearing [10].

The consequences of head necrosis depend on its extent, location and, in particular, on any growth plate



**Fig. 3.235. Vascular supply:** The femoral head is supplied by three vascular systems during growth, the artery of the ligament of the head of femur (1), the medial circumflex femoral artery (2) and the lateral circumflex femoral artery (3). The two latter arteries may show age-related anastomoses, hence the much better prognosis for femoral head necrosis up to the age of approx. 4–5 years compared to later childhood and adolescence

involvement. Symptoms and radiological changes generally occur in children within a few weeks or months after the trauma, i.e. earlier than in adults. Pain and restricted movement, radiological joint space widening and head sclerosis are the initial signs and will require further clarification by MRI. From the therapeutic standpoint, neither a prolonged period of protected weight-bearing nor core decompressions have any appreciable influence on the often disastrous course of the condition. The value of circulation-promoting drugs, e.g. iloprost (Ilomedin), a prostaglandin, has not yet been clearly established. In addition to assisted mobilization by physical therapy in order to preserve mobility, the possibility of realignment procedures such as pelvic and/or femoral osteotomies should be considered if there are problems of femoral head centering and acetabular coverage.

### Growth disorders and posttraumatic deformities

Given the extensive anatomical spread of the femoral head and trochanteric growth plates, they are likely to be directly involved in the event of trauma or indirectly involved as the result of a circulatory problem. A continuum initially exists from the medial femoral head via the lateral femoral neck to the lateral greater trochanter, but this rapidly breaks up between the ages of 8 and 10. Thereafter, the head (femoral neck growth) and trochanteric physes (trochanteric growth) are anatomically and functionally autonomous. The following changes can occur depending on the time and extent of the damage:

- Trochanteric elevation, trochanteric hypoplasia.
- Coxa valga as a result of lateral physeal closure.

- A *coxa vara* may be the result of instability or an inhibiting growth disorder. Whether the determining factor in the latter case is the trauma itself, the subsequent circulatory impairment or iatrogenic trauma is unclear. Except in very small children, a varus deformity of 110° or less persists and will therefore require correction.
- Femoral neck shortening.
- Leg-length discrepancies: The proximal femoral epiphyseal plate contributes approx. 30% to femoral growth, which means that physeal closure results in a length loss of approx. 4–5 mm per year.
- Pseudarthroses are usually the result of inadequate or absent fixation of the fracture and represent an indication for a revision procedure.
- Infections with femoral neck osteomyelitis and/or septic arthritis occur in 10–20% of patients. The main risk factors are aseptic necrosis and open reduction [10].

### 3.2.9.3 Diaphyseal femoral fractures

#### Occurrence

Around 1% of all pediatric fractures are diaphyseal femoral fractures. Causes to be ruled out in children below walking age are child abuse and osteogenesis imperfecta. Older children usually suffer these fractures in accidents as pedestrians or cyclists.

#### Diagnosis

#### Clinical features

Pronounced swelling of the thigh as a result of shortening and the fracture hematoma is the most striking finding in complete fractures. Concomitant neurovascular damage and soft tissue injuries are frequent after direct traumata. In particular, the possibility of rupturing of the underlying muscles should be investigated if skin lacerations are present.

**!** A drop in blood pressure is rarely explained by a single femoral fracture, but usually occurs as a result of some additional injury, whether pelvic, intra-abdominal or intrathoracic.

#### Imaging investigations

If a clinically visible deformity is present, a single projection plane will suffice, otherwise standard AP and lateral views to include the hip and knee are indicated.

#### Fracture types

- Greenstick fractures: primarily in the area of the distal femoral diaphysis with compression of the medial cortical bone.
- Compression fractures.
- Transverse fractures and short oblique fractures are the most common type.
- Spiral fractures.

### 3.2.9 · Fractures of the pelvis, hip and thigh

- *Pathological fractures* principally affect the metadiaphyseal junction in unicameral bone cysts and *fibrous dysplasia*.
- *Stress fractures* rarely occur in the femur.

#### Treatment

The treatment of choice depends mainly on the fracture type and the age and weight of the patient. Multiple injuries, local soft tissue contusion, psychosocial and economic factors are also taken into account. The various options should be presented in detail to the parents and – adapted to the child's age – the patient, so that they can be actively involved in the therapeutic decision.

#### Femoral shaft fractures: Treatment of choice according to age group

- **0- to 5-year olds:**
  - Shortening up to a maximum of 2 cm: Immediate hip spica, unless contraindicated as a result of substantial swelling or concomitant injuries.
  - Shortening >2 cm (rare): 5 days of adhesive tape traction followed by a hip spica or, alternatively, intramedullary nailing in consultation with the parents.
  - Polytrauma, open fracture or in combination with severe craniocerebral trauma: External fixator.
- **Over 5-year olds:**
  - Stable, not dislocated: Pelvic ring cast.
  - Unstable: Prévot nailing or external fixator (Fig. 3.236).
  - After physeal closure: Solid medullary nailing.

#### Spontaneous corrections

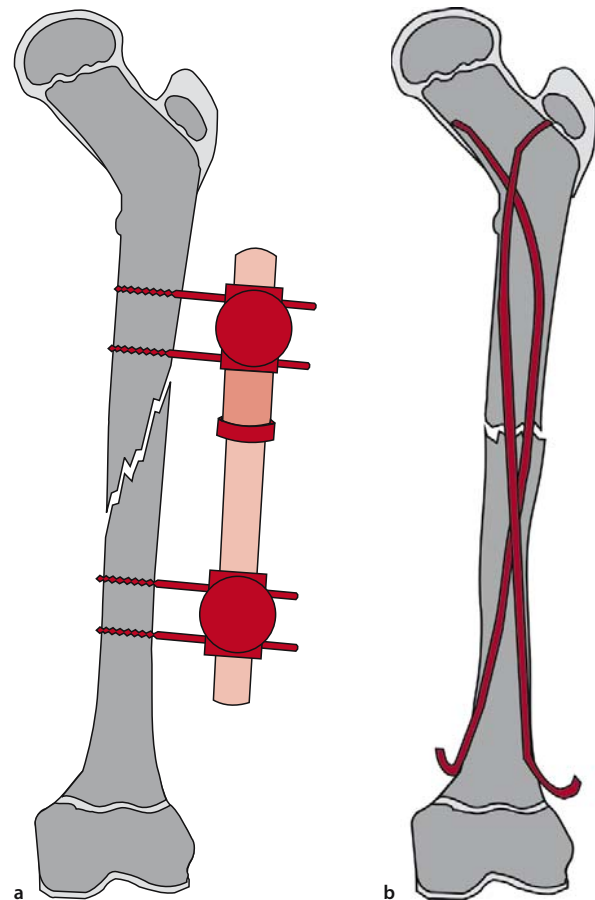
Axial deviations and shortening should be avoided in view of posttraumatic leg length alterations as a result of stimulatory growth disturbances. As regards possible spontaneous correction, the following critical values may be assumed in relation to the extent and level of the deviation and the patient's age:

- Side-to-side deviations: toddlers – full, subsequently up to half, shaft width.
- Shortening: toddlers – 1–2 cm, subsequently no shortening.
- Valgus and antecurvation deformities: up to approx. 10°.
- Varus and rotation deformities: in toddlers up to approx. 20°, subsequently up to 10°.
- Retrocurvation hardly ever occurs.

#### Conservative treatment

##### Hip spica

For closed unstable femoral fractures in under 5-year olds we immediately apply a hip spica, under sedation,



■ **Fig. 3.236a, b.** Treatment of femoral shaft fracture between the age of 5 and epiphyseal closure. Since it is better from the weight-bearing standpoint we prefer stabilization with the external fixator (Monotube yellow, blue or red) for spiral, comminuted and long oblique fractures. Sedation usually proves sufficient for implant removal, and a general anesthetic is only required in around a third of cases. We consider intramedullary flexible nailing to be indicated for proximal fractures or transverse and short oblique fractures in the middle of the shaft. Since a general anesthetic is always required for implant removal in such cases, the overall process is rather more complicated than the external fixator method

in 90° hip and knee flexion and 30° hip abduction, i.e. a »sitting spica«. This is the best position for the purposes of hygiene, sitting and carrying. The spica extends on the non-fractured leg to just above the knee (Bermuda cast) and, on the fractured side, to just above the ankle. In children of all ages with stable fractures, a pelvic ring cast will allow them to mobilize independently on crutches. The cast basically involves a circular thigh cast supplemented by a pelvic ring.

#### Surgical treatment

The traction table is used only if the correct femoral length cannot be restored by moderate manual longitudinal traction on a relaxed patient.



### Retrograde flexible intramedullary nailing (Prévoit nailing)

- Transverse fractures,
- Short oblique fractures,
- Pathological fractures.

Fractures at the distal metadiaphyseal junction can be managed in a descending direction, inserting the nails via an access created at the distal lateral greater trochanter.

### Unilateral external fixation

- spiral fractures,
- multifragmented and comminuted fractures,
- open fractures.

### Solid anterograde medullary nailing

We perform solid anterograde medullary nailing at the earliest shortly before physeal closure in view of the risk of femoral head necrosis, possible growth disturbances at the greater trochanter, the limited width of the medullary canal and the valid alternatives [1, 12].

### Radiographic controls

Positional control 1 week after the trauma for initially non-dislocated fractures according to the age of the child. Consolidation x-ray depending on the age in each case: toddlers 4 weeks, 5- to 10-year olds: 6–8 weeks, >10-year olds: 8–12 weeks and every 4 weeks until consolidation.

### Immobilization and consolidation period

From approx. 2 weeks for non-dislocated fractures in neonates up to approx. 12 weeks in adolescents.

### Implant removal

- External fixator: when at least 3 sections of cortical bone in AP and lateral standard projections show consolidation.
- Flexible intramedullary nails: approx. 6–12 months after implantation.
- Solid medullary nails: 1 year postoperatively.

### Follow-up management

External fixator: Pin care. Dynamization from approx. 3–4 weeks. Physical therapy: initially, for instruction on walking with weight-bearing crutches according to the level of pain; after implant removal, for instruction on walking, muscle strengthening, coordination training and, for patients who take part in sport, gradual rehabilitation until the load-bearing level specific to the sport is reached.

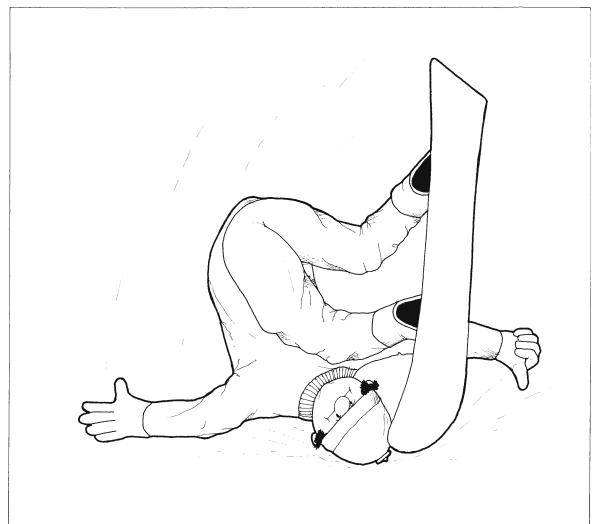
After the removal of a hip spica, the toddler is left to mobilize spontaneously and the parents are advised that it may take a few days before their child develops sufficient strength and confidence to be able to stand and walk.

### Follow-up controls

Up to 2 years after the trauma to ascertain any posttraumatic leg-length discrepancy.

### Complications

- *Avascular femoral head necroses* are a possible threat after anterograde solid medullary nailing while the growth plates are still open. Although it has only been reported in isolated cases, this complication is serious enough to warrant recourse to other options, e.g. flexible retrograde nailing or external fixation [1].
- *Delayed healing and pseudarthroses* are rare. They are a possible threat after serious open fractures with bone loss or extensive soft tissue injuries, osteomyelitis or unstable fixations, but can also occur after the external fixation of transverse or short oblique fractures, particularly during adolescence [18].
- The value of early dynamization in order to accelerate consolidation during external fixation is probably overestimated, as has been demonstrated by one prospective randomized study [5].
- *Refractures* can occur particularly after external fixation: In contrast with intramedullary nailing, the patient can feel irritated by his »frame« and pressurize doctors into premature removal of the implant. The premature resumption of sport predisposes to refracture after removal of the fixator. As a rule, therefore, the child must refrain from taking part in contact or ball-based sports, skiing, snowboarding or jumping for 4 to 6 weeks after the removal of an implant from the femur.
- *Growth disturbances and posttraumatic deformities:* Premature closure of the greater trochanter physis after anterograde nailing can lead to coxa valga and subluxation of the femoral head [12].



### 3.2.9 · Fractures of the pelvis, hip and thigh

- *Posttraumatic leg-length discrepancies* are the result of growth stimulation in under 10-year olds or of posttraumatic shortening in over 10-year olds. The commonest causes of stimulatory growth disturbances are the remodeling of ad latus and shortening deformities, postprimary manipulation at the fixation callus and traction treatment. The stimulation lasts for less than 2 years, even for substantial remodeling. After the age of 10, the growth plate nearest the fracture usually closes prematurely, which also influences the final leg-length discrepancy. Consequently, correct fracture reduction in terms of axis and length within the first few days is the top priority [6]. Failure to observe these rules may result in a length change of up to 3 cm.
- *Axial deformities* should be prevented from the outset. Consolidated deviations remodel themselves subject to the critical values specified above. Parents of children under 10 years of age are advised to wait until completion of this phase of spontaneous correction in order to avoid any unnecessary corrective procedures.
- *Rotational deformities* usually manifest themselves as external rotational deformities of the distal fragment. Deviations up to approx. 20° can be actively compensated for by rotation at the hip and are therefore rarely of clinical significance even if they persist after completion of growth. Rotational deformities are at least partially corrected spontaneously in connection with the physiological derotation that occurs up until the pubertal growth spurt [3]. The quality of the intraoperative fracture reduction and the rotation situation during follow-up are ideally determined by comparing the extent of internal and external rotation with the hip flexed at 90°. This check does not apply in the case of conservative treatments, but is essential at the end of surgical fixation.
- *Restricted mobility:*

  - After Prévot nailing: Usually caused by an irritating nail end at the medial femoral epicondyle beneath the vastus medialis muscle.
  - After external fixation: Can largely be avoided by a) flexing the knee to its maximum extent at operation to facilitate the passage of the pins through the fascia lata, and b) positioning the knee intermittently in 90° hip and knee flexion for several days postoperatively (using a foam block).
- *Infections*  
Pin-track infections can be expected to occur in patients with external fixation in 5%–10% of cases, even with a good standard of care/instruction. Pain, seropurulent secretion and reddening at the pin insertion sites are the initial warning signs. Oral broad-spectrum antibiotics, daily baths or showers and local pin care usually reduce the inflammation promptly. Only in rare cases does the skin incision need to be

opened up or, even more rarely, a loose pin need to be changed.

If the infection fails to respond to local or medical measures, or if general symptoms such as high fever, malaise, tiredness and loss of appetite occur, the possibility of osteomyelitis will need to be ruled out by further diagnostic investigation [erythrocyte sedimentation rate, C-reactive protein, leukocyte count (differential), bone scan, possibly MRI].

- Osteomyelitis is rare after the percutaneous fixation of closed fractures, but common after severe open fractures or in cases of defective or delayed healing [8].

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### 3.2.10 Transient synovitis of the hip

#### ➤ Definition

Transient synovitis is a hip joint effusion that occurs in small children in connection with other illnesses (e.g. viral infections of the upper respiratory or gastrointestinal tract). The effusion is transient and disappears after a few days. Transient synovitis is a symptom rather than a separate illness.

— *Synonym: Toxic synovitis*

#### Etiology

Since transient synovitis occurs as a symptom in association with other, usually viral, infections, there is no uniform etiology [1, 16, 24]. It involves a reaction to a process outside the hip [17], most commonly a viral infection of the upper respiratory or gastrointestinal tract. Such disorders can produce effusions in the hip or just synovial swellings, which trigger the symptoms described below.

The condition is characterized by an accumulation of fluid in the hip and its transient nature. The correct diagnosis must be stated, e.g.: *Transient synovitis* in association with a trivial, general viral infection, transient synovitis in association with Legg-Calvé-Perthes disease, etc. Ultrasound studies have shown that a (slight) effusion is also present, without producing symptoms, in the other hip in around a quarter of cases [3].

There has been much discussion as to whether Legg-Calvé-Perthes disease can develop from transient synovitis. This idea was postulated in the 1980's, but has not been confirmed by subsequent investigations [6, 7, 10, 17]. While Legg-Calvé-Perthes disease may be accompanied by an effusion, the underlying disease itself can already be diagnosed at this stage sonographically (on the basis of cartilage thickening) and radiologically (condensation, cartilage thickening, poss. subchondral fracture). The effusion is never the cause but, at most, a concomitant symptom of the Legg-Calvé-Perthes disease.

In a follow-up study involving 119 children with transient synovitis, Legg-Calvé-Perthes disease did not subsequently occur in a single case [10]. While a femoral head necrosis can be generated experimentally in animals by producing excessive pressure in the hip [11], the pressure situation in transient synovitis is not comparable with these experimental conditions. What is striking, however, is the fact that children with transient synovitis are smaller than average for their age – precisely the same finding has also been observed for Legg-Calvé-Perthes disease [22]. No explanation has yet been found for this phenomenon.

#### Occurrence

Transient synovitis is the commonest hip disorder in growing children. The frequency peaks around the age of 5 or 6, but it can also affect children at any age between 1 and 12/13 [1]. The annual risk of transient synovitis calculated in a major study in Sweden was approx. 0.2%, while the cumulative risk for a child was approx. 3% [17]. The risk of recurrence is high as 90% of the children affected subsequently experienced a second episode of transient hip effusion. Another study in Germany calculated an annual incidence of approx. 0.076% [23]. A recurrence risk of 15% was determined in a British study [20].

#### Clinical features, diagnosis

The joint effusion causes pain, which manifests itself as limping and restricted hip movement. Depending on the extent of the effusion, the pain may be sufficiently severe as to render the child incapable of walking.

The clinical picture of transient synovitis is short-lived in 80% of all cases connected with a trivial viral infection – in accordance with the declining effusion and thus the declining pressure [9, 17] in the hip. This means that the symptoms steadily regress and disappear, at the latest, by the 5th day after the onset of the illness, with or without treatment [1, 16]. The onset is usually acute, producing a spontaneous limp. The children with transient synovitis are always healthy and are not feverish, nor do they have a history of high fever in the previous 14 days. Confusion can be caused by cases that are superimposed by a current viral infection with subfebrile temperatures (e.g. of the upper respiratory tract).

In 20% of cases the symptoms are not short-lived, but persist or recur without treatment or after the discontinuation of anti-inflammatory measures for periods exceeding two weeks [16]. But even these children are invariably in good health with no clinical signs of a serious illness. The patient presenting with transient synovitis shows a distinct limp and significant restriction of hip mobility, initially in terms of flexion/extension, subsequently extending to internal/external rotation, right up to complete locking of the hip. The limp usually occurs spontaneously, although transient episodes of limping during the 10 days preceding the initial consultation are also sometimes reported.

The primary diagnosis is always based on the effusion, which is best detected by an *ultrasound scan* [4,19], although this cannot differentiate between a serous effusion and pus.

❗ **The most important differential diagnosis is septic arthritis of the hip. This diagnosis must not be missed under any circumstances, since an infection in a joint that persists for more than 4 days inevitably results in permanent damage.**

### 3.2.10 · Transient synovitis of the hip

Our diagnostic efforts therefore focus primarily on the exclusion of this diagnosis [14]. An American study has shown that four parameters can be used to diagnose a purulent process in the hip:

- high fever,
- inability to walk,
- elevated erythrocyte sedimentation rate (>40 mm in the 1st hour),
- white blood cell count higher than 12,000 [14].

The diagnostic accuracy was 40% for two predictors, 93% for three predictors and 99% when all four predictors were present. The validation of these statements has shown contradictory results in two recent studies [15, 18]. We consider C-reactive protein (CRP) to be a more suitable parameter than erythrocyte sedimentation since the latter reacts more slowly to an infection.

Fever and the inability to walk can readily be ascertained clinically.

**! The white cell count and the CRP (or the erythrocyte sedimentation rate) must be determined in every child with hip pain and restricted hip movement. A delay in the diagnosis of a single case of septic arthritis of the hip is not justified by the cost savings resulting from the omission of laboratory tests for hundreds of negative cases – particularly since the costs of treating a purulent hip condition rapidly spiral if it is not diagnosed in good time.**

An experienced clinician is usually able to complete the differential diagnosis with a high degree of certainty on the basis of the child's general condition alone. A child with a septic process is seriously ill, whereas the child with transient synovitis can appear quite cheerful. On the other hand – every experienced clinician was once inexperienced...

If an effusion is present in the hip and if the clinical parameters suggest an infectious process, the hip effusion must be *aspirated* and the aspirated fluid forwarded for bacteriological investigation. Under no circumstances may antibiotics be administered before the aspiration.

If the aspirated fluid is clear but there are definite clinical signs of infection, the possibility of *acute hematogenous osteomyelitis* should be considered. Further investigations are indicated in this situation: bone scan, poss. MRI (► Chapter 3.2.11).

If there are no signs of infection but the illness and/or effusion is protracted, we consider the following differential diagnoses [1, 2, 5, 8, 16]:

- juvenile rheumatoid arthritis of the hip,
- leukemia,
- Lyme disease,

- Legg-Calvé-Perthes disease,
- Crohn's disease and ulcerative colitis,
- multiple epiphyseal dysplasia,
- slipped capital femoral epiphysis.

#### Other investigations

- *X-rays (AP and axial)*: clarification in respect of Legg-Calvé-Perthes disease, epiphyseal separation, osteomyelitis, epiphyseal dysplasia.
- *MRI*: Legg-Calvé-Perthes disease, epiphyseal dysplasia, osteomyelitis, effusion, epiphyseal separation, inflammatory soft tissue changes.
- *Bone scan*: osteomyelitis, soft tissue disorders associated with rheumatological processes.
- *Laboratory*: leukemia, infections (blood culture, CRP, erythrocyte sedimentation, differential white blood cell count), juvenile rheumatoid arthritis (rheumatoid factors) etc.

#### Treatment, course

The following therapeutic options are available for transient synovitis:

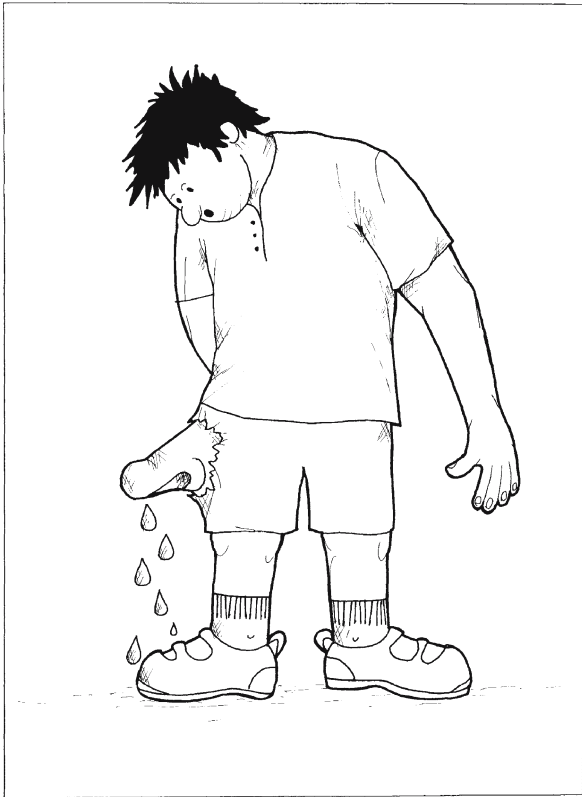
- Resting the hip,
- Aspiration,
- Analgesics/anti-inflammatory drugs.

*Resting the hip* is always indicated. The child automatically rests the affected leg in any case since it is painful. Small children have an excellent instinct in relation to pain. They spontaneously avoid weight-bearing on a painful extremity until the symptoms have disappeared (in contrast with adults, some of whom like to play the hero while others suffer from inertia and do not risk weight-bearing at all even when the pain has subsided).

The benefit of *aspiration* is disputed. Although the removal of the fluid relieves the joint and also the pain, the effusion often recurs after aspiration [10, 21]. However, one randomized study has shown that the course of the illness is significantly shortened by aspiration [13]. The drawback of aspiration is the need for a general anesthetic. We therefore aspirate only in those cases involving a distinct restriction of movement and with sonographic evidence of a substantial effusion.

*Analgesics/anti-inflammatory drugs* are useful. One randomized study comparing the administration of ibuprofen vs. placebo showed that the anti-inflammatory agent shortened the duration of the illness from 4.5 days to 2 days [12].

In our view, the constantly repeated recommendation to *follow-up* a case of transient synovitis after 3–6 months on the grounds that Legg-Calvé-Perthes disease could develop from the effusion is not appropriate. Any incipient Legg-Calvé-Perthes disease found at this point will have already been present beforehand but had simply not been visible or had been overlooked [17] and did not



develop from the effusion. What is important however, if incipient Legg-Calvé-Perthes disease is involved, is that recurrent episodes of limping can be expected and these will naturally require follow-up. The patient and the parents should be informed of this possibility. But a transient effusion of itself does not trigger Legg-Calvé-Perthes disease.

*Recurrences* can occur in up to a third of cases. Usually the parents do not even bother bringing the patients for a consultation since they treat the condition themselves at home. If they do attend, however, the procedure is the same as that for the initial episode.

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### 3.2.11 Infections of the hip and the femur

#### 3.2.11.1 Septic arthritis of the hip

##### Definition

Hematogenous bacterial infection of the hip, usually in infants or toddlers, with or without involvement of the proximal femoral metaphysis.

— **Synonyms:** Septic coxitis, infectious arthritis of the hip

##### Occurrence

Septic arthritis of the hip is the commonest septic joint condition during growth, reaching a distinct peak in frequency during infancy.

##### Etiology

The joint is infected during childhood via hematogenous transmission, resulting in colonization of the joint with bacteria. See ► chapter 4.3.2 for more detailed information on the etiology of hematogenous joint infections.

##### Growth prognosis and complications

If the effusion – and thus the intra-articular pressure – persists for several days or weeks, this can lead to subluxation or dislocation of the hip for purely mechanical reasons [8] (■ Fig. 3.237). Furthermore, any purulent effusion in a joint that persists for more than 4 days will naturally result in irreversible damage to the joint cartilage and the growth plates. The shorter the duration of the effusion, the less serious the damage. The longer the joint infection lasts, the greater the direct damage to the femoral head and neck, potentially resulting in complete destruction and disintegration of the proximal end of the femur [3, 10] (■ Fig. 3.238).

While the damage to the joint cartilage only manifests itself as early osteoarthritis at a late stage, the damage to the growth cartilage is apparent sooner: The premature partial or total closure of the proximal growth plate leads to deficient growth and shortening of the proximal end of the femur, with or without additional increasing axial deviation of the femoral neck [12]. Since the trochanteric apophysis is not usually involved, the trochanter can grow excessively long (■ Fig. 3.239).



■ Fig. 3.237a–d. Acute septic arthritis of the hip in an infant with defective healing. **a** Septic arthritis of the hip with a history of more than 5 days was diagnosed at the age of 2 months. The radiograph shows that the left femoral head has already subluxated. **b** At the age of 15 months an attempt was made to center the hip by means of a Salter pelvic osteotomy and an intertrochanteric varus/derotation osteotomy. **c** At 6 years the hip is still clearly off center. **d** By 10 years, a neoacetabulum has formed above the original acetabulum as an adaptation to the subluxated femoral head



**Fig. 3.238.** Defect after septic arthritis of the hip in a toddler. 5-year old girl with septic arthritis of the hip. The femoral head has been completely destroyed by the infection



**Fig. 3.239.** Trochanteric overgrowth after a growth disorder due to septic arthritis of the hip. 14-year old boy after an episode of septic arthritis of the hip 5 years previously, with femoral head necrosis and a growth disorder. Overgrowth of the greater trochanter persists as a residual defect

Femoral head necrosis is less a result of pressure-induced vascular damage than the consequence of direct destruction. Acetabular involvement can occur in extended and protracted cases with the possibility of damage to the triradiate cartilage [16]. Secondary dysplasias also occur. The joint destruction can be serious enough to warrant a total hip replacement soon after reaching adulthood [6].

If septic arthritis of the hip is promptly detected and treated effectively, the prognosis can certainly be considered good. Apart from an occasionally observed case of coxa magna and slight (in the context of idiopathic differences) leg-length discrepancies, no serious late-occurring sequelae would be expected [7]. While there are isolated reports of bony recurrences after septic coxitis, these appear rather to be cases of unhealed concomitant osteomyelitis which have become chronic and which can flare up even after several years. It is not certain whether the isolated descriptions of late-occurring growth disorders are likewise the consequences of chronic concomitant bone infections or whether these can also occur after long-healed arthritis.

### Clinical features

The clinical findings in septic arthritis of the hip correspond with those of bacterial inflammations in other major joints and are discussed in detail in ► chapter 4.3. Features particular to the hip will only be mentioned at this point. The diagnosis can often be difficult in infants since septic temperatures are not always present [17]. Indicative signs include the poor general condition and septic appearance of these small patients.

The signs and symptoms in the hip, which are often first discovered as incipient pain in the hip when the baby's diaper is being changed, increase over time and are not transient. A nearby pelvic osteomyelitis can simulate the signs and symptoms of septic arthritis of the hip, as can psoas abscesses [11], or can also lead to a sympathetic sterile effusion in the hip. However, this does not affect the subsequent management. Not infrequently – particularly in infancy – the condition affects more than one joint, and this should be clarified by careful general clinical examination of the patient.

### Diagnosis and treatment

**! Any hip pain in an infant (even without fever) and in patients older than 1 year accompanied by fever must be considered to be septic coxitis until proven otherwise!**

The diagnostic strategy for septic arthritis is discussed in detail in ► chapter 4.3. When it affects the hip, the restriction of movement is very easy to check at any age. Internal rotation, in particular, is reduced in a side-to-side comparison in the presence of a joint effusion. An ultrasound

scan is particularly suitable for diagnosing septic arthritis in the hip [9] (■ Fig. 3.240).

Treatment is likewise based on the principles outlined in ► chapter 4.3. Disagreement exists about the local treatment. The argument in favor of immobilization (► Chapter 4.3) can be dismissed straightaway. Basi-

cally, any joint, particularly the infected joint, should be moved and not immobilized. This statement is supported by numerous experimental and clinical reports of children and adults. On the other hand, there is no objection to immobilization in a splint for a few days for the purposes of pain relief. There is much more vigorous debate about the type of evacuation of the purulent joint effusion.

The majority of authors support an urgent arthrotomy with the insertion of an irrigation drain [2, 17], although repeated arthroscopic irrigation is gradually gaining favor [4, 18]. Some authors just aspirate on a single occasion [5, 18]. Only a single track, rather than the whole joint, can be irrigated with an irrigation drain. As a result pockets of infection can cause extensive cartilage damage despite the treatment. Those authors who perform arthrotomies also stress that the viscous pus – in cases of a prolonged history – and connective tissue septa cannot be removed by arthroscopic irrigation alone, but only by a generous arthrotomy. All authors essentially agree, however, that the prognosis is better, and that the amount of treatment can be reduced, the earlier the condition is diagnosed and treatment started [1, 18], so that arthroscopic irrigation is sufficient and arthrotomy is not usually required [18]. A simple needle aspiration is not a sufficient treatment of septic arthritis of the hip.

#### Our therapeutic strategy

If an *acute stage* with a short history and no radiologically visible complications is present, the joint – even in infants – is *arthroscopically irrigated* with 4–5 l of irrigation fluid. If an arthroscope for infants is not available the irrigation should be performed via 2 wide cannulas. Antibiotic treatment is only initiated after fluid has been aspirated and forwarded for bacteriological testing. The arthroscopic irrigation is repeated at intervals of two days if necessary, i.e. if the effusion recurs, until the irrigation fluid is clear and no more bacteria are detected.

The antibiotic treatment is switched to targeted monotherapy as soon as the culture and sensitivity test results are available. The antibiotic is administered intravenously until the clinical inflammation parameters and C-reactive protein (CRP) have returned to normal. CRP therefore has to be checked on the 2nd, 5th and 8th days after the start of the antibiotic treatment and then at 8-day intervals until it has completely returned to normal. The minimal duration of i.v. treatment is two weeks.

Immobilization should be avoided as a rule. If the patient is large enough for a dynamic splint, he is placed on the splint immediately after the operation and moved continuously. Small children move spontaneously without passive appliances. The patients are given appropriate analgesic medication for any initial pain.



■ Fig. 3.240a, b. Acute septic arthritis of the hip in a 9-year old boy. a Incipient thickening of the joint capsule is visible on the axial and AP x-rays. b The ultrasound scan clearly shows the joint effusion on the left side (top) (for comparison purposes the other side is shown at bottom)

If the condition has reached a *chronic stage* with a long history, radiologically visible destruction or even dislocation of the head (■ Fig. 3.237), we consider that *arthrotomy*, revision, lavage and open reduction of the joint are indicated. We do not insert an irrigation drain, preferring to drain the tube using two tubes, which can subsequently be used if necessary to irrigate the joint periodically.

For a *defective situation* involving widespread destruction of the femoral head and femoral neck and an elevated greater trochanter, Paley has perfected a femoral osteotomy that was originally developed by Schanz and subsequently modified by Ilizarov [13]. This buttresses the femur against the ischial bone and effectively corrects the Trendelenburg limp and the leg shortening (■ Fig. 3.241).

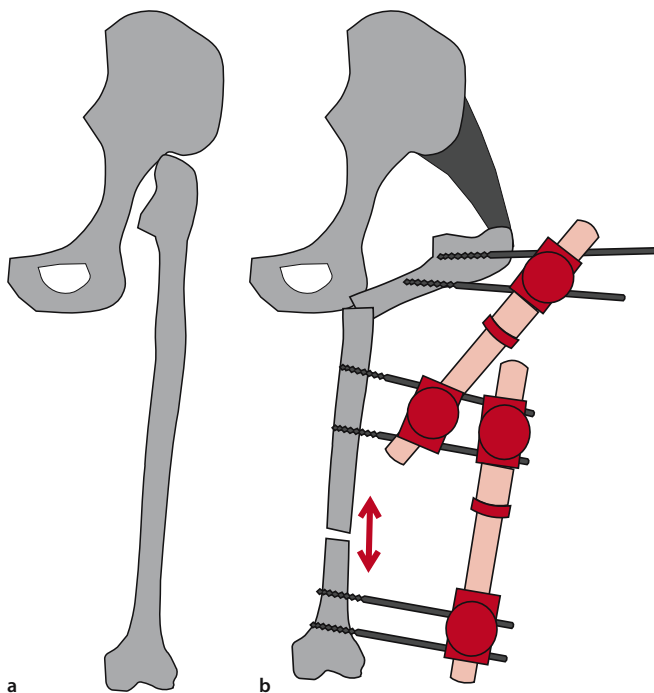
### 3.2.11.2 Osteomyelitis of the thigh

In infants, an osteomyelitis of the femoral shaft can spread extremely rapidly (■ Fig. 3.242). Nevertheless, the prognosis is very good, despite the impressive radiographic findings, if it is treated adequately at a sufficiently early stage. In older children, the femoral shaft tends to be affected by a hematogenous osteomyelitis only in the chronic stage. The osteomyelitis always starts in the metaphyses.

Osteomyelitis, excluding posttraumatic and postoperative forms, is discussed in ► chapter 4.3. Since the distal femoral epiphyseal plate is the most active growth zone in the body and since turnover at the proximal femur is also relatively high, femoral osteomyelitis is a fairly common condition.



■ Fig. 3.242. Osteomyelitis of the femur in an infant. 1 month old boy with osteomyelitis of the whole femur and a pronounced periosteal reaction, as typically occurs at this age



■ Fig. 3.241a, b. Principle of the angulation osteotomy according to Paley. a In preparation, an x-ray of the hip in maximum adduction is recorded. This determines the angle and level of the osteotomy. b This level is selected such that the femur is buttressed against the ischial



bone and the greater trochanter is placed in the maximum lateral position. As a rule, the valgization angle is 40–50° and maximum extension is sought in order to achieve passive standing. A lengthening osteotomy is performed more distally in the same session. c Clinical example



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### 3.2.12 Rheumatoid arthritis of the hip

#### ► Definition

Hip disorder associated with juvenile rheumatoid oligo- or polyarthritis.

#### Occurrence

The incidence of juvenile rheumatoid monoarthritis and polyarthritis (► Chapter 4.4) calculated in a recent study in South Germany was 14.8 per 100,000 children and adolescents under 16 years [10]. The hip was affected in approximately 9% of cases [9].

### Clinical features

The involvement of the hip in juvenile rheumatoid polyarthritis is the principal reason for the loss of the patient's ability to walk. The diagnosis at hip level is made at a relatively late stage since an effusion is not outwardly visible and is difficult to detect. The key symptoms are pain and restriction of movement. An ultrasound scan will often help in detecting the effusion at an early stage [2].

In a study involving 386 children and adolescents with juvenile rheumatoid arthritis affecting the hip, 50% were mono- or oligoarticular, 30% were polyarticular and 20% were systemic [6]. The prognosis in the monoarticular group was good, particularly if the condition started before the age of 6. With systemic involvement, however, this age group had the worst prognosis. Another study described 13 regressive, 2 recurrent and 14 progressive forms [4].

Rheumatoid factors provide an indication of the prognosis: a positive test result is an inauspicious sign. The following structural deformities can be expected in the progressive form of the condition: coxa magna, shortening of the femoral neck, subluxation and cystic erosions of the femur. These result in narrowing of the joint space and – as a particularly typical change – in acetabular protrusion (► Fig. 3.243). An important complication is femoral head necrosis – usually occurring as a result of treatment (steroids), but can also occur spontaneously [8]. Only a



► Fig. 3.243. X-ray of the left hip of a 14-year old female patient with severe juvenile rheumatoid arthritis of the hip with typical acetabular protrusion



small proportion of cases of juvenile rheumatoid arthritis of the hip are progressive.

### Differential diagnosis

The following conditions should be considered in the differential diagnosis: transient synovitis (▶ Chapter 3.2.10), Legg-Calvé-Perthes disease (▶ Chapter 3.2.5), multiple epiphyseal dysplasia (▶ Chapter 3.2.7.1), Waldenström's disease (idiopathic chondrolysis), spondyloepiphyseal dysplasia tarda (▶ Chapter 3.2.7.2), femoral head necrosis in the adolescent (▶ Chapter 3.2.5).

In the initial stages, juvenile rheumatoid coxitis can be difficult to differentiate from transient synovitis. If the symptoms persist for more than 1 week, the possibility of a rheumatoid process should always be considered. If rheumatoid serology tests prove negative, the diagnosis can often only be made after a trial treatment. A good symptomatic response to acetylsalicylic acid suggests the presence of a rheumatoid disorder.

### Treatment

#### Conservative treatment

The conservative treatment of juvenile rheumatoid arthritis of the hip is no different from that for other joints, and the drug therapy is outlined in ▶ chapter 4.4. The local injection of steroids can lead to fairly prolonged remissions. The risk of femoral head necrosis seems to be more associated with systemic treatment and is not increased by local treatment [8].

#### Surgical treatment

##### Soft tissue interventions

##### Joint lavage, mobilization under anesthesia and botulinum toxin injection

If the patient only shows an incipient joint contracture without significant osteoarthritis, joint lavage can prove beneficial. For this procedure we use the arthroscope on the extension table, which is introduced under image-intensifier control. The joint is filled with fluid, causing the capsule to stretch. This procedure has proved effective particularly for adduction contractures, and is often combined with botulinum toxin injection into the adductors (▶ Chapter 3.2.5).

Postoperatively the child is placed in a body cast in a position of maximum abduction. There follows a period of intensive physical therapy, which is made possible by an epidural catheter that is left in situ for several days (also ▶ chapter 3.2.5, ■ Fig. 3.189). This method, often involving relatively little effort and minimal morbidity, can improve mobility, and particularly walking ability, for several months and sometimes years. Part of the effect is achieved by the flushing out of the cartilage breakdown products that are partly responsible for chronic synovitis. A long-term effect cannot be expected, however, in the progressive form.

### Muscle and tendon lengthening

Muscle and tendon lengthening procedures can be useful for patients with contractures. The adductors (particularly the adductor longus muscle) are often contracted, while the tensor fasciae latae, rectus femoris and psoas muscles can be affected in flexion contractures. As a rule, we never perform tenotomies, since the loss of effect of a complete muscle is uncontrollable and can have negative consequences. We therefore prefer aponeurotic lengthening, in which the aponeurosis at the transition between the muscle and tendon is divided by several incisions and the underlying muscle is stretched. Good results can be achieved in the long term with this method [12], although a subsequent deterioration has been observed in isolated cases after 3 years [7].

### Synovectomy

The once commonly practiced procedure of synovectomy has produced disappointing results in juvenile rheumatoid arthritis of the hip. Although it can relieve the pain in the short term, hip mobility, and thus everyday function, is not improved. Radiographs have been shown a faster progression of the osteoarthritis [5]. Therefore, in contrast with the lengthening of muscles and tendons, synovectomy is no longer a recommended procedure.

### Hip implants

If the progressive arthritis leads to a loss of the ability to walk, only a total hip replacement will be able to restore mobility. Hip implants are even inserted in adolescents who are still growing in some centers. We do not have any experience with operations at this age and insert a hip implant at the earliest when growth is complete (■ Fig. 3.244). Substantial experience has been accumulated worldwide with these operations [1, 3, 11].

Compared to total hip replacements for idiopathic osteoarthritis of the hip, the complication rate in this group of young patients is relatively high. Particularly common complications are infections as a result of the long-term treatment with steroids and cytotoxic drugs. We have had the misfortune to experience the death of a female patient from acute sepsis after the insertion of bilateral hip implants. The durability of prosthetic anchorage is relatively high compared to other adolescent groups. Although radiological loosening occurs after the usual period, particularly of the acetabular component as a result of osteoporosis, the prosthesis needs to be changed, on average, only after the same period applicable to older patients with idiopathic osteoarthritis of the hip. This is explained by the relatively low mobility and lower than average weight of rheumatoid patients. The Kaplan-Meier survival curves for total hip replacement patients with juvenile rheumatoid arthritis are similar to those for old patients with idiopathic osteoarthritis of the hip.



■ Fig. 3.244. 16-year old female patient with coxarthrosis in juvenile rheumatoid arthritis of the hip: left AP x-ray of the left hip, right Situation after a total hip replacement

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## 3.2.13 Tumors of the pelvis, proximal femur and femoral shaft

### ► Definition

Primary bone tumors originating in the pelvis and proximal femur, and soft tissue tumors arising from the muscles, connective tissues, blood vessels or nerve tissues in the immediate vicinity of the pelvis, hip and proximal femur.

### Occurrence

#### Bone tumors

Around 5% of all bone tumors in children and adolescents are located in the *pelvic area* (adults: 10%; ■ Table 3.40). Bone tumors are also often located in the proximal femur. After the distal femur and proximal tibia, this region is the third most commonly affected site (► Chapter 4.5.1).

The frequency in a particular part of the body correlates with the growth activity of the epiphyseal plates. Only 15% of tumors in the proximal femur are malignant, compared to more than a third in the pelvis. A malignant tumor that particularly affects adolescents is Ewing's sarcoma, whereas chondrosarcomas are the predominant malignancies in adults [5]. The pelvis is one of the commonest sites affected by Ewing's sarcoma: Out of 200 Ewing's sarcomas, 42 originated in the pelvis [6].

Pelvic osteosarcomas are extremely rare in children and adolescents, and slightly more common in adults. Some of these tumors develop in connection with an underlying Paget's disease [19]. The principal benign pelvic tumor in children and adolescents is an aneurysmal bone cyst [1]. Other very common benign tumors are osteochondromas (cartilaginous exostoses) and Langerhans cell histiocytosis. Fibrous tumors, osteblastomas and osteoid osteomas in the pelvis are rarer in adolescents than in adults [2] (■ Table 3.40).

In the *proximal femur*, osteochondroma, fibrous dysplasia, osteblastoma and juvenile bone cysts are the commonest benign tumors or tumor-like lesions. The proximal femur is the second most frequent site, after the proximal humerus, for juvenile bone cysts. Because of the prevailing shear forces, fibrous dysplasia in the proximal femur leads to a typical curvature that has been compared to the shape of a »shepherd's crook« (■ Fig. 3.245).

The hip itself can be affected by synovial chondromatosis [8] and pigmented villonodular synovitis [16]. In the epiphysis (i.e. in the femoral head) the possibility of a chondroblastoma should be considered (■ Fig. 3.246). Occasionally such a tumor in the femoral head can be confused with a case of Legg-Calvé-Perthes disease. Differentiating between the two should not pose any difficulties however, as in Legg-Calvé-Perthes disease the head is always flattened, which is not always the case with a tumor. Malignant tumors of the proximal femur are fairly rare – usually involving osteosarcomas, and occasion-

ally Ewing's sarcomas, chondrosarcomas or a malignant hemangiopericytoma (■ Table 3.40).

The *femoral shaft* is primarily affected by osteoid osteomas, enchondromas and osteochondromas that grow from the metaphysis into the diaphysis (■ Table 3.40). Of the malignant tumors in adolescents, we have observed several osteosarcomas, but surprisingly few Ewing's sarcomas exclusively in the femoral shaft, even though this tumor forms in the medullary space. It does have a tendency, however, to start its growth at the margins of the medullary space, in which case it becomes a metaphyseal/diaphyseal tumor.

### Soft tissue tumors

A *desmoid tumor* is a benign but very active soft tissue tumor that is frequently located in the buttocks area. This tumor is not all that rare in children and adolescents and its treatment usually poses major problems.

A *rhabdomyosarcoma* is the commonest malignant soft tissue tumor that affects this age group (■ Fig. 3.247).

Less frequent are liposarcomas, while malignant fibrous histiocytomas and fibrosarcomas occur in later life.

### Diagnosis

Tumors in the vicinity of the pelvis and proximal femur are surrounded by large soft tissue masses. Since these only become palpable when they reach a very respectable size they are extremely difficult to diagnose and often only detected at a very late stage.

❗ **If unclear, non-load-related pain occurs in the area of the pelvis or thigh an x-ray should always be recorded in doubtful cases, if necessary followed by a bone scan. Tumors in this region are often overlooked for inexcusably long periods because of the large soft tissue masses.**

The primary imaging technique is always the *plain x-ray*. If the radiographic findings are unclear, a *bone scan* should be arranged. This cost-effective investigation can reveal the presence of a neoplastic process with a very

■ Table 3.40. Primary bone tumors of the pelvis, proximal femur and femoral shaft in children and adolescents (n=281) compared to adults (n=492). (According to the Basel Bone Tumor Reference Center)

	Children and adolescents						Adults					
	Pelvis		Proximal femur		Femoral shaft		Pelvis		Proximal femur		Femoral shaft	
Osteochondroma	9	13.0%	9	5.3%	8	19.5%	19	11.7%	22	8.7%	2	2.7%
Chondromyxoid fibroma			1	0.6%	1	2.4%	5	3.1%				
Chondroblastoma			11	6.4%			1	0.6%	4	1.6%		
Enchondroma			7	4.1%	5	12.2%	4	2.5%	23	9.1%	2	2.7%
Aneurysmal bone cyst	12	17.4%	9	5.3%	3	7.3%	9	5.5%	22	8.7%	1	1.4%
Non-ossified bone fibroma	1	1.4%	3	1.8%	3	7.3%	1	0.6%	1	0.4%	2	2.7%
Giant cell tumor			5	2.9%			9	5.5%	10	3.9%		
Fibrous dysplasia			24	14.0%			7	4.3%	39	15.4%		
Osteoid-osteoma/osteoblastoma	2	2.9%	27	15.8%	5	12.2%	4	2.5%	12	4.7%	4	5.4%
Other benign tumors	2	2.9%	5	2.9%	1	2.4%	10	6.1%	10	3.9%	17	23.0%
<i>Langerhans cell histiocytosis</i>	6	8.7%	7	4.1%			1	0.6%	2	0.8%		
<i>Unicameral bone cyst</i>	1	1.4%	35	20.5%	3	7.3%	10	6.1%	6	2.4%		
<i>Other tumor-like lesions</i>	9	13.0%					14	8.6%				
<b>Osteosarcoma</b>	<b>2</b>	<b>2.9%</b>	<b>11</b>	<b>6.4%</b>	<b>6</b>	<b>14.6%</b>	<b>11</b>	<b>6.7%</b>	<b>22</b>	<b>8.7%</b>	<b>14</b>	<b>18.9%</b>
<b>Ewing's sarcoma/PNET</b>	<b>20</b>	<b>29.0%</b>	<b>5</b>	<b>2.9%</b>	<b>2</b>	<b>4.9%</b>	<b>9</b>	<b>5.5%</b>	<b>2</b>	<b>0.8%</b>		
<b>Chondrosarcoma</b>	<b>3</b>	<b>4.3%</b>	<b>4</b>	<b>2.3%</b>	<b>1</b>	<b>2.4%</b>	<b>34</b>	<b>20.9%</b>	<b>30</b>	<b>11.8%</b>	<b>3</b>	<b>4.1%</b>
<b>Other malignant tumors</b>	<b>2</b>	<b>2.9%</b>	<b>8</b>	<b>4.7%</b>	<b>3</b>	<b>7.3%</b>	<b>16</b>	<b>9.8%</b>	<b>49</b>	<b>19.3%</b>	<b>29</b>	<b>39.2%</b>
<b>Total</b>	<b>69</b>	<b>100.0%</b>	<b>171</b>	<b>100.0%</b>	<b>41</b>	<b>100.0%</b>	<b>164</b>	<b>100.0%</b>	<b>254</b>	<b>100.0%</b>	<b>74</b>	<b>100.0%</b>

## 3.2.13 · Tumors of the pelvis, proximal femur and femoral shaft

high degree of probability and also provide an indication of its location. In the pelvic area, however, one should also always consider the possibility of soft tissue tumors (■ Fig. 3.247).

If a tumor is suspected, more extensive investigations, e.g. *MRI* and *CT scans* should be arranged. The *MRI* scan is essential for malignant processes. In fact, an *MRI*

scan should be recorded generally for all tumors that can emerge from bone and spread into the surrounding soft tissues with significant consequences. By contrast, tumors that remain within the bone are better visualized with the *CT scan*. This particularly applies to the osteoblastoma, but also to the aneurysmal bone cyst, which commonly occurs in the pelvis.



■ Fig. 3.245. AP x-ray of the proximal femur of a 19-year old male patient with *fibrous dysplasia* and abnormal curvature of the femur (»Shepherd's crook deformity«)



■ Fig. 3.246. Conventional AP tomogram of the right hip of a 14-year old male patient with a *chondroblastoma* in the femoral head



■ Fig. 3.247. *MRI* scan of an 8-year old girl with a large *rhabdomyosarcoma* in the gluteal muscles and resting on the iliac bone. a Frontal section, b horizontal view ( $t_2$ -weighting)



## Site

### Pelvis

Although, in anatomical respects, the pelvis is formed from the ilium, pubis and ischium, the following classification for the site of bone tumors has proved more effective since it is based on the needs of resection, reconstruction and function [23]:

- iliosacral,
- acetabular,
- ischiopubic.

Two-fifths of malignant tumors in each case are located in the first two regions, while the remaining fifth are located in the ischiopubic area [23]. ■ Table 3.40 and ■ Fig. 3.248 show the sites of pelvic tumors recorded in our own register.

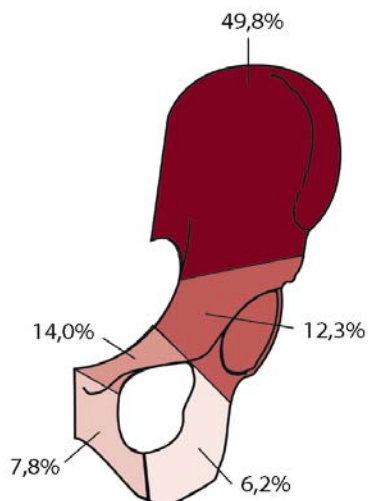
### Proximal femur

In the proximal femur we distinguish between the following sites: epiphyseal (4%), epiphyseal/metaphyseal (15%), metaphyseal (49%), metaphyseal/diaphyseal (13%), epiphyseal/metaphyseal/diaphyseal (4%), diaphyseal (15%; the percentages in brackets relate to the distribution of 491 bone tumors of the proximal femur recorded by the Bone Tumor Reference Center in Basel).

## Treatment of pelvic tumors

### Benign tumors

The common pelvic tumor of aneurysmal bone cyst is usually treated by simple curettage [24]. Recurrences are rare if this procedure is performed with care. Vascularized soft tissues, in particular, must be removed;



■ Fig. 3.248. Distribution of tumors in the pelvis (n=248). The ilium is the most commonly affected site, followed by the pubis and the acetabulum. The ischium is rarely the site of origin of a bone tumor. (Figures from the Bone Tumor Reference Center in Basel)

flat bone and long bones appear to behave differently in this context. Other benign tumors, also, rarely pose therapeutic problems. Osteblastomas must likewise be thoroughly curetted. Osteochondromas should only be removed if they 1) bother the patient, 2) are very large or 3) change in size. In case of doubt, removal is indicated since malignant degeneration occurs rather more frequently close to the trunk compared to the extremities. This particularly also applies to enchondromas, which can occur in the pelvis for example in connection with Ollier enchondromatosis.

## Malignant tumors

### Therapeutic strategies

#### Bone tumors

The therapeutic strategies for the treatment of bone tumors are discussed in detail in ► chapter 4.5.6. Only the regional features will be mentioned at this point. Of the malignant pelvic tumors that can occur in children and adolescents, *Ewing's sarcoma* is the commonest. Since these are usually very large by the time they are diagnosed, the possibility of (micro-)metastases should be considered. As with other sites, chemotherapy is initially administered for 3 months.

If imaging investigations and clinical examination do not show any reduction in the tumor mass, preirradiation may be considered [27]. This possibility must be checked particularly if the tumor cannot be completely resected with a margin of healthy tissue because of its location (e.g. if it grows into the sacrum). A dose of 30–40 Gy is administered for the preirradiation, whereas 60–70 Gy would be required for irradiation of the tumor.

Another option is the combination of preirradiation with hyperthermia. Hyperthermia sensitizes the tumor for subsequent radiotherapy (and incidentally also for chemotherapy [3]). The value of hyperthermia is difficult to estimate. The enthusiastic reports dating back to the 1980's have not been followed up by more recent publications on sarcoma treatment. The drawback of irradiation is the subsequently increased bleeding tendency during resection and the increased postoperative infection risk.

The option of preirradiation does not apply to *osteosarcomas*, nor can chemotherapy even be used in *chondrosarcomas*. All malignant tumors – where resectable – are surgically removed. The surgeon should always aim for a wide resection, with the cut margins extending into healthy tissue. This objective is not always achievable, particularly if the tumor grows into the sacrum. If the accompanying resection of the sacral roots cannot be avoided, then substantial functional deficits must be expected. Distinguishing between healthy tissue and tumor tissue at operation can often prove very difficult precisely in the sacral area. If the resected margins are doubtful, subsequent irradiation is possible in the case of Ewing's



sarcoma, provided the patient has not already been irradiated preoperatively.

### Soft tissue tumors

In the case of rhabdomyosarcomas and other highly malignant soft tissue tumors, the chemotherapy protocols for adolescents and young adults are similar to those for osteosarcoma or Ewing's sarcoma. Here, too, a wide resection is desirable during their removal. Unfortunately, the technique of isolated limb perfusion (see below) cannot be used for tumors in the pelvic area. One possible option for weakly malignant tumors is the drug imatinib mesylate (Gleevec), which has already been used successfully for leukemias and is likewise effective for certain weakly malignant soft tissue tumors. The potential efficacy can be tested with a tumor marker on the tumor specimen. However, the drug is still undergoing clinical trials for the indication of soft tissue tumors.

### Reconstruction options

A hemipelvectomy is extremely mutilating. Because of the lack of anchoring options, a subsequent prosthetic implant is almost impossible. Even just sitting can prove problematic for the patient. Such a disfiguring procedure should therefore be avoided wherever possible. Consequently, an »internal hemipelvectomy« with preservation of the extremity is almost invariably performed nowadays.

If the pelvic ring is interrupted as a result of a tumor resection, a *reconstruction* will be required. The following *options* are available:

- bridging with autologous fibular graft
- removal of the tumor with the pelvic bone, irradiation of the bone and reinsertion at the site of removal
- bridging with allogeneic pelvic bone (allograft)

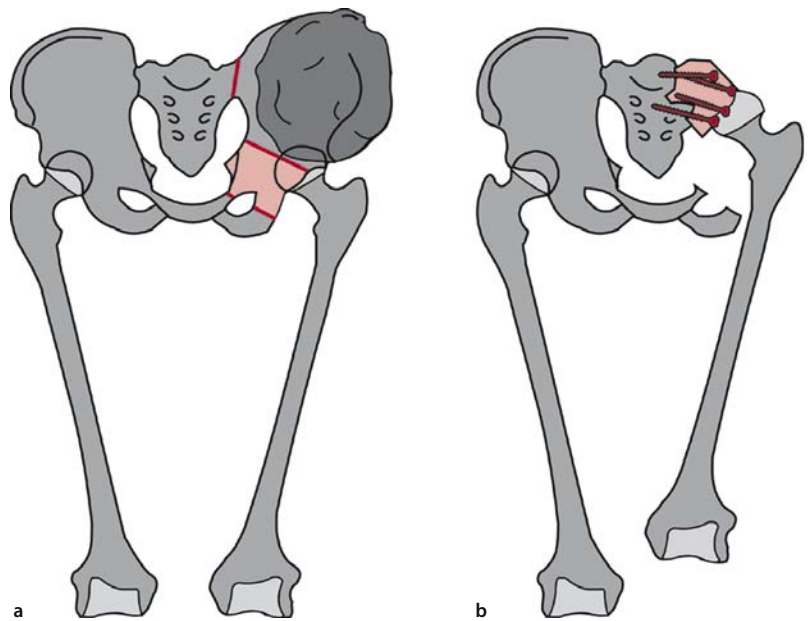
- combination of allogeneic pelvic bone (allograft) with total hip replacement
- bridging with plastic or metal pelvic prosthesis
- fixation of a saddle prosthesis to the residual portion of the ileum
- transposition of the hip to the sacrum [30]

The use of plastic or metal *pelvic implants* has not proved effective, since the anchoring options in the soft pelvic bone and the sacrum are inadequate and unable to provide permanent support. The fixation of a saddle prosthesis to the residual cranial portion of the ileum offers a more durable solution [22]. The best results have been achieved with the use of an *autologous fibular graft*. While the pelvic ring can be reconstructed with fibular segments, this method can only be used if the hip is not (significantly) also affected (■ Fig. 3.249).

If the acetabulum is also involved, the method described by Winkelmann [30] is recommended. In this technique the residual part of the acetabulum is rotated and screwed to the sacrum (■ Fig. 3.250 and 3.251). Although this results in shortening of the leg by a few centimeters, it does produce a stable and permanent situation after the healing phase. The removal of the tumor with the pelvic bone and the *reinsertion* at the site of removal after irradiation is only possible if the tumor has significantly impaired the stability of the bone. Highly osteolytic tumors such as Ewing's sarcoma weaken the bone, whereas this does not apply as much to chondrosarcomas. Extracorporeal irradiation is a good option for the pelvis, provided sufficient stability can be preserved, because the bone fits exactly and offers good conditions for revascularization (similar to that for non-vascularized fibula) [7, 20].



■ Fig. 3.249a, b. 20-year old female patient with Ewing's sarcoma. a AP x-ray of the left pelvis. b 2 years after resection and reconstruction with autologous fibula



**Fig. 3.250a, b.** Principle of transposition of the hip according to Winkelmann after resection of a tumor of the ilium and parts of the acetabulum [30]. **a** Preoperatively; **b** postoperatively



**Fig. 3.251a, b.** 12-year old female patient with a large *Ewing's sarcoma* in the area of the left iliac wing and parts of the acetabulum. **a** AP x-ray. **b** 1 year after tumor resection and *Winkelmann hip transposition*

Another option for reconstruction is the insertion of an allogeneic portion of pelvis (*allografts*; **Fig. 3.252**). Although the mechanical strength of the allograft is less than that of a metal or plastic prosthesis, the anchorage is better. If the pelvic bone is well supplied with blood and a good fit is achieved, the allogeneic bone is gradually transformed into autologous bone over a section measuring 1–2 cm, thus creating the conditions for long-term anchorage [21, 26]. If an allograft is used, the hip should be replaced by a (standard) hip implant. Because of the



**Fig. 3.252.** 20-year old male patient with *clear cell chondrosarcoma* in the left half of the pelvis. **a** AP x-ray. **b** 1 year after reconstruction with an allograft and conventional total hip replacement

better anchorage we prefer this option to an artificial pelvis, even though it likewise does not offer a really permanent solution.

### **Treatment of tumors of the proximal femur and femoral shaft**

#### **Benign and semimalignant tumors**

Surgery may be indicated for a tumor of the proximal femur for the following reasons:

- pain,
- tumor growth,
- mechanical hindrance,
- risk of malignant degeneration,
- loss of stability.

For most of these parameters the indication for treatment does not differ from that for other body regions. The *loss of stability* on the other hand is particularly important for the proximal femur, for example, where it may be an indication for the treatment of tumors which would otherwise not need treatment. This particularly applies to a *unicameral bone cyst*. This tumor-like lesion occurs primarily in the humerus and does not require treatment at this site. Any pathological fractures usually result in healing of the lesion. Spontaneous healing almost always occurs by completion of growth. A spontaneous fracture of the proximal femur, on the other hand, is not so favorable since it does not usually respond adequately to conservative treatment. Accordingly, we occasionally perform a »prophylactic« stabilization with a thin medullary nail (Prévo<sup>t</sup> nail) for juvenile bone cysts [25] (■ Fig. 3.253). As well as providing reinforcement, this procedure also perforates and relieves the cyst.

#### **Malignant tumors**

##### **Therapeutic strategies**

The therapeutic strategies for malignant tumors of the proximal femur are no different from those at other sites (► Chapter 4.5.6).

##### **Resection and reconstruction procedures**

The following *reconstruction options* are available after a limb-preserving resection:

- bridging with autologous fibular graft,
- removal of the tumor with the proximal femur, extra-corporeal irradiation of the bone and reinsertion at the site of removal,
- bridging with allogeneic femur (allograft),
- combination of allogeneic femur (allograft) with total hip replacement,
- bridging with a hip tumor prosthesis.

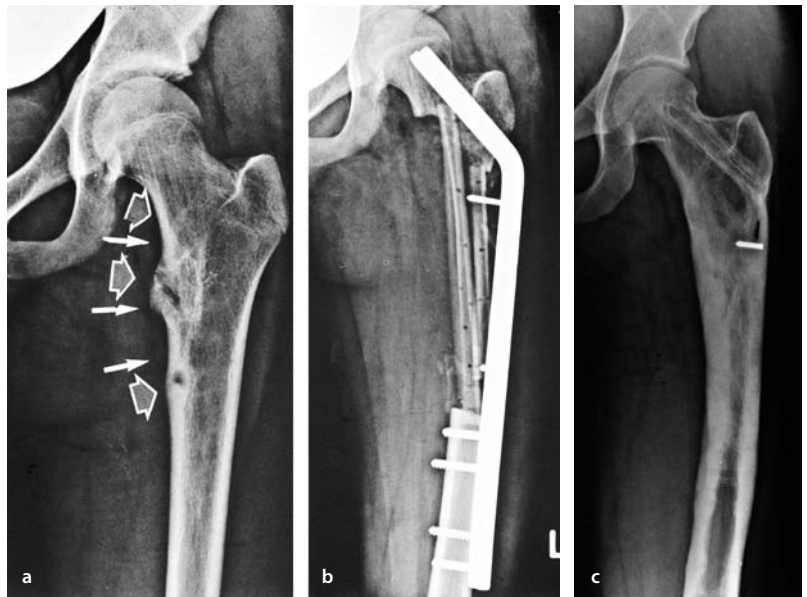
Provided the hip itself can be preserved, bridging with *autologous fibula* is the most suitable method. In adolescents the implanted fibula undergoes remodeling over



■ Fig. 3.253. AP x-ray of the proximal femur of a 10-year old boy with unicameral bone cyst and prophylactic stabilization with Prévo nails

the course of several years, eventually forming an autologous long bone (■ Fig. 3.254). If the hip is also affected, an artificial joint must be inserted. We do not have any experience with the use of an allogeneic hip replacement on its own, although we have employed the *combination of a femoral allograft with a tumor prosthesis* (► Chapter 4.5.6, ■ Fig. 4.64) [9]. The allograft provides better and longer lasting anchorage for the muscles and also for the prosthesis in the proximal part of the femur. The prosthesis for the hip is more durable and mechanically stronger than if the joint was replaced by an allograft alone.

With very extensive tumors it can sometimes prove necessary to replace the femur, including the hip and knee, completely. Allogeneic bone is not a suitable solution for mechanical reasons, and a modular tumor prosthesis must be used (► Chapter 4.5.6, ■ Fig. 4.61).



**Fig. 3.254a–c.** 13-year old girl with a *chondrosarcoma*. **a** AP x-ray of the proximal femur. **b** Situation after resection, bridging with autologous fibula and stabilization with an angled blade plate. **c** 7 years after the operation, the fibula fragments have been remodeled into a femoral long bone with a corresponding medullary cavity

In small children with substantial growth potential, bridging with a standard tumor prosthesis or allogeneic bone is not an effective solution since considerable shortening can occur as growth continues. Moreover, since the femoral shaft also grows in width, loosening of the prosthetic anchorage occur even without the influence of mechanical factors. Although *extendable prostheses* have been developed, these have only been inserted in a few centers as they are associated with a high complication rate [10, 11].

The best and most durable solution therefore is the so-called *rotationplasty*. This technique was first described by Borggreve [4] and later by Van Nes [28] in connection with the treatment of congenital defects. It is also suitable for the treatment of tumors in the knee area. Winkelmann published a modification [29] that is suitable for the treatment of tumors in the region of the proximal femur. After the hip is resected, the distal femur is rotated through 180° and anchored in the pelvis. The knee then functions as a hip, while the rotated foot serves as a knee (▶ Chapter 4.5.6, ■ Fig. 4.66). This method produces excellent, and also lasting, results in terms of function, but poses problems in psychological respects. The child and the parents do not find it easy to accept the rotated foot. Nevertheless, the functional gain, compared to a hip disarticulation or even a hemipelvectomy, is so great that the psychological problems can be overcome, particularly because extendable prosthesis still does not represent a viable alternative.

*Malignant soft tissue tumors* are treated according to the principles described in ▶ chapter 4.5.4.3. For tumors in the lesser pelvis this can mean that nerve roots, or even

the whole of the sciatic nerve, may also have to be resected as well, although the patient still retains the ability to walk. For soft tissue tumors enveloping the sciatic nerve but located below the greater trochanter one possible solution is isolated limb perfusion and the subsequent administration of tumor necrosis factor (TNF) + melphalan (M) [12]. The isolation of the tumor area means that much higher doses of cytotoxic drugs can be administered than would be possible systemically. Low-grade malignant tumors also respond to drug treatment administered in this way.

### Prognosis

Compared to tumors of the extremities, those affecting the pelvis have a much poorer prognosis [14, 15, 17, 18]. This is partly because they are diagnosed at a relatively late stage because of the large surrounding soft tissue masses and partly because they rapidly enter blood vessels in the richly perfused pelvic tissues. Furthermore, it can prove extremely difficult to resect into healthy tissue particularly with large tumors located close to nerves and vessels. Nor is it easy sometimes to evaluate the resected margins at operation in this blood-rich tissue. Thus, a survival rate of just 15–40% has been reported for Ewing's sarcoma at this site [14, 15], i.e. much lower than that reported for the same sarcoma in the extremities.

Most authors believe that resection improves the survival rate [14, 15, 17, 18]. The survival rates for tumors of the proximal femur (depending on the type and size of tumor and the patient's age) are comparable with those affecting the distal femur, where most experience has been acquired (▶ Chapters 3.3.12 and 4.5).



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### 3.2.14 Differential diagnosis of hip pain

Table 3.41 shows the differential diagnosis of hip pain.

Table 3.41. Differential diagnosis of hip pain			
Age group	Signs and symptoms	Tentative diagnosis	Additional measures
Infant (0–2 years)	Poss. fever	<b>Septic arthritis of the hip</b>	Laboratory (infection parameters) X-ray Ultrasound
Toddler/child (2–10 years)	Pain (movement-related) restricted movement, poss. limping	<b>Transient synovitis of the hip</b>	Ultrasound, poss. laboratory (infection parameters)
	Pain (load-related)	<b>Legg-Calvé-Perthes disease</b>	Ultrasound, if no effusion: AP and axial x-rays
	Pain day and night, restricted movement, poss. limping, fever, general illness	<b>Septic arthritis of the hip</b>	Laboratory (infection parameters) AP x-ray, poss. leukocyte scan
	Pain (movement-related)	<b>Juvenile rheumatoid arthritis of the hip</b>	Laboratory (infection parameters, rheumatoid factors), ultrasound
	Restricted movement, poss. limping Pain (day and night) Poss. restricted movement Poss. limping	<b>Tumor</b>	AP x-ray Poss. bone scan Poss. MRI/CT
Adolescent (from 10 years)	Pain (load-related) Restricted movement Poss. limping Poss. obesity	<b>Slipped capital femoral epiphysis</b>	AP and axial x-rays
	Pain (load-related) Restricted movement, poss. limping	<b>Hip dysplasia</b>	AP and Dunn x-rays
	Pain (day and night) Poss. restricted movement Poss. limping	<b>Tumor</b>	AP x-ray Poss. bone scan Poss. MRI/CT
	Pain (movement-related) Restricted movement, poss. limping	<b>Juvenile rheumatoid arthritis of the hip</b>	Laboratory (infection parameters, rheumatoid factors), ultrasound AP x-ray
	Pain day and night, Fever, general illness	<b>Septic arthritis of the hip</b>	Laboratory (infection parameters), AP x-ray, poss. leukocyte scan
	Pain (load-related), restricted movement, poss. limping	<b>Avascular necrosis of the femoral head</b>	Ultrasound, if no effusion: AP and axial x-rays
	Pain movement-related, tenderness around the pubic bone, patient keen on sport	<b>Pectineal bursitis</b>	
	Pain movement-related, (particularly on internal rotation and abduction) Tenderness around the psoas, patient keen on sport	<b>Psoas pain</b>	

### 3.2.15 Differential diagnosis of restricted hip movement

■ Table 3.42 shows the differential diagnosis of restricted hip movement

■ Table 3.42. Differential diagnosis of restricted hip movement			
Age group	Restricted direction of movement	Tentative diagnosis	Additional measures
Infant at birth	Full extension (20°–30° flexion contracture)	Normal findings	None
Infant (from 2 months)	Abduction only up to 70°	Hip dysplasia/dislocation	Ultrasound
Infant (0–2 years)	Internal rotation, poss. all directions of movement	Septic arthritis of the hip	Laboratory (infection parameters) AP x-ray poss. leukocyte scan
	All directions of movement	Arthrogryposis	AP x-ray
Toddler/child (2–10 years)	Internal rotation	Transient synovitis of the hip	Ultrasound, poss. laboratory (infection parameters)
	Internal rotation, abduction	Legg-Calvé-Perthes disease	Ultrasound, if no effusion: AP and axial x-rays
	Internal rotation, abduction, poss. all directions of movement	Septic arthritis of the hip	Laboratory (infection parameters), AP x-ray, poss. leukocyte scan
	Internal rotation, abduction, poss. all directions of movement	Juvenile rheumatoid arthritis of the hip	Laboratory (infection parameters, rheumatoid factors), ultrasound, AP x-ray
	Internal rotation, abduction	Tumor	AP x-ray Poss. bone scan Poss. MRI/CT
Adolescent (from 10 years)	Internal rotation, abduction	Slipped capital femoral epiphysis	AP and axial x-rays
	Poss. internal rotation	Hip dysplasia	AP and Dunn x-rays
	Internal rotation, poss. extension	Tumor	AP x-ray Poss. bone scan Poss. MRI/CT
	Internal rotation, extension, abduction	Juvenile rheumatoid arthritis of the hip	Laboratory (infection parameters, rheumatoid factors), ultrasound, AP x-ray
	Internal rotation, extension, abduction	Septic arthritis of the hip	Laboratory (infection parameters), AP x-ray, poss. leukocyte scan
	Internal rotation, extension, abduction	Femoral head necrosis	Ultrasound, if no effusion: AP and axial x-rays
	Internal rotation, flexion, adduction	Impingement, labrum lesion	AP and Dunn x-rays, Arthro-MRI with radial sequences, poss. CT

### 3.2.16 Indications for imaging procedures for the hip

The indications for imaging procedures for the hip are presented in [Table 3.43](#).

**Table 3.43. Indications for imaging procedures for the hip**

Age	Circumstances/Indication	Tentative clinical diagnosis	Imaging procedures
Infant	Positive family history, positive clinical examination findings, additional malformations	Hip dysplasia	Ultrasound, AP hip x-ray
Infant, toddler, child	Fever, restricted movement, pain, limping, positive laboratory results (CRP, erythrocyte sedimentation rate, white cell count)	Septic arthritis of the hip	Ultrasound (effusion?), AP and axial hip x-rays (Lauenstein), bone scan
Childhood	If, after 1 week, no improvement or fever or positive laboratory results (CRP, erythrocyte sedimentation rate, white cell count)	Transient synovitis of the hip	Ultrasound (effusion?), AP and axial hip x-rays (Lauenstein) bone scan
Childhood	Hip or knee pain, limping, abduction and internal rotation restricted	Legg-Calvé-Perthes disease	AP and axial hip x-rays (Lauenstein), ultrasound (lateral or anterior calcification? subluxation?)
Adolescence	Reduced internal rotation and abduction, Drehmann sign, unclear knee pain and clinical hip findings	Slipped capital femoral epiphysis	AP and axial hip x-rays (Lauenstein)
Any age	After start of walking Any age if pain present	Status after treated DDH	AP hip x-ray AP hip x-ray
Any age	Pain, poss. also in the knee area, poss. inability to walk	Femoral fracture	X-ray in 2 planes
Any age	Pain mainly at night, poss. also in the knee area, poss. also restricted movement	Tumor	X-ray in 2 planes, poss. bone scan, CT or MRI
Any age, neurogenic disorder	Asymptomatic Evidence of pain, clinical instability	Neuromuscular hip dislocation	AP hip x-ray every 2–3 years AP hip x-ray every 6–12 months
Adolescence or later	Pain on walking, sitting	Impingement	AP and Dunn x-rays, Arthro-MRI with radial sequences, poss. CT

### 3.2.17 Indications for physical therapy in hip disorders

The indications for physical therapy in hip disorders are presented in [Table 3.44](#).

**Table 3.44. Indications for physical therapy in hip disorders**

Disorder	Indication	Goal/type of treatment	Duration	Additional measures
Septic arthritis of the hip	Defective healing and restricted mobility, as long as progress is possible	Improve mobility, particularly abduction, but also rotation, improve gait	As long as mobility is restricted and progress is still possible	Poss. exemption from gymnastics, cycling and swimming
Legg-Calvé-Perthes disease	During the first two years of the illness always if movement is restricted. Subsequently, if movement is restricted, as long as progress is possible	Improve mobility, particularly abduction, but also rotation, improve gait	Generally 2 years, poss. longer	Exemption from gymnastics until end stage is reached. Frequent swimming and cycling
Developmental dysplasia of the hip (DDH)	In the older child with persistent dysplasia, poss. postoperatively	Improve gait Improve mobility	As long as mobility is restricted and progress is still possible	–
Intoeing gait	None	Encouragement of sporting activity more useful than physical therapy	–	Operation very rarely indicated; watch for tibial torsion
Slipped capital femoral epiphysis	Postoperatively	Strengthen the muscles (extensors/abductors), improve mobility	Until the patient walks without a limp, mobility is unrestricted or no further progress is possible	No strenuous sport until completion of growth
Femoral fractures	If gait pattern is not normal after 3 months	Walking exercises	As long as patient is symptomatic	–

### 3.3 Knee and lower leg

#### 3.3.1 Examination of the knees

The examination protocol for the knees is shown in **Table 3.45**.

##### History

##### — Has *trauma* occurred?

*If so:*

- When did the trauma happen?
- During what type of activity (sport, play, daily routine)?

- Was direct or indirect trauma involved?
- What movement was involved (flexion? hyperextension? external/internal rotation? varus or valgus stress)?
- Did anything »give way« during the trauma, or was dislocation of the patella observed?
- Was any locking present at the time of the trauma?
- Pain history: Where is the pain located? When does it occur? Is the pain load-related, movement-related, or does it also occur at rest (e.g. while sitting) or even at night? If so, does the pain only occur when the patient changes position or does the patient awake at night because of the pain? For movement-related pain:

**Table 3.45. Examination protocol for the knees**

	Examination	Question
<b>I. History</b>	Ask about trauma	Type of trauma? Rotation component?
	Pain history	Site? Duration? Load-related?
	Ask about stability	Feeling of instability? Giving way?
	Ask about joint locking	Meniscal lesion? Joint mouse? Patellar dislocation?
<b>II. Inspection</b>	Leg position, pelvic tilt, symmetry	Genua valga/vara? Contractures? Leg lengths? Symmetry?
	Position of the patella	Patella alta? Patella baja?
	Contours, indentations	Swelling? Effusion? Popliteal cyst?
	Gait	Limping? Kneeing-in? Range of motion?
	Lateral contours of the supine patient with 90° knee flexion	Lesion of the posterior cruciate ligament?
<b>III. Palpation</b>	Circumference measurements (level of the joint space, 10 cm or 15 cm above)	Trophic status? Swelling/effusion? Quadriceps atrophy?
	Palpation of the patellar margins, shifting of the patella	Anterior knee pain? Bipartite patella? Sinding-Larsen-Johansson?
	Palpation of the joint space	Meniscal injury? Osteochondritis dissecans?
	Palpation of the tibial tuberosity	Osgood-Schlatter disease?
	Palpation of the medial femoral condyles	Medial shelf?
	Palpation of the popliteal fossa	Popliteal cyst?
<b>IV. Range of motion</b>	Dancing patella, milking	Effusion?
	Passive and active, comparison with other side	Locking? Contracture?
<b>V. Ligamentous apparatus</b>	Patellar tracking	Hypermobility?
	Lachman test (drawer test with almost full extension, i.e. in approx. 15° flexion)	Lesion of the anterior and/or posterior cruciate ligament?
	Varus stress	Lateral opening? Lesion of the lateral collateral ligament?
	Valgus stress	Medial opening? Lesion of the medial collateral ligament?
	Drawer test in 60° flexion	Lesion of the anterior and/or posterior cruciate ligament?
<b>VI. Meniscal signs</b>	Pivot shift test	Insufficiency of the anterior cruciate ligament?
	Palpation of the joint space	Backward migration of tenderness during increasing flexion?
	External rotation with increasing flexion	Lesion of the medial meniscus? (the more the knee is flexed the further back the pain migrates)
	Internal rotation with increasing flexion	Lesion of the lateral meniscus? (the more the knee is flexed the further back the pain migrates)



What specific movements elicit the pain (flexion?, external/internal rotation? varus or valgus stress?)?

- Does locking or pseudolocking occur? If genuine locking is present, the knee can neither be flexed nor extended from a particular position for a prolonged period (occurs particularly after a bucket-handle tear of the meniscus). In pseudolocking the knee remains »fixed« in a particular position for a short period, but can be extended again (e.g. in subluxation of the patella, medial shelf syndrome, etc.) by means of certain manipulations (e.g. careful traction).
- Does giving way occur? The patient reports that the knee »gives way« suddenly and unexpectedly during certain movements (typical of anterior cruciate ligament insufficiency).

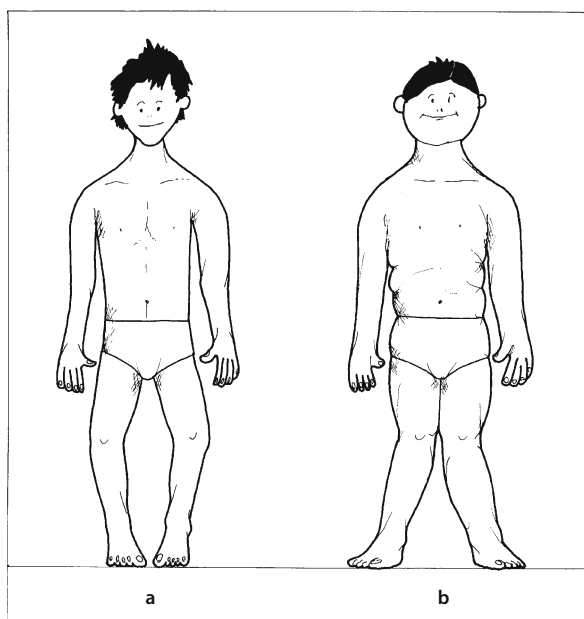
### Inspection

#### Examination of the walking patient

- Is a *limp* present (protective limp or stiff limp)?
- During walking, are the knees extended normally (up to approx. 5° flexion in the stance phase), overextended (complete extension or even hyperextension in the stance phase) or insufficiently extended (remain in flexion of more than 15° during the stance phase)?

#### Examination of the standing patient

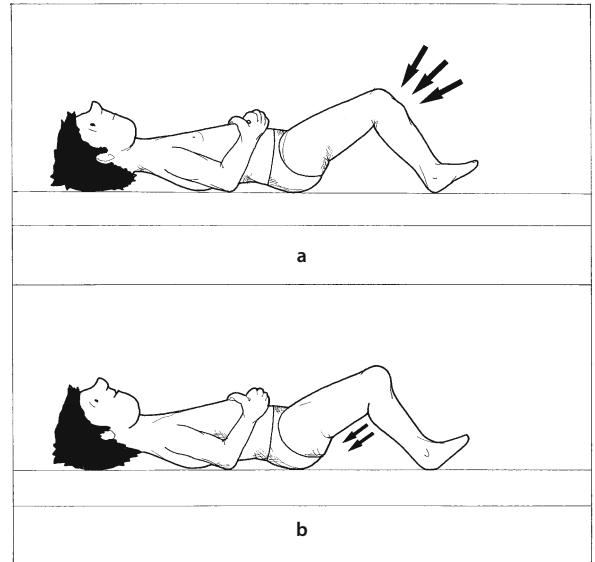
- *Axial deviation* (genu varum, valgum, flexum, recurvatum or combination of several deviations; Fig. 3.255)?
- *Joint contours* (symmetrical or bulging on one side = evidence of local swelling, effusion)?



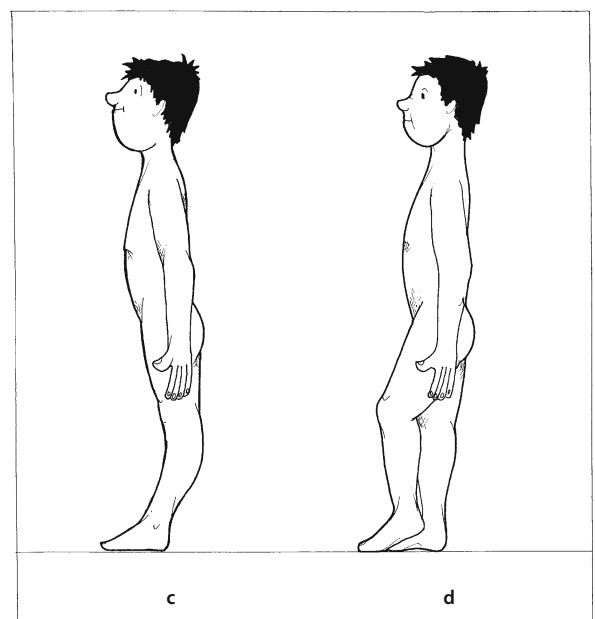
■ Abb. 3.255a–d. Axial deviations. a Genu varum, b Genu valgum, c Recurvated knee, d Genu flexum

#### Examination of the supine patient

- Lateral contours of the tibial tuberosity with 90° flexion of the knee with the patient in the supine position (posterior displacement of the tibial tuberosity compared to the other side is a sign of a lesion of the posterior cruciate ligament; Fig. 3.256)?

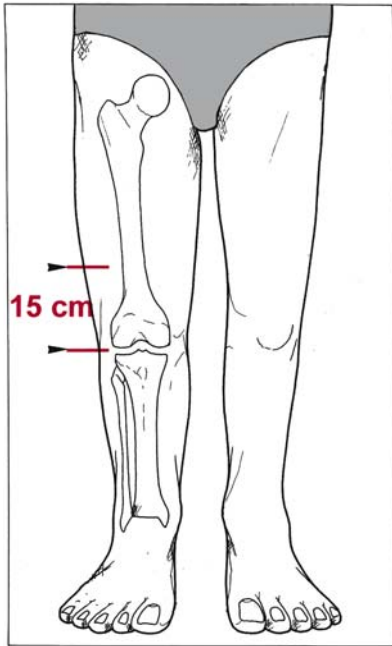


■ Fig. 3.256a, b. Examination of the lateral contours of the proximal lower leg (*posterior drawer*): Viewed from the side, the tibial tuberosity must protrude slightly (a). Posterior displacement (b) – this is particularly easy to see in a comparison of both knees – is a sign of a posterior drawer

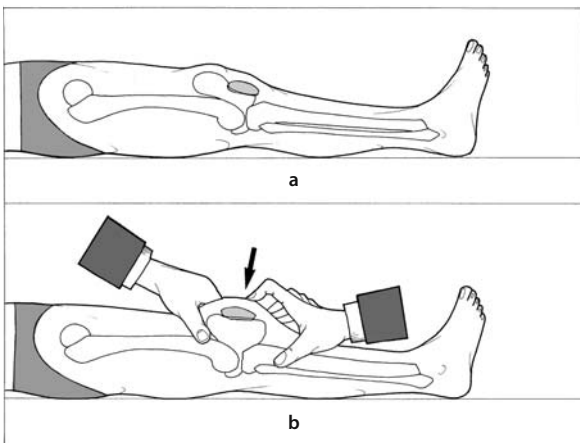


## 3.3.1 · Examination of the knees

- **Circumference measurement:** Mark the knee with a felt pen or ballpoint pen at the level of the joint space and 15 cm above the joint space: measure the circumference at the marks with a tape measure (evidence of muscle atrophy; ■ Fig. 3.257).



■ Fig. 3.257. *Circumference measurement:* Mark the knee with a felt pen or ballpoint pen at the level of the joint space and 15 cm above the joint space: measure the circumference at the marks with a tape measure

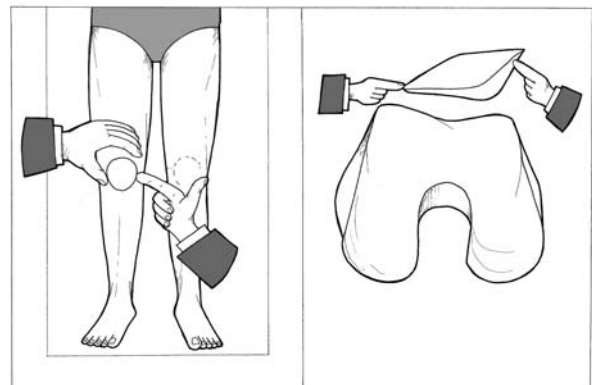


■ Fig. 3.258a, b. *Intra-articular effusion:* Test for dancing of the patella. If a knee effusion is present, the patella will rise from its base. The effusion tends to move into the suprapatellar pouch. The examiner uses both hands to milk the effusion in the direction of the patella (and shift the fluid out of the joint pockets and underneath the kneecap). The »dancing« of the patella on the fluid is now readily discerned by the palpating index finger

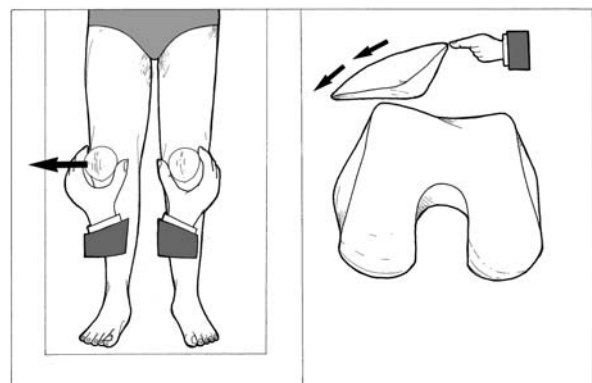
**Palpation**

The knee is usually palpated while the patient is supine.

- **Joint space:** The medial and lateral joint spaces are palpated with a finger from front to back (tenderness, bulging?). If tenderness is present, the examiner establishes whether the painful point migrates posteriorly during increasing flexion (evidence of a meniscal lesion).
- **Joint capsule:** The joint capsule can be palpated at the reflection on the medial and lateral sides of the condyles at the front (soft, thickened, uneven?).
- **Swelling of the periarticular soft tissues** (bursae), tenderness?
- **Intra-articular effusion:** Filling of the joint capsule, »dancing« of the patella during milking of the suprapatellar pouch (■ Fig. 3.258).
- **Palpation of the patellar facets** (■ Fig. 3.259): Push the patella in the lateral direction and check whether this causes pain medially (evidence of tearing of the medial retinacula; ■ Fig. 3.260).



■ Fig. 3.259. *Testing for painful patellar facets:* The index finger of one hand palpates the undersurface of the patella, while the other hand stabilizes the patella from the other side



■ Fig. 3.260. *Testing for stability of the patella:* The patella is pushed away in the lateral direction to check how stably the patella is guided by the retinacula. If this manipulation causes pain medially this is a sign of tearing of the medial retinacula

**Range of motion**

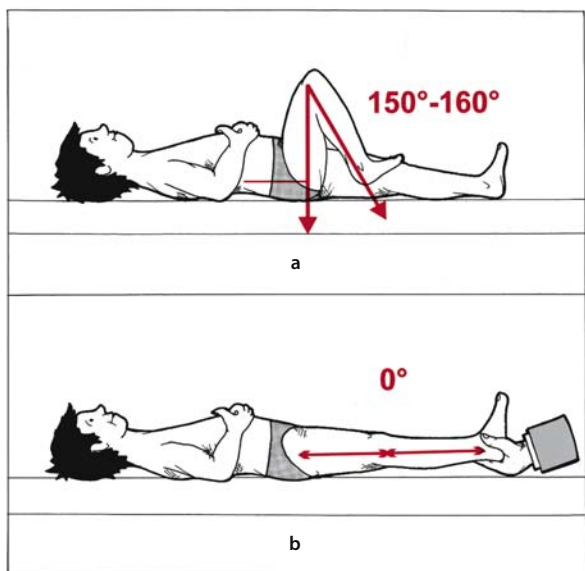
**!** Both sides should always be measured when checking range of motion of the knee.

- Neutral-0 position = extended knee.
- Flexion/extension: The patient is examined in the supine position with the hips flexed (■ Fig. 3.261). Contracture of the thigh muscles is checked by performing the functional test such that the shortened, two-joint muscles are relaxed (flexion with flexed hip and extension with extended hip; ■ Fig. 3.262). Flexion in the child is normally 150–160°. Only in very obese children can this angle be smaller without the presence of any knee pathology. A hyperextensibility of 5–10° is common, particularly in girls where it is often associated with general ligament laxity.
- Rotation: This is locked in the extended position. At 90° flexion the maximum external rotation is approx. 25°, and internal rotation approx. 10°.

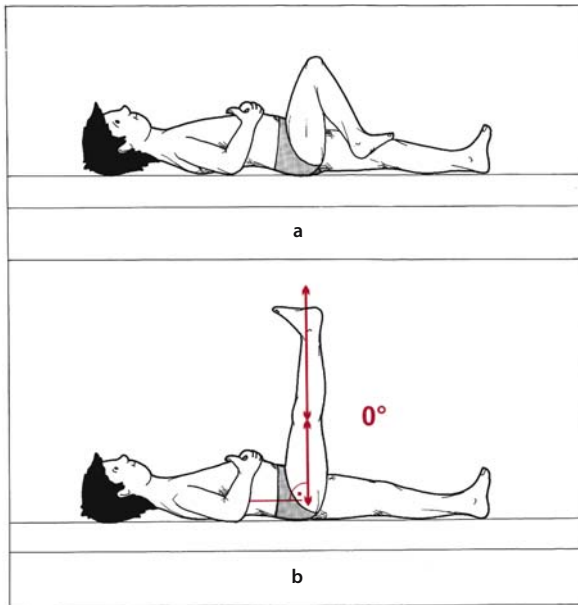
**Stability testing**

- Lachman test, i.e. drawer test with almost full extension: The examiner grasps the thigh with one hand and the lower leg with the other with the knee in 10–20° flexion and tests for the drawer sign anteriorly and posteriorly (■ Fig. 3.263). As well as assessing the extent of movement, the examiner should also note the quality of the end point (whether »firm« or »soft«). This test must always be performed on the other side for the purposes of comparison. This inves-

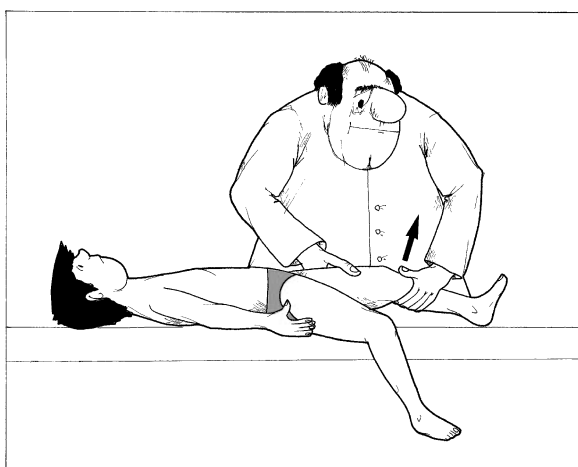
tigation is more reliable for showing the presence of a drawer sign than the same test with more pronounced flexion (see below), since tensing of the hamstrings in the latter case can impair the anterior translation of the lower leg.



■ Fig. 3.261a, b. Knee flexion/extension: a flexion is tested with the patient in the supine position and the hip in 90° flexion. b The leg must be raised slightly at the foot to test for full extension



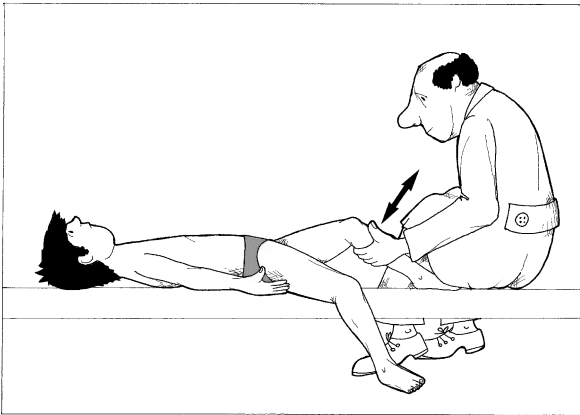
■ Fig. 3.262a, b. Contracture of the hamstring muscles is checked by having the patient flex the hip and knee (a) and then straighten the knee (b). If contracture of the hamstrings is present, this is only possible up to approx. 60° from a position of full flexion



■ Fig. 3.263. Lachman test (drawer test with almost full extension): The examiner grasps the distal thigh with one hand and the proximal lower leg with the other and pulls and pushes the lower leg forwards and backwards in relation to the thigh. Since, in this position of almost full extension, the patient is unable to prevent the drawer movement with his hamstrings, the test is particularly suitable for investigating a recently traumatized joint

## 3.3.1 · Examination of the knees

- **Drawer test** in approx. 60° flexion: With the patient's knee slightly bent, the examiner sits on the foot, grasps the proximal end of the lower leg with both hands and pulls the lower leg forward and then pushes it backward (■ Fig. 3.264). The baseline position is inspected generally beforehand (posterior drawer?). The extent of movement in the AP direction is estimated



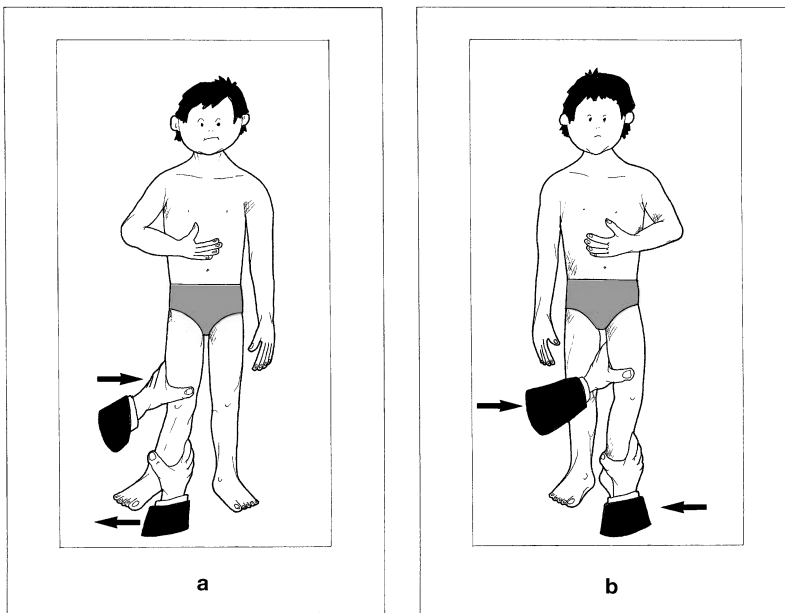
■ **Fig. 3.264.** *Drawer test in approx. 60° flexion:* In the classic drawer test, the examiner stabilizes the foot by sitting on it. The rotational position of the lower leg can then be adjusted appropriately (internal, neutral or external rotation). The examiner grasps the proximal end of the lower leg with both hands and pulls it forward (»anterior drawer«) or pushes it backward (»posterior drawer«). As well as estimating the extent of the translation movement in mm (or using plus signs: + up to 5 mm, ++ up to 10 mm, +++ >10 mm), the examiner also notes the quality of the anterior and posterior end points (»firm«, »soft«)

in millimeters and the quality of the end point is assessed (whether »firm« or »soft«). This test should also be performed on the other side for comparison purposes.

- **Test for lateral opening:** The lower leg and thigh are each grasped with one hand and a valgus or varus stress is applied in approx. 20° flexion (■ Fig. 3.265). Note that a slight degree of lateral (but not medial) opening is normal. If the collateral ligaments are injured, no opening is detected in the extended leg, provided the cruciate ligaments are intact.
- **Test for the pivot shift phenomenon:** The examiner holds the foot on the affected side in one hand and presses the other hand laterally against the proximal lower leg to apply valgus stress. The hand at the foot rotates the lower leg inwardly. The extended knee is now slowly flexed. If anterior cruciate ligament insufficiency is present, an impressive (and painful) reduction click occurs at approx. 30–40° flexion after subluxation. The insufficiency can also be quantified with this test: If it is positive during internal rotation of the lower leg the result is termed +. If it can also be elicited in neutral rotation the severity is termed ++, whereas pronounced instability is present (+++) if the subluxation also occurs in external rotation.

#### Testing for the meniscus signs

Different parts of the menisci are subjected to compression or tension in differing positions of knee rotation and flexion. If a lesion occurs at a specific site, pain can be elicited by rotation and flexion. External rotation places the medial meniscus under tension, while inter-



■ **Fig. 3.265a, b.** *Test for lateral opening:* The thigh and lower leg are each grasped with one hand and a valgus (a) or varus (b) stress is applied

nal rotation exerts tension on the lateral meniscus. The more the knee is flexed, the more the dorsal sections of the menisci are compressed. To test for the meniscus signs we rotate the lower leg in differing flexion positions. The previously mentioned posterior migration of the painful point at the joint space during increasing flexion is characteristic of a meniscal lesion. However, the symptoms are less typical in children and adolescents than in adults.

### References

1. Debrunner HU (1982) Orthopaedisches Diagnostikum. Thieme, Stuttgart New York, pp. 102–105
2. Ruwe PA, Gage JR, Ozonoff MB, De Luca PA (1992) Clinical determination of femoral anteversion. A comparison with established techniques. *J Bone Joint Surg (Am)* 74: 820–30

### 3.3.2 Radiographic techniques

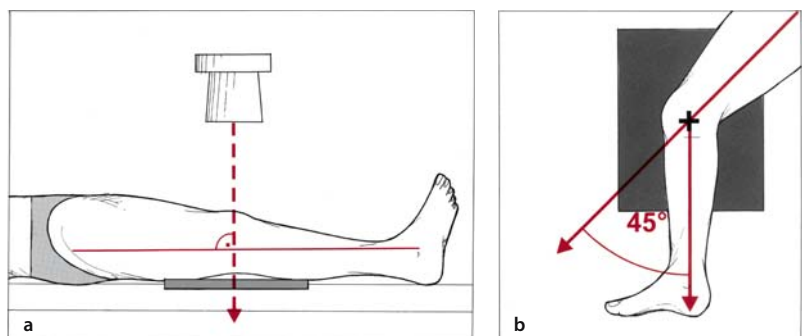
Images of the knee area are usually recorded while the patient is supine, except where axial deformities are involved. In the latter case, x-rays of the standing patient are needed, if possible during single-leg stance. All x-rays are prepared without a grid. Questions about cartilaginous or ligament lesions should be clarified by an MRI scan before arthroscopy. If tumors are diagnosed on a plain x-ray, an MRI is often indicated. Exceptions to this rule are non ossifying fibromas and osteochondromas, which can be diagnosed of plain x-rays, and is the osteoid osteoma, for which computed tomography is the preferred option since it provides a better view of the nidus and enables percutaneous treatment to be administered.

The following standard x-rays of the knee are prepared.

#### AP and lateral view of the knee in the supine position

This is the most frequently used position (■ Fig. 3.266). The lateral view is recorded with the knee in 45° flexion.

■ Fig. 3.266a, b. Recording x-rays of the knee: AP (a) and lateral (b) views in the supine position. For the lateral view the knee should be flexed by 45°

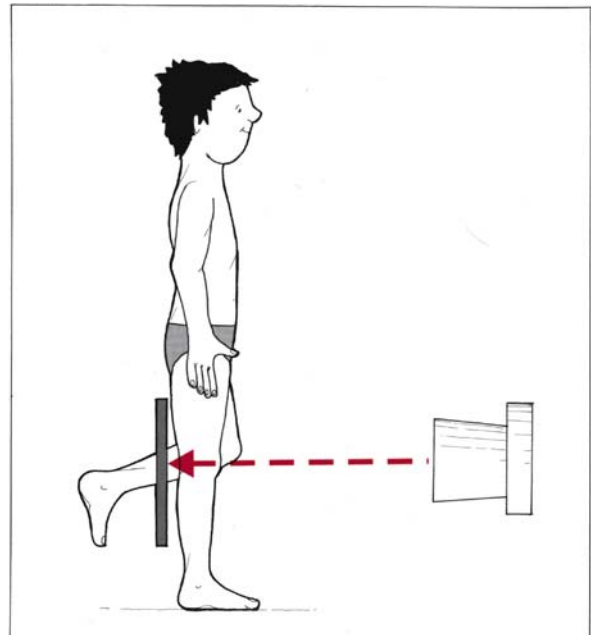


#### Knee with adjacent upper and lower leg in the standing position

An x-ray recorded in the single-leg stance is particularly indicated prior to any scheduled correction osteotomy (■ Fig. 3.267).

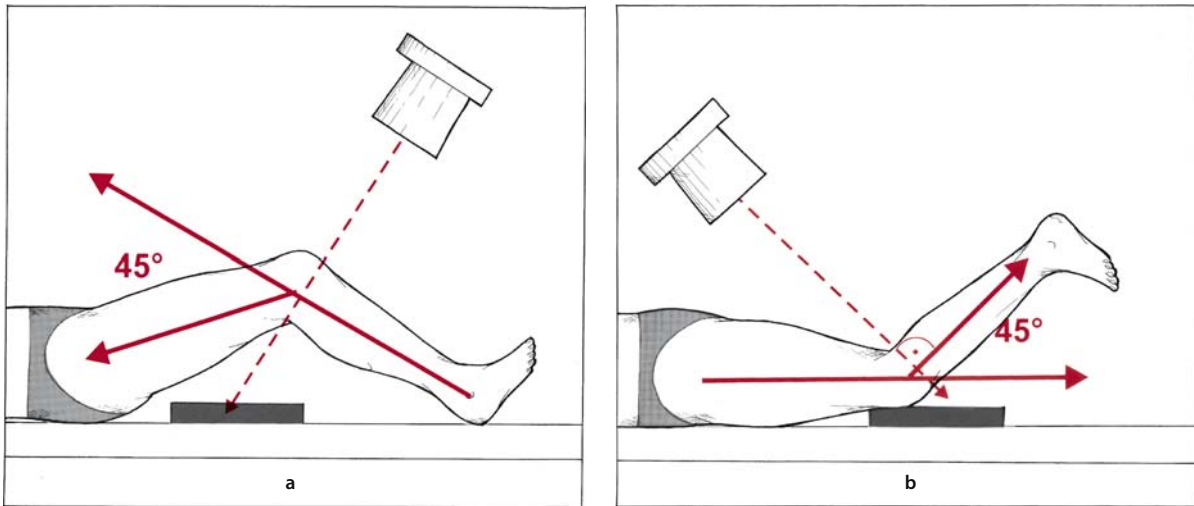
#### Tunnel view according to Frick

This x-ray is indicated in a suspected case of osteochondrosis dissecans (■ Fig. 3.277a in ► chapter 3.3.4), but can also be useful for assessing the intercondylar notch if a proximal bony avulsion of the anterior cruciate ligament is suspected or following cruciate ligament surgery. The patient lies in the supine position with the knee in 45° flexion. The central beam forms is perpendicular to the longitudinal axis of the lower leg and is centered over the inferior pole of the patella (■ Fig. 3.268a). Alternatively, this view can also be recorded with the

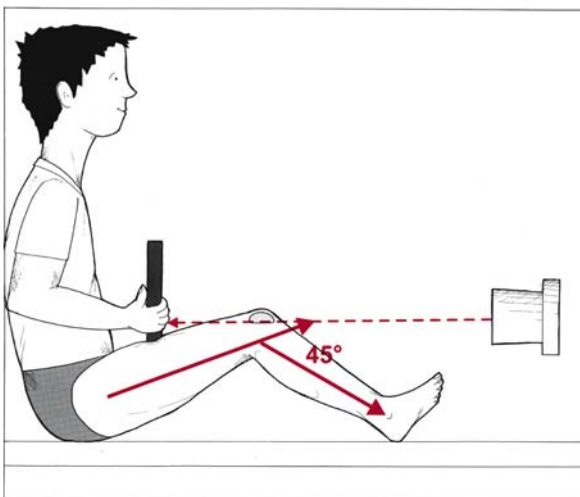


■ Fig. 3.267. Knee with adjacent thigh and lower leg: AP with single-leg stance





■ Fig. 3.268a, b. Recording the tunnel view according to Frick: a in the supine position; b in the prone position



■ Fig. 3.269. Recording an axial x-ray of the patella (tangential) with the knee in 45° flexion

patient in the prone position and the knee flexed by 45° (■ Fig. 3.268b).

#### Oblique view of the knee in the supine position and with the leg extended

The leg is placed in 45° internal or external rotation. These views are useful for clarifying doubtful fractures of the distal femur and proximal tibia and for providing a better view of the medial or lateral edge of the patella.

#### Axial view of the patella (tangential)

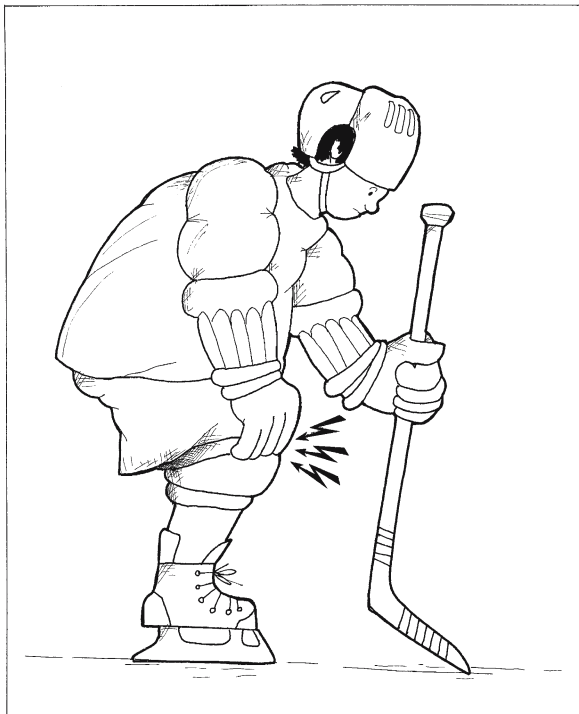
The beam is directed in a caudal to cranial direction. The knee is flexed by at least 30° (■ Fig. 3.269). Views can also

be recorded in 45°, 60° and 90° flexion. The alternative direction, i.e. from cranial to caudal, is also possible.

### 3.3.3 Knee pain today – sports invalid tomorrow? – Pain syndromes of the knee and lower leg

Pain in the knee area is particularly common during adolescence. The numerous reports in the mass media about knee problems in top-class athletes and the occasional case of premature invalidity as a result of a knee injury often raise fears in parents of sporty children with knee pain that their offspring will one day suffer a fate similar to that of some famous sports personality who, according to a television report, was forced to end his or her career prematurely. But knee pain not infrequently occurs in children as well as adolescents. The various causes of knee pain are discussed in this chapter.

The knee is a distinctive joint, whose form and function serve as a symbol for a wide variety of activities and properties in our everyday speech. As with the back and posture (► Chapter 3.1.3), psychology also plays a certain role in this context, and very opposing properties can be described in relation to the knee. When we aggressively reduce someone to a state of submission we »bring them to their knees«, but when we submit to the will of others we »bow the knee« before them. When we are overcome by a strong feeling we »go weak at the knees«. We learn about life »at our mother's (or father's) knee«. When we wish to show special respect or even devotion to someone, we go down »on bended knee«. While the psychology associated with the development and course of knee disorders plays a much less significant role compared to



back disorders, we should not completely disregard this aspect.

### 3.3.3.1 »Growing pains«

#### ► Definition

Growing pains refer to pain that occurs spontaneously and sporadically during early childhood, usually at night, in the lower extremities, predominantly in the knee area. Both sides are alternately affected.

#### Clinical features

»Growing pains« are a little investigated and uncertain phenomenon that occurs during early childhood. Children aged between 3 and 8 wake up during the night and complain of pain, usually in the area of the knee, but also occasionally in the lower leg or foot. Usually one side only is painful, although the other side may cause symptoms another night.

#### Etiology

The etiology of »growing pains« is not fully understood. The idea that growth is responsible for the symptoms is not completely convincing since growth remains relatively constant during early childhood and does not occur in »spurts« as parents repeatedly assume. On the other hand, the fact that the pains occur in the zone of the two epiphyseal plates (distal femur and proximal

tibia), where growth is at its most active, suggests that the increase in height is responsible for the pain. Although overall growth proceeds more slowly during early childhood than during puberty, the increase in the length of the extremities is greater at this stage, whereas spinal growth predominates during adolescence. On the other hand, the greatest growth in the length of long bones occurs at about the age of 10, i.e. rather later than the period of »growing pains«. Cell growth is more pronounced at night than during the day since the growth hormone is secreted primarily at night [3], which would explain the nocturnal occurrence of the pains. Since the condition is harmless and does not have any negative consequences, there is no strong incentive to investigate the etiology with any scientific rigor.

#### Diagnosis

! The most important diagnoses to be considered in the differential diagnosis of »growing pains« are tumors and inflammation.

If the pains occur alternately on the right and left sides, and if the child's age is typical and the knees are clinically normal (normal range of motion, no tenderness, no redness or swelling), no further diagnostic investigations are required, i.e. no x-rays need be taken. Nor will any abnormal findings be detected by other imaging procedures if growing pains are diagnosed.

! If the pains consistently occur on one side, a x-ray is always indicated, and a bone scan is also appropriate in case of doubt.

#### Treatment

The most important treatment is »tender loving care« from the mother. In addition to the pain, children at this age also suffer feelings of anxiety at night and it is important for the mother, or father, to comfort and stay with the child. If drugs need to be prescribed, an anti-inflammatory ointment is better than an analgesic since the physical contact involved in massaging provides another opportunity for showing affection. The oral administration of magnesium also appears to produce a beneficial effect.

### 3.3.3.2 Anterior knee pain

#### ► Definition

Distinctly exertional pain around the patella (usually on the medial rather than the lateral side) that occurs predominantly during adolescence and usually disappears again after the completion of growth.

— Synonyms: Patellofemoral syndrome, , chondromalacia of the patella

### Etiology

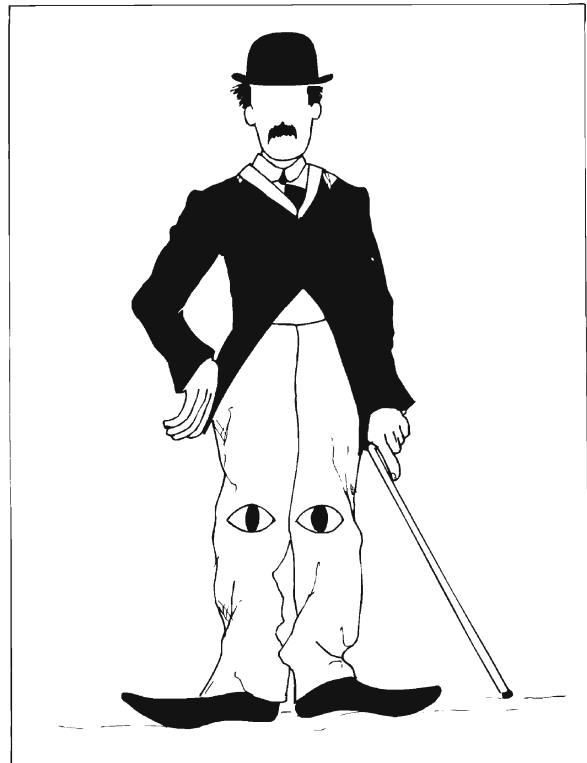
The cause of anterior knee pain is unknown. In the early 1970's, when the era of arthroscopy was just beginning, irregularities of the retropatellar cartilage were often seen in patients with anterior knee pain. Doctors concluded that this was the cause of the pain and accordingly named the condition »chondromalacia of the patella«. It subsequently emerged, however, that such irregularities also frequently occur in patients without any form of retropatellar pain (who were arthroscopied for completely different reasons) and that these cannot therefore explain the symptoms. Based on the epidemiological findings, the following tentative conclusions can be drawn in respect of the etiology: The pain occurs during the pubertal growth spurt, is particularly severe when retropatellar pressure is high (walking downhill), and particularly affects tall (asthenic) girls with proportionately weak muscles. The symptoms improve on conclusion of the growth phase. An imbalance must exist, therefore, between the rapidly rising retropatellar pressure situation resulting from the growing lever arms on the one hand and muscle (and ligament) control on the other. We can now say for sure that a »disorder« of the retropatellar cartilage is not present in most patients and that the term »chondromalacia« should not therefore be used (apart from a few arthroscopically confirmed, usually posttraumatic, cases).

New findings of relevance to the etiology have emerged in recent years. Investigations with computed tomography have shown that patients with anterior knee pain have significantly higher degrees of femoral anteversion compared to a control group [4, 6]. No other differences between the two groups were observed in respect of knee parameters (shape of the condyles and the patella). The authors also noted a connection between femoral anteversion and osteoarthritis of the knee [5]. While, in the 1970's, we still believed that increased anteversion was a problem for the hip, it now appears to be more of a problem for the knee. Although the connections with tibial torsion have not been studied sufficiently to date, since the knee position during walking correlates more closely with the rotation of the lower leg and feet than the rotation of the thigh, such a connection probably does exist. Abnormal rotation of the knee during walking leads to abnormal loading.

**!** **Rotation anomalies probably have a greater influence on the loading of the knee than axial deviations – a problem that has been almost completely ignored to date.**

### Clinical features

Anterior knee pain is common in adolescence and is typically characterized by the following factors: Girls



Charlie had no anterior knee pain because he turned outwards to make his knees point straight ahead; the problem occurs primarily when the knees do not face the direction of walking...

are more frequently affected than boys, they tend to be keen on sports but are typically of an asthenic rather than an athletic build and general ligament laxity is also often present. The pain is at its strongest after physical exertion. It is particularly pronounced when walking downhill and, to a slightly lesser extent, when walking uphill. *Clinical examination* reveals a normal-looking knee with tenderness over the patellar facets, usually on the medial rather than the lateral side. One specific test is for the »Zohlen sign«: With one hand the examiner grasps the top of the patella and presses it against the femoral condyles. The patient is then asked to tense the quadriceps muscle. The reporting of pain by the patient suggests the presence of a patellofemoral syndrome. Since, in my experience, this examination proves painful in every case if sufficient pressure is applied, its value in differential diagnosis is doubtful. Crepitation is a non-specific sign and is not an indication of retropatellar pathology in young patients. In an investigation involving 123 young adults, crepitation was noted in 60% of cases, whereas retropatellar pain was present in only 3% [1]. Contracture of the quadriceps muscle (restricted knee flexion with hips extended) is also occasionally observed. Anteversion (■ Fig. 3.135) and tibial torsion /

the foot axis (■ Fig. 3.352) should also always be checked as rotation anomalies are frequently associated with anterior knee pain. Since anterior knee pain can usually be diagnosed on clinical examination, *radiographic investigations are not generally needed*. Axial x-rays of the patella are often prepared, occasionally as »défilé« views in flexion positions of 30°, 60° and 90° flexion. However, none of these views is suitable for assessing subluxation of the patella, since a subluxating patella reduces itself even in 30° flexion. Since axial x-rays of the patella in less than 30° flexion are not technically feasible, only *computed tomography* is helpful in this situation. CT images of the extended leg, with and without tensing of the quadriceps muscle, are useful for assessing the centering of the patella (► Chapter 3.3.5). Much attention has been paid to the shape of the patella as observed on axial views. The classification according to Wiberg is based on differences in the medial patellar facet. However, these differing shapes are of no relevance to the course of the painful symptoms and, therefore, to the treatment. If a CT scan is ordered and a rotation anomaly is clinically suspected, then the corresponding rotations should, if possible, be measured by means of appropriate sections (■ Fig. 4.8).

### Treatment

❗ **In view of the generally benign course of anterior knee pain during adolescence, we would explicitly warn against any overtreatment!**

Useful measures include strengthening of the quadriceps muscle, and possibly also the stretching of this muscle, since the etiology assumes the presence of an imbalance between muscle strength and the growing lever arm. This can be achieved (in the short term) by physical therapy. In the long term, however, graduated sports-based stamina training is indicated. Although the pain occurs mainly after sporting activities, a complete ban on sports should not be ordered. Rather, the quadriceps muscle should be strengthened in a targeted and graduated manner. Whether the vastus medialis muscle plays a particular role is the subject of debate. This is probably the case in only a few specific situations, e.g. if the muscle is attached to the patella at a very high point and general ligament laxity also results in inadequate centering of the patella. Another conservative treatment option is the use of a knee support incorporating a pad surrounding the patella (■ Fig. 3.270). Since this pad is designed to guide the patella it is particularly indicated for patients with flaccid ligaments. Although its efficacy is based more on psychological than mechanical factors, we have nevertheless been able to achieve success with such supports in a few intractable cases.

Extreme caution is indicated when it comes to surgery. The English surgeon Goodfellow once said that he



■ Fig. 3.270. A knee support with a pad surrounding the patella can be used in patients with ligament laxity and peripatellar pain

had never seen a patient who had *not* undergone surgery become disabled as a result of anterior knee pain. This sentence implies that the opposite is perfectly possible [17]. Such patients often undergo further surgical procedures, because of the unsatisfactory initial result, eventually impairing the circulation in the patella and causing dystrophy. In such cases, the ultimate (and very bad) solution is patellectomy.

This problem is discussed in ► chapter 3.3.5 in connection with patellar dislocation and is even more serious after a patellofemoral syndrome, since the latter (compared to patellar dislocation) has a better prognosis if left untreated. Nevertheless, a long-term UK study involving adolescents in Oxford who had previously been investigated for anterior knee pain [17] showed that, while 90% were still taking part in sports after an average of 16 years, 3/4 of the patients still had significant symptoms [13]. On the other hand, there is no evidence to date to indicate that surgery is capable of improving this prognosis. Although a link appears to exist between rotation anomalies and anterior knee pain we cannot generally recommend



surgical correction of rotation anomalies. In ► chapter 4.2 however, we describe the correction of a pathologically increased tibial torsion in a patient under 10 years of age by means of a supramalleolar osteotomy, as this procedure is safe and simple and produces a cosmetically convincing result, and the femoral anteversion still has time to normalize itself spontaneously.

On the other hand, children with anterior knee pain are almost always older than 10 years and the corrective procedure much more elaborate since it often has to include both the upper and lower legs. Insufficient data are available concerning osteoarthritis of the knee in later life to enable such a procedure to be recommended.

### 3.3.3.3 Osgood-Schlatter disease

#### Definition

Avascular necrosis and traction-induced inflammation in the area of the tibial tuberosity in adolescents. Boys are more commonly affected than girls.

— *Synonym:* Osteochondritis of the tibial tuberosity

#### Historical background

The disease was first described by Osgood [16] and Schlatter [18], independently of each other, in 1903.

#### Occurrence

In one epidemiological study, Osgood-Schlatter disease was observed in 21% of cases in a group of school students who were very active in sports. In another group of students who were not very active in sports, the same disease was only found in 4.5%. Both knees were affected in approx. 25% of cases [10].

#### Etiology

Osgood-Schlatter disease is an avascular necrosis and aseptic inflammation triggered by repetitive stress as a result of traction exerted by the patellar tendon on the immature, still cartilaginous apophysis. It occurs during a phase of particularly strong growth activity (puberty). The mechanical traction eventually produces microtraumas in the hormonally weakened growth plate. One recent investigation has shown a link between patella alta and Osgood-Schlatter disease [2].

#### Clinical features, diagnosis

Exertional pain is experienced in the area of the tibial tuberosity, typically *after* sporting activity. Clinical examination reveals tenderness on palpation of the tibial tuberosity. It may also be possible to elicit the pain by asking the patient to raise the extended leg against resistance. A doughy swelling, generally unaccompanied by any inflammation, is also occasionally observed in the area of the tibial tuberosity. The history and examination

usually leave no doubt about the diagnosis. A lateral *x-ray* is non-specific in most cases, although fragmentation of the tibial tuberosity is sometimes observed (■ Fig. 3.271), possibly as a result of repair attempts after the occurrence of microtraumas. On the other hand, fragmentation of this apophysis is also occasionally seen in completely asymptomatic patients. More extensive imaging investigations are not necessary.

#### Treatment

Osgood-Schlatter disease is treated conservatively. The parents and child should understand that the healing period in the bradytrophic tissue of a growth plate under traction can last 1–2 years. In the acute stage, ice treatments, physical therapy and massaging with ointments can be beneficial. Anti-inflammatory drugs are not advisable since they hardly influence the course of the disease and have to be taken for a very long time. By contrast, a cylinder cast worn on the extended leg for 6 weeks can be useful in cases of very persistent pain. The efficacy is based less on the immobilization itself, than the fact that the – usually very sporting adolescents – are prevented from practicing their sport for a prolonged period and thus subjecting the apophyseal plate



■ Fig. 3.271. Lateral *x-ray* of the left knee of a 15-year old boy with fragmented tibial tuberosity in *Osgood-Schlatter disease*



to constantly repeated microtraumas. Surgical treatment is only indicated in Osgood-Schlatter disease in one situation: If a loose and irritating fragment in the area of the patellar tendon attachment is still protruding in the full-grown patient, the removal of this sequestrum may be indicated.

### 3.3.3.4 Sinding-Larsen-Johansson disease

#### ► Definition

Aseptic bone necrosis in the area of the distal pole of the patella in adolescents.

#### Historical background

The disease was described in 1921 by Sinding-Larsen [19] and in 1922 by Johansson [8].

#### Etiology

Like Osgood-Schlatter disease, this disorder also involves an avascular necrosis of the cartilaginous tendon attachment resulting from repetitive microtraumas. In Sinding-Larsen-Johansson disease, however, it is the proximal rather than the distal attachment of the patellar tendon that is affected.

#### Clinical features, diagnosis

The patients complain of pain in the patella region after strenuous sporting activity. Clinical examination reveals tenderness in the area of the distal pole of the patella and occasionally slight swelling as well. Pain can also be elicited at this site if the patient tries to elevate the stretched leg against resistance. In contrast with Osgood-Schlatter disease, the *radiographic findings* in Sinding-Larsen-Johansson disease are significant. Osteolysis in the area of the distal pole of the patella is not normal and is characteristic of this disease (■ Fig. 3.272). A tumor is the only other possible alternative explanation. If doubt exists, a bone scan can clarify the issue. Uptake is only slightly increased in Sinding-Larsen-Johansson disease, in contrast with the situation for a tumor. If clinical symptoms are present but the x-ray is normal, one possible diagnosis to consider is »jumper's knee«. This is similar to the pathological condition observed in full-grown patients, but instead of occurring at the cartilaginous tendon attachment, the necrosis affects the tendon itself and is not visible on the x-ray. This condition is typically seen in athletes who practice jumping sports (long jump, high jump, triple jump, volleyball, basketball, etc.).

#### Treatment

Since this pathological condition is similar to Osgood-Schlatter disease, but simply occurs at the other end of the same tendon, the same therapeutic measures are indicated. In active jumpers, the cause is not infrequently



■ Fig. 3.272. Lateral x-ray of the left knee of a 12-year old girl with avascular bone necrosis at the distal pole of the patella (*Sinding-Larsen-Johansson disease*)

an incorrect jumping technique. In collaboration with the trainer, the error should be identified and eliminated by careful training.

### 3.3.3.5 Bipartite patella

#### ► Definition

The presence of two or more ossification centers in the patella.

#### Occurrence

The incidence is not known. Boys are nine times more frequently affected than girls, and the condition is usually unilateral. The superior lateral pole of the patella is affected in 75% of cases, the lateral margin of the patella in 20% and the inferior pole of the patella in 5% of cases [20].

#### Etiology

The bipartite patella is a congenital condition. The fact that a bipartite patella is hardly ever seen on x-rays of adults indicates that unification of the ossification centers occurs during the course of maturation. The symptoms occur when the synchondrosis is loosened as a result of trauma or chronic stress.

**Clinical features, diagnosis**

The existence of more than one ossification center in the patella is not, in itself, a cause of symptoms. Only if trauma loosens the cartilaginous joint does pain result. Clinical examination usually reveals a fairly large patella with tenderness over the synchondrosis between the accessory and main fragments. If the tenderness is highly localized and not pronounced, the radiological diagnosis of »bipartite patella« should be classed as a chance finding with no pathological significance. The condition is diagnosed on the basis of the AP x-ray (■ Fig. 3.273), and no further diagnostic investigations are required. In contrast with a fracture, the fragment has smoothly defined margins. Neither a bone scan nor an MRI scan will be able to show whether the synchondrosis is loosened or not.

**Treatment**

Conservative treatment with local anti-inflammatory measures and possibly immobilization in a cylinder cast should be tried initially. Surgery may be considered if symptoms persist. In the past we used to remove the separated fragment. Although this usually relieved the symptoms, we still do not know enough about the long-term effect of this partial resection. While we ourselves have never observed any adverse effects, a more recent method for fragments that are not particularly mobile seems preferable in my view. In this procedure only the insertion of the vastus lateralis muscle is detached subperiosteally. This is a reliable method for relieving the symptoms [14]. There are also reports of successful screw fixation of the fragment [7].

**3.3.3.6 Medial shelf syndrome****► Definition**

Connective tissue septum running from the medial recess towards the patella and which can rub on the edge of the medial femoral condyle.

**Etiology**

The mediopatellar plica is an embryonic remnant. During fetal development circulation to the knee is ensured by means of a vessel that passes through this plica, while a similar connective tissue fold with an artery is present laterally. The vessel is obliterated during the course of subsequent development and is no longer present to any appreciable extent in the neonate, although the plica persists. While its actual existence is a normal finding, its anatomical configuration can vary. Its presence was first established with the introduction of arthroscopy. Evaluating its pathophysiological significance, however, can prove problematic.

**! Very few plicae are responsible for symptoms in the adolescent knee.**

In isolated cases, a plica with a very sharp edge in a fairly tight knee can rub over the medial femoral condyle during increasing flexion, producing cartilage damage or synovitis at this point. The lateral plica hardly ever causes painful sensations.

**Clinical features, diagnosis**

Patients complain of exertion-related knee symptoms on the medial side of the patella. Pseudolocking and snap-

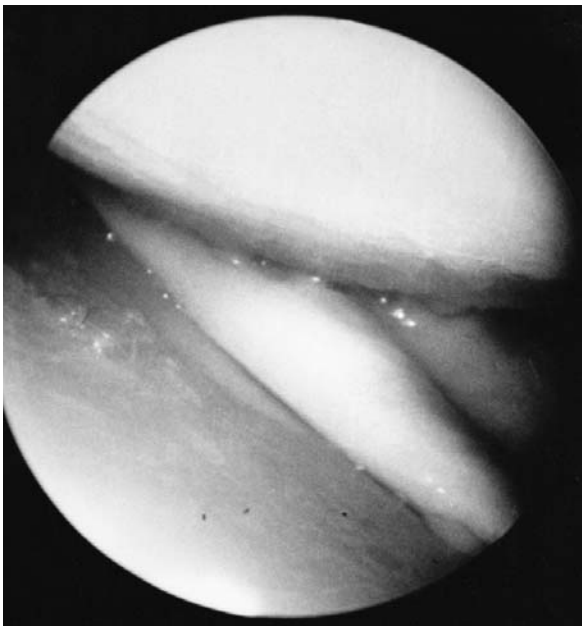


■ Fig. 3.273a, b. 10-year old girl with *bipartite patella* (separate ossification center of the superior lateral pole of the patella). a AP x-ray of the left knee, b axial x-ray of the patella

ping phenomena are occasionally present. On clinical examination, a band running over the medial femoral condyle that moves during flexion and extension is palpated in an otherwise normal knee. For diagnostic purposes, it is very important to establish whether the patients experience this pain as a diffuse or localized symptom during palpation of this band. Snapping may occur during active flexion between 30° and 60°. If the examiner pulls the patella towards the lateral side, traction on the plica is increased, potentially eliciting pain. Patients with a symptomatic mediopatellar plica tend to have fairly tight knees with no general ligament laxity. A tentative diagnosis of medial shelf syndrome is confirmed on clinical examination. While the mediopatellar plica is visible on MRI, such a scan does not provide any information about its pathological significance. Since the plica is a physiological phenomenon and always present, we consider that an MRI scan is not indicated for confirming a tentative diagnosis. Since other imaging procedures are not helpful either, the definitive diagnosis must be confirmed by arthroscopy. This shows a sharply defined white plica running from the medial recess toward the patella (■ Fig. 3.274); cartilage damage, including an inflamed synovial membrane, is observed at the edge of the medial femoral condyle.

### Treatment

There is no known conservative treatment for medial shelf syndrome.



■ Fig. 3.274. Arthroscopic view of a *mediopatellar plica* in a 12-year old girl: top patella; bottom medial femoral condyle, between the two is the cord-like plica, which rubs over the femoral condyle

❗ **The decision to proceed to arthroscopic resection must be taken with extreme caution.**

We consider that arthroscopy is indicated only if the following conditions are satisfied:

- palpable mediopatellar band,
- pronounced, very localized tenderness at this site,
- duration of symptoms more than 3 months,
- snapping between 30° and 60° flexion.

Resection during arthroscopy is indicated only if the following conditions are fulfilled:

- very sharply-defined, tight medial plica,
- cartilage damage at the medial femoral condyle,
- with adjacent synovitis.

Provided these conditions are observed and plica resection is cautiously indicated, a high success rate can be achieved in treating the medial knee symptoms of these patients [9]. While the plica resection can be performed effectively during arthroscopy, the problem can likewise be solved with minimum morbidity by means of open resection via a small incision.

### 3.3.3.7 Stress fractures of the lower leg

#### ➤ Definition

Tibial fracture or fissure resulting from repetitive bending loads, associated with chronic pain and characterized by reactive new bone formation.

- *Synonyms:* Fatigue fracture, shin-splint syndrome

#### **Etiology, localization, occurrence**

Stress fractures occur, particularly in adolescent bone, as a result of repetitive bending loads [11]. In one study of 369 stress fractures among recruits of the Finnish army, the tibia was the commonest site, occurring in 52% of cases. The metatarsals represented another common site (13%), whereas all other bones were only rarely affected. 3–5% of all recruits suffered a stress fracture [11]. But such fractures occur not just in young adults, but also occasionally in very active sporting adolescents.

#### **Clinical features, diagnosis**

The patient reports a history of chronic, exercise-related pain roughly at shin level. The symptoms occur particularly in very active adolescents and can last for months. Clinical examination and palpation may reveal protuberance of the anterior tibial margin and local tenderness. Thickening of the cortical bone, usually on the anterior side, is observed on the *x-ray* (■ Fig. 3.275). The actual fracture is not always visible – and even if it is visible rarely manifests itself as a typical fracture gap –, but only diffuse osteolysis of varying degree as the consequence of

repair processes. A *bone scan* shows pronounced uptake. Other imaging procedures (CT, MRI) are not usually needed.

The most important *differential diagnosis* is an *osteoid osteoma*. Here, too, thickening of the cortical bone occurs and the uptake on the bone scan is similar. The osteolysis of the stress fracture can easily be misinterpreted as the nidus of an osteoid osteoma. The most important distinguishing feature is the fact that the pain resulting from a stress fracture is clearly exercise-related, whereas the osteoid osteoma very typically produces nocturnal pain that responds very well to the administration of acetylsalicylic acid.

### Treatment

Treatment involves the exclusion of the causal stress, i.e. a temporary ban on the excessively practiced sport. The



■ Fig. 3.275. AP and lateral x-rays of the lower leg in a 13-year old boy with a *stress fracture* beneath the center of the tibia with anterior cortical thickening. The slight osteolysis corresponds to the organizing fracture and should not be confused with the nidus of an osteoid osteoma

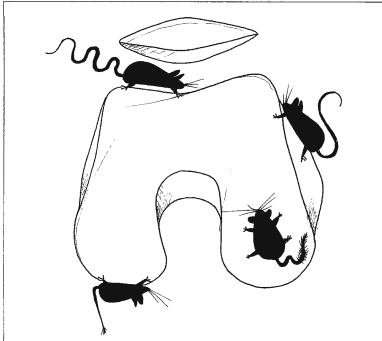
most effective treatment is the fitting of a short-leg cast as this is the best way of imposing the sports ban. We use the »Sarmiento cast«, a short-leg cast that supports the patella, thereby substantially relieving the tibia without immobilizing the knee. A check x-ray is taken after 4 weeks, by which time the fatigue fracture has usually healed. A change in footwear may be useful as a prophylactic measure. Soles with a high proportion of polyurethane and air cells can provide effective prophylaxis for runners against excessive bending moments [12].

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### 3.3.4 Osteochondritis dissecans

» If the mouse is still in its lair, sport will soon drive it from there. «



#### ➤ Definition

Necrotic focus usually localized on the lateral curve (facing the intercondylar notch) of the medial femoral condyle with surrounding sclerosis of the bone, which can then become detached and remain in the joint as a loose body (»joint mouse«).

#### Historical background

- 1558: Ambroise Paré observes loose joint bodies in the knee [31].
- 1879: Paget suspects that loose joint bodies are caused by avascular necrosis [30].
- 1887: König explains the etiology in terms of trauma and hereditary factors [20].

#### Etiology

The most important etiological factors are:

- hereditary factors,
- trauma.

*Hereditary factors* probably play the most important role. In an investigation of 122 lesions, half of the patients had multiple lesions and 30% were of small stature [25]. Other factors such as endocrine dysfunction, collagen disorders and epiphyseal anomalies (e.g. multiple epiphyseal dysplasia, discoid menisci [2]) were also more frequently associated with osteochondritis dissecans. Other authors report a familial occurrence of the disease in several joints [21].

Other studies showed an increased incidence among very physically active patients, suggesting that *trauma* also plays a role in its development [4]. One biomechanical study with a three-dimensional model showed that high shear forces, particularly around the medial femoral condyle, occur during flexion under load. These forces reach a peak at 60° flexion. The deformation of the carti-

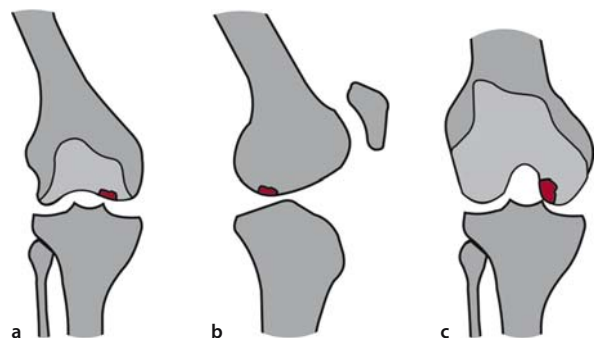
lage surface at the typical site of osteochondritis dissecans, i.e. the medial femoral condyle, is very marked, indicating that mechanical factors play a significant part in the etiology [26]. The shape of the loose body allows certain conclusions to be drawn about the etiology: i.e. in the traumatic form the walls of the loose body are less steep than in the idiopathic form [6]. Ossification of the femoral condyle takes place in very differing ways. In many children, for example, isolated ossification centers or islands are observed, and these can subsequently develop into a case of osteochondritis dissecans.

#### Classification, occurrence

We make a distinction between a more common *juvenile form of the disease* (with open epiphyseal plates at the start) and a rarer *adult form* (with closed or premature plates) [33]. The juvenile type rarely begins before the age of 10 and has a better prognosis than the adult form. Since the prognosis deteriorates even a year before plate closure, we include boys up to the age of 14 and girls up to the age of 13 in the juvenile form. A *systemic form* with multiple lesions affecting several joints also exists [25]. In this form, both knees, and frequently both elbows as well, are usually affected. In the latter case, the condition is known as Panner's disease. In a multicenter study of the European Paediatric Orthopaedic Society directed by the author, the male:female ratio in 798 cases of osteochondritis dissecans was approx. 2:1 [17]. Both sides were affected with equal frequency and both knees were affected in 11% of cases. While there are no precise figures about its occurrence, the disease is fairly rare.

#### Site

The typical site is the lateral curve, facing the intercondylar notch, of the medial femoral condyle. This is a concave surface with a relatively small curve radius. Generally speaking, osteochondritis dissecans only occurs on concave surfaces. In the above-mentioned multicenter study of 798 cases, 70% were located at this site (■ Fig. 3.276 and 3.277a). 7% were on the medial side of the medial femoral



■ Fig. 3.276a–c. Osteochondritis dissecans: *Typical site* on the laterodorsal section of the medial femoral condyle



## 3.3.4 · Osteochondritis dissecans



■ Fig. 3.277a–c. 14-year old girl with *osteochondritis dissecans* on the lateral section of the medial femoral condyle. a (x-rays: AP, lateral and tunnel view according to Frick). The large fragment in the classi-

cal site is fully dissected, and a slight varus axis is present. b Situation after screw fixation of the fragment and *transcondylar tibia valgization osteotomy*. c Healed result after 2 years



■ Fig. 3.278. Lateral x-ray of a 16-year old boy with *osteochondritis dissecans* of the patella

condyle, 16% on the lateral femoral condyle, 6% on the patella (■ Fig. 3.278) and 0.3% on the lateral tibial plateau (in the concave part).

### Diagnosis

The symptoms in osteochondritis dissecans are non-specific. Exercise-related pain is usually present, and possibly pseudolocking as well. Genuine locking and effusions are observed particularly in the presence of loose joint bodies. During clinical examination, the test described by Wilson [37] is helpful in establishing the diagnosis. With the leg slightly flexed, pain is elicited when forced external rotation is applied since the osteochondrotic focus is compressed by the anterior cruciate ligament. The above-mentioned multicenter showed that neither pain nor effusion were reliable indicators of a dissected fragment [17].

The diagnosis is primarily confirmed with *plain x-rays*: In addition to the AP and lateral views, we require a tunnel view according to Frick (▶ Chapter 3.3.2). This view is used for visualizing the intercondylar notch and usually shows the osteochondrotic focus better than the AP view, since this is generally located on a relatively posterior part of the lateral margin of the medial femoral condyle. A typical feature of osteochondritis dissecans is sclerosis around the necrotic zone. The disease should



■ Fig. 3.279a, b. 7-year old boy with *ossification disorders* of the lateral femoral condyle (x-rays: a lateral, and b AP). This is a harmless variant of normal ossification of the epiphysis and not a case of *osteochondritis dissecans*

not be confused with ossification irregularities, which are common particularly in smaller children and are of no clinical significance (■ Fig. 3.279). The  $^{99}\text{Tc}$  *bone scan* was used in the past for monitoring progression, but has superseded by the MRI scan with its wide range of possibilities.

The *MRI investigation* always shows the osteochondrotic focus very clearly. While the intact appearance of the cartilage can also be seen on the MRI scan, this evaluation is not always reliable in view of the inadequate resolution. The scan shows an enlarged focus as a result of the surrounding edema. The most important question to be answered by the MRI concerns the stability of the lesion. This can be answered most convincingly if fluid (effusion or gadolinium injected into the joint) is seen to flow around the focus – in this case the dissected fragment is obvious. Accordingly, the most reliable information is provided by an arthro-MRI. But even intravenously injected gadolinium is helpful, since it reveals the circulation in and around the focus [28].

The following simple classification has been proposed by Bohndorf [5]:

- Stage I: intact cartilage, contrast enhancement of the lesion, absence of »cystic« changes.
- Stage II: discontinuity in the cartilage surface. Fluid around the fragment / dissected fragment.

### 3.3.4 · Osteochondritis dissecans

This classification provides clear indications for the subsequent therapeutic strategy.

The cartilage surface can best be assessed by *diagnostic arthroscopy*. An initial sign of incipient dissociation is the yellowish discoloration of the cartilage. The initial detachment of the cartilage-bone fragment can very readily be palpated with a probe.

#### **Prognosis, indication for type of treatment**

Several studies have shown a substantial risk of arthritis after osteochondritis dissecans. One study with a follow-up period of more than 33 years showed that arthritis was clearly present in 32% of the patients and that only 50% of the patients were without symptoms [36]. Other investigations have reported an arthritis risk of a similar level [8]. The prognosis appears to correlate closely with the age of the patient at the onset of the condition [33]. If the first symptoms occur more than a year before closure of the epiphyseal plate (juvenile form), the course is much better compared to a later onset of the disease. Various studies have attempted to compare conservative and surgical treatments [9]. Although most of these investigations have not shown any advantages of surgery over conservative treatment, the significance of such studies is questionable given the poor comparability of the patient populations.

In the above-mentioned *multicenter study* of the European Paediatric Orthopaedic Society directed by the author [17] and involving 798 cases of osteochondritis dissecans, suitably comprehensive documentation with an adequate follow-up period was available in 509 cases. The evaluation of these cases enabled the *following conclusions* to be drawn:

- The *prognosis is age-related*: It is better in children and adolescents than in patients with premature or closed plates. Nevertheless, even in the younger patient group, an abnormal knee was observed in 22% of cases after an average follow-up period of 3.2 years (compared to 42% in the older patients), indicating that the disease is not completely benign in the young. A poor prognosis exists particularly if fragment dissection occurs. In a patient with pain and/or effusion, the dissection can only be detected with sufficient reliability by MRI or arthroscopy. Conventional radiography is not sufficiently reliable.
- Osteochondrotic focuses at the *classical site* have a *better prognosis* than those at an atypical site.
- The *prognosis is worse in patients who actively participate in sports* than in less active patients.
- Provided *no dissection* has occurred, the *results of conservative treatment are better* than those observed after surgery.
- Simply *drilling the focus* – if no dissection has occurred – *does not have any positive effect on the end result*.

- If *dissection* has occurred, the *results of surgical treatment are better* than those of conservative treatment.

Accordingly, the crucial question concerning the indication for treatment is whether the lesion is stable. A reliable answer can usually be provided by MRI [13].

#### **Treatment**

##### **Conservative treatment**

The following options are available:

- Reduction of sporting activity,
- Relief,
- Physical therapy,
- Cast immobilization, splints.

The most important measure is probably the *reduction of sporting activity* or a *sports ban*. The risk that the focus does not heal spontaneously is much greater if sporting activity is continued than if it is suspended for a certain period. Other conservative methods, for example a specific *exercise program* and *relief*, may be useful in the short term [9]. The objective of temporary *immobilization* in a *cylinder cast* is not so much to facilitate the reintegration of the fragment as to effectively impose the sports ban. However, the cast should not be applied for longer than 4–6 weeks, as a more prolonged period of immobilization is harmful for the joint generally. Since the (radiological) healing usually takes a lot longer than 4–6 weeks, the use of removable *splints* is useful. A splint in a near fully-extended position worn throughout the day avoids rotational movements, relieves the – usually dorsally located – osteochondrotic focus and prevents sporting activity.

##### **Surgical treatment**

The following procedures are possible:

- Drilling of the sclerotic zone (forage, Pridie drilling),
- Refixation of the dissected fragment,
- Reconstruction after defect formation.

##### **Drilling of the sclerotic zone (Pridie drilling)**

A sclerotic zone forms around the necrotic, dissociating focus. This is probably a reaction to, rather than the cause of, the circulatory impairment. Drilling (or forage) of the sclerotic zone is designed to improve the circulation to the dissected fragment. The drilling can proceed in a *retrograde* i.e. from outside the joint surface in the direction of the dissected fragment, or *anterograde* direction, i.e. through the cartilage covering and right through the avascular zone. As noted in our multicenter study, we were unable to demonstrate the benefit of the very common [19] procedure of drilling [17], despite the positive results reported in the literature [1, 6]. If no dissection has occurred, this surgical measure is not useful on its own.

### Refixation

The following refixation methods are available:

- normal small fragment screws,
- cannulated screws (e.g. »Herbert screw«) [12],
- special screws made of allogeneic bone,
- special hooks [18],
- fibrin glue,
- absorbable pins or screws made of polyglycolic acid or other polylactates.

The key requirement is that the fixation should be stable. This is possible only if the dissected fragment consists exclusively of cartilage and the bony surface is not covered with a thick layer of connective tissue. Only fragments that have not yet completely broken off, or that have left their bed within the recent past can be refixed. The type of refixation is of secondary importance.

We ourselves have experience with all of the above-mentioned methods. The earlier types of absorbable pins were not suitable, in our view, since they did not produce any compression between the fragment and the femoral condyle. Nor has our experience with hooks been positive, since they are relatively complicated to assemble and likewise do not produce any effective compression. Furthermore, they show a certain tendency to loosen and their removal is relatively time-consuming. A recent technique involves the use of screws made of allogeneic bone that do not need to be removed. On the other hand, these screws retain their hardness, which is substantially higher than that of the surrounding cartilage and bone, for a long time, which can lead to problems [14]. Fixation with a single screw continues to remain the best solution (■ Fig. 3.277b). Screws made of polyglycolic acid do not need to be removed and produce good compression between the dissected fragment and the mouse bed. »Smart pins« that produce good compression have also recently been developed.

### Reconstruction after defect formation

Extensive research is currently being conducted in this field, and a variety of new methods have been proposed in recent years. The following procedures are discussed and also employed in some cases:

- allogeneic cartilage/bone graft (allograft),
- autologous cartilage/bone graft in the form of ground tissue, with or without fibrin glue,
- autologous cartilage graft combined with periosteum or perichondrium (periosteal flap reconstruction),
- autologous cartilage/bone graft (mosaicplasty),
- cartilage replaced by cultured cartilage tissue.

None of these methods has gained widespread acceptance. All have their own disadvantages and none represents a fully adequate replacement of the defective part of

the joint. We have acquired (limited) personal experience with most of these methods.

*Allogeneic cartilage/bone grafts (allografts)* can be harvested from cadavers. Graft availability, exact matching and the fixation method can prove problematic, although the reports in the literature are very positive [3, 11, 15]. Our own experience from tumor surgery (► Chapters 3.3.12 and 4.5.5) suggests that problems occur at an early stage if the radii of curvature of the adjacent surfaces do not match exactly.

*Autologous cartilage/bone grafts* must be taken from other sites in the knee, whether from the tibia [35] or parts of the femoral condyle far away from the dissection [38]. This is based on the idea that there are important and less important loading zones in the knee. For a long time it was claimed that the classical site of osteochondritis dissecans was not in the loading zone. We consider this view to be incorrect.

**!** On the human body there is no site with hyaline cartilage that does not experience loading corresponding to the thickness of the cartilage.

This restricts the options for autologous cartilage grafting. For many years we have used a paste made from crumbled chondrocytes (taken from the region of the intercondylar notch), autologous cancellous bone and fibrin glue. Postoperatively, we have consistently used the passive motorized splint. One problem with this procedure is ossification of the graft up to the joint surface.

This problem also occurs after *periosteal flap reconstruction*. While the transplantation of periosteum or perichondrium (from the ribs) into the defect offers the potential of a repair tissue that is probably equivalent to hyaline cartilage [10, 29], reports have also recently described the failure of this method [23, 27]. Better conditions are produced if the periosteum is seeded with autologous, cultured chondrocytes [7, 32]. This costly technique looks promising, and considerable research effort is currently focusing on the field of cell cultures. The main problem lies in the anchoring of the chondrocytic layer on the carrier material (bone, periosteum, synthetic material).

There is considerable discussion nowadays about »mosaicplasty« [16]. In this procedure, cylinders of cartilage and bone are taken from the edge of the femoral condyles using a special device and inserted into the defect. The advantage of this method is that the replacement graft of full-thickness hyaline cartilage is well anchored in the underlying bone. The follow-up studies conducted to date (histological and MRI studies) have shown good integration of the bone, although the cartilage only appears to bind with the adjacent connective tissue. No investigations have been conducted to date by independent authors, i.e. who are not promoters of the method.



### 3.3.4 · Osteochondritis dissecans

In particular, no clear solution has been proposed for the problems that can occur in the long term at the harvesting site – *for, as mentioned above, on the human body there is no site with hyaline cartilage...*

We are therefore rather skeptical about this method, since it involves the risk of further damage to the knee.

#### Correction of the leg axes

If a pronounced valgus or varus deformity is present, a correction osteotomy can be particularly useful if osteochondritis dissecans is present in the overloaded zone (■ Fig. 3.277b) [34]. The osteotomy must be performed at the site of the deformity, i.e. either at the distal femur or proximal tibia.

#### Our therapeutic strategy for osteochondritis dissecans

Our therapeutic strategy for osteochondritis dissecans is shown in ■ Table 3.46.

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■ Table 3.46. Our therapeutic strategy for osteochondritis dissecans

Signs and symptoms	Treatment
Asymptomatic	No treatment
Mild symptoms, no effusion, no suspicion of dissection on the x-ray	Sports ban, poss. temporary »hindering« splint or cylinder cast. No »forage« of the focus
Clear clinical symptoms and/or radiological signs of dissection	MRI or arthroscopy, if dissection confirmed, refixation with (polyglycolic acid) screw(s) or fragment removal and defect filling. Poss. axial correction if varus axis present



32. Peterson L, Minas T, Brittberg M, Nilsson A, Sjogren-Jansson E, Lindahl A (2000) Two- to 9-year outcome after autologous chondrocyte transplantation of the knee. *Clin Orthop* 374: 212–34
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### 3.3.5 Dislocation of the patella

#### ➤ Definition

Dislocation of the patella from the patellofemoral groove in a lateral direction.

#### Classification

We distinguish between the following:

- *Acute traumatic dislocation of the patella*: Lateral dislocation of the patella caused by proportionate trauma.
- *Acute constitutional dislocation of the patella*: First dislocation of the patella in the presence of predisposing factors. Usually progresses to the recurrent form.
- *Recurrent dislocation of the patella*: Repeatedly occurring lateral dislocations of the patella in the presence of predisposing factors.



- *Habitual dislocation of the patella*: The patella can be dislocated laterally at will and only remains in the patellofemoral groove when the leg is extended.
- *Chronic dislocation of the patella*: The patella dislocates during the course of childhood and is then permanently dislocated, the extensor mechanism is displaced laterally (usually combined with genu valgum).
- *Congenital dislocation of the patella*: The patella is dislocated at birth and the extensor mechanism is displaced laterally (usually combined with genu valgum).
- *Neuromuscular dislocation of the patella*: Lateral (usually permanent) dislocation of the patella caused by abnormal traction on the vastus lateralis muscle.
- *Iatrogenic dislocation of the patella*: Medial subluxation or dislocation of the patella after surgery to correct a lateral dislocation of the patella.

#### Etiology

As may be seen from the classification, the etiology of dislocation of the patella is not uniform.

#### Acute traumatic dislocation of the patella

A proportionate level of trauma is needed to produce an acute dislocation of the patella. The injury usually occurs when the knee is in a position of flexion, valgus and external rotation. This is the same mechanism that most commonly leads to rupture of the anterior cruciate ligament in adults. Although injuries of this ligament are fairly rare in children and adolescents, this connection must be borne in mind. Other concomitant injuries are also often observed, for example disruption of the medial retinacula and shear fractures of the medial patellar facet and lateral femoral condyle. Acute traumatic dislocation of the patella without any predisposing factors is rare in children and adolescents.

#### Acute constitutional dislocation of the patella

This injury is observed much more frequently in children and adolescents than acute traumatic dislocation. In contrast with the latter, the trauma of the triggering accident is not proportionate, few concomitant injuries are observed and predisposing factors are present (these are described in detail for the recurrent form). The acute predispositional form almost always progresses to a recurrent form.

#### Recurrent dislocations of the patella

In this common condition, recurrent lateral dislocations of the patella occur with increasing frequency. The dislocations are promoted by the following predisposing factors:

- *General ligamentous laxity*: Most patients show signs of a general weakness of the ligaments. Typically, recurrent dislocations of the patella are also common in hereditary disorders associated with a diminished

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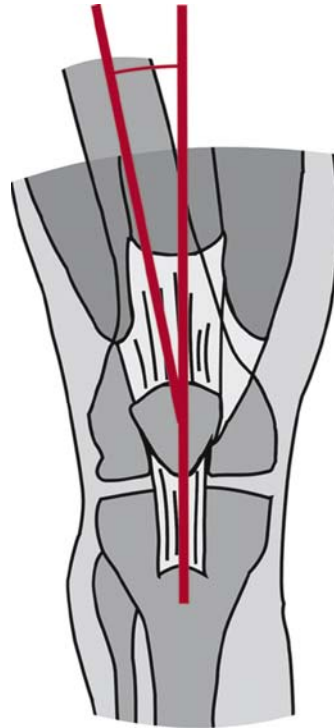
quality of collagen (e.g. Ehlers-Danlos syndrome, arachnodactyly or Marfan syndrome, osteogenesis imperfecta, Turner syndrome [17], Down syndrome, Kabuki syndrome) [7] (see also ► chapter 4.6).

- *Muscle imbalance with subluxation of the patella:* If, when the leg is extended without tensing of the quadriceps, the patella slides laterally in the patellofemoral groove, then dislocation of the patella is more likely to occur during external rotation of the lower leg and increasing flexion. Individual authors have also found primary muscle changes. A fairly recent MRI-based study has shown the regular presence of fibrosis of the vastus lateralis muscle in patients with habitual dislocation of the patella [12].
- *Dysplasia of the femoral condyles:* The lateral femoral condyle normally protrudes anteriorly slightly more than the medial condyle. Flattening of the lateral femoral condyle and a reduced indentation of the patellofemoral groove will promote dislocation.
- *Changes in the retinacula:* The retaining apparatus is very important for guiding the patella. Tearing of the medial ligaments and shortening of the lateral ligamentous apparatus will promote dislocation.
- *Patella alta:* If the patient happens to have a high patella, the patella will show a delay in sliding into the patellofemoral groove during increasing flexion and thus make a dislocation more likely.
- *Q-angle:* The determination of this angle is shown in ■ Fig. 3.280. A high Q-angle is thought to increase the probability of dislocation. However, CT measurements have shown that this angle was not increased, but rather reduced, in a fairly large sample of patients with patellar dislocation [21, 24]. This was partly due to the fact that the patella in these patients was in a more lateral position than normal [24].
- *Axial and rotational deformities:* A genu valgum or increased lateral torsion of the tibia will promote dislocation of the patella. The same applies to a recurvated knee. Additionally, increased rotation between the femur and tibia is a typical feature of patients with recurrent dislocation of the patella [21].

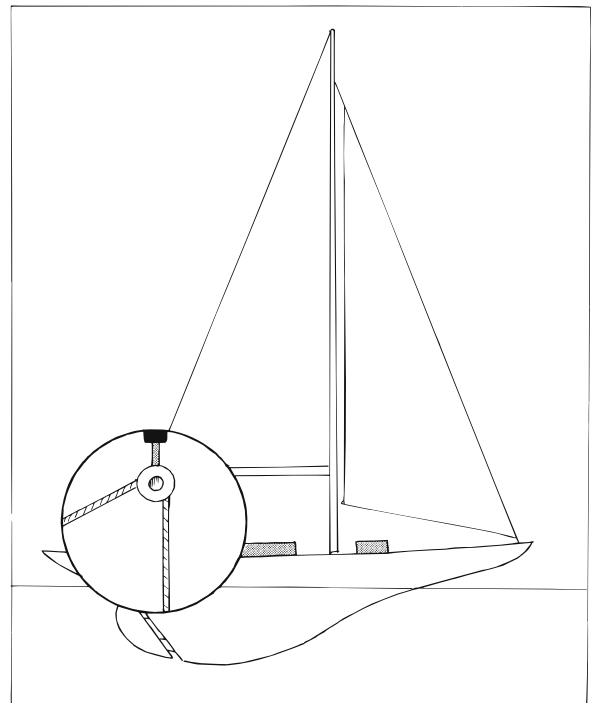
In most cases it is not one individual factor that leads to the establishment of recurrent dislocation of the patella, but rather a combination of various elements.

#### Habitual dislocation of the patella

In this form of the condition the patient can dislocate the patella at will by exerting lateral traction on the vastus lateralis muscle. The kneecap slides to the side during increasing flexion and then reduces itself again on extension. The predisposing factors are even more pronounced compared to the recurrent form. Nor is it possible to make a clear distinction between habitual and congenital dislocation.



■ Fig. 3.280. Determination of the Q-angle: The angle between the axes of the quadriceps and patellar tendons. An angle of more than  $15^\circ$  is considered to be pathological, although the measurement is very inaccurate



The patella is like a pulley. Without it the sail cannot be hoisted or the knee properly extended..

### Chronic dislocation of the patella

This dislocation occurs during the course of childhood. The patella is permanently dislocated and is often (particularly after early dislocation) smaller than normal. The permanent dislocation is usually preceded by a period of recurrent or habitual dislocation. The quadriceps muscle is secondarily too short and displaced laterally, in some cases functioning as a flexor. Typically, chronic dislocation occurs in patients with pronounced ligament laxity, e.g. in Down syndrome.

### Congenital dislocation of the patella

This dislocation is already present at birth. The patella is very small and dysplastic, and in some cases is even completely absent. The quadriceps muscle is too short and displaced laterally, in some cases functioning as a flexor [2]. Congenital dislocation of the kneecap is rare and occasionally occurs in connection with the »nail-patella syndrome« or arthrogyposis.

### Neuromuscular dislocation of the patella

Recurrent or even chronic lateral dislocation of the patella not infrequently occurs in spastic tetraparesis. In addition to predisposing factors (see recurrent form), the abnormal muscle forces play an important role (► Chapter 3.3.7).

### Iatrogenic dislocation of the patella

Medial subluxation or dislocation can occasionally occur after poorly indicated and/or inadequately performed patella-centering operations [16].

### Occurrence

A Finnish epidemiological study calculated an incidence of 43 patellar dislocations per 100,000 children and adolescents under 16 years [19]. In patients with hemarthrosis, arthroscopy revealed a previous episode of patellar dislocation in 10% of cases [22].

### Clinical features, diagnosis

Dislocations of the patella usually occur in adolescents. Except in cases of acute traumatic dislocation (which is

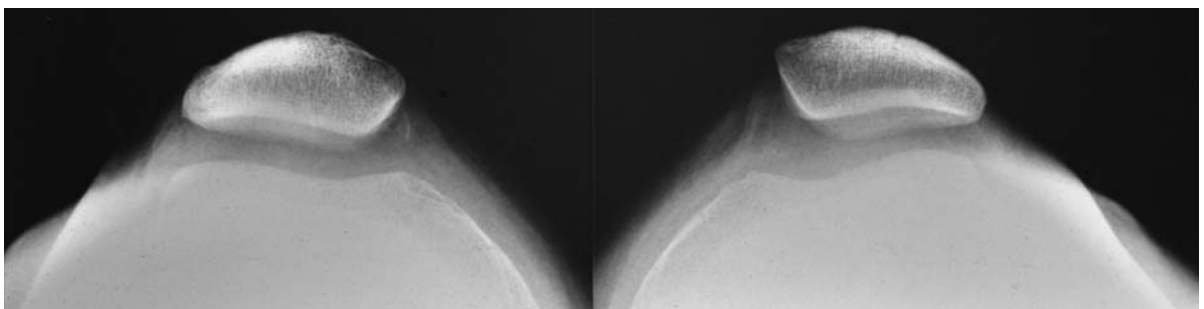
very rare), a precise history of the accident cannot usually be reconstructed. The incident is often described as »giving way«, »locking up«, and occasionally as »going out«. The dislocation occurs during flexion under load with external rotation of the tibia. Occasionally, the patients notice the patella dislocating and then reducing again after stretching the knee. Hemarthrosis frequently occurs not only after acute traumatic dislocation, but also after the first constitutional dislocation. Concomitant injuries must accordingly be ruled out (► Chapter 3.3.8). Patients with recurrent dislocations report this event occurring with increasing frequency. Effusions only form in rare cases. Chronic peripatellar symptoms with pain on prolonged sitting and walking downhill are often observed. Boys and girls are affected with approximately equal frequency [18].

The *clinical examination* should start with an evaluation of the leg axes while standing. Predisposing factors are valgus knees and recurvated knees and increased lateral torsion of the tibia. Atrophy of the vastus medialis muscle as an expression of a functional deficit is frequently observed. The painful lateral subluxation can be reproduced by pushing the patella laterally (apprehension sign«). Typically, the patient moves his hands as if to try and prevent the examiner from continuing with this manipulation [18]. Hypermobility of the patella can be checked at approx. 30° flexion. The examiner establishes whether, in this position, the kneecap can be moved over the lateral femoral condyle. Typically there is tenderness over the medial femoral condyle, which is also characteristic of other painful peripatellar conditions (► Chapter 3.3.3.2).

The axial and rotational situation should be noted: Genu valgum and a lateralized tibial tuberosity caused by rotation of the knee promote dislocation. Contrary to the prevailing view, the Q-angle (■ Fig. 3.280) is not usually increased, but rather decreased as the patella is lateralized [21, 24].

### Radiographic diagnosis

AP and lateral *x-rays*, as well as tangential views of the patella (with the least possible flexion; ■ Fig. 3.281, see also



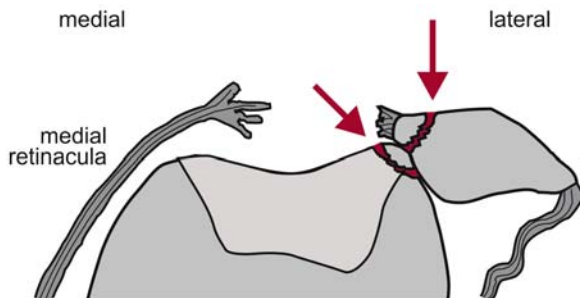
■ Fig. 3.281. Axial *x-ray* of both patellas of a 16-year old girl in 30° flexion of the knees. The femoral condyles are slightly dysplastic, while the right patella is slightly subluxated laterally

### 3.3.5 · Dislocation of the patella

► chapter 3.3.1) are recorded. The orthopaedist first checks all the x-rays for the presence of osteochondral fragments. Typical sites are the medial patellar facet and the edge of the lateral femoral condyle (■ Fig. 3.282 and 3.283). On the lateral view we measure the height of the kneecap according to Insall [8] (■ Fig. 3.284). On the tangential view of the patella, we can determine the lateral patellofemoral angle according to Laurin [13] (also known as the tilt angle) to establish any subluxation (■ Fig. 3.285).

In one major epidemiological study this tilt angle was an average of  $12^\circ$  in symptomatic patients, compared to just  $4^\circ$  in asymptomatic patients [4]. An even better indication of patellar subluxation is provided by CT or MRI scans with the leg extended, with and without tensing of the quadriceps. Muscle tension is visualized by this investigation (■ Fig. 3.286), which is particularly useful if the tangential x-rays fail to provide any clear findings. The dislocation movement generally takes place during the first  $20^\circ$  of flexion [11].

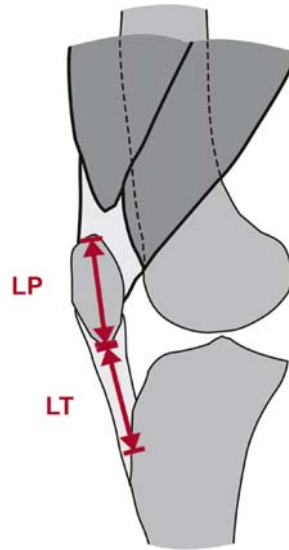
The evaluation of leg axes and leg rotation is also important. If the clinical findings are indicative of a pathology, we measure the axial, linear and rotational parameters on the CT scans, although these have the drawback



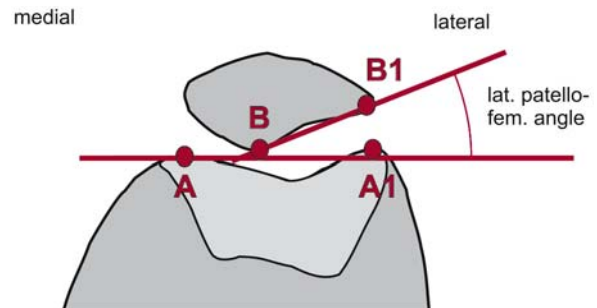
■ Fig. 3.282. Typical injuries associated with patellar dislocation: Tearing of the medial retinaculum, osteochondral fractures of the medial patellar facet and the lateral femoral condyle



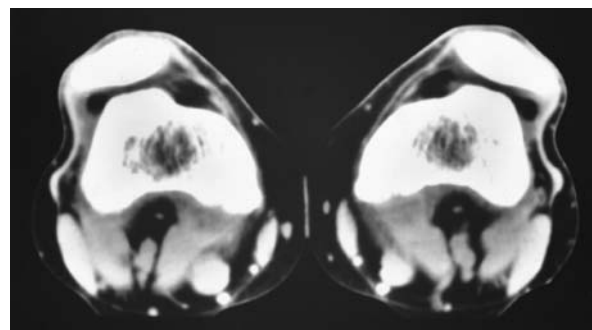
■ Fig. 3.283. Intraoperative photograph of a large osteochondral fragment on the back of the patella after traumatic dislocation in a 14-year old girl



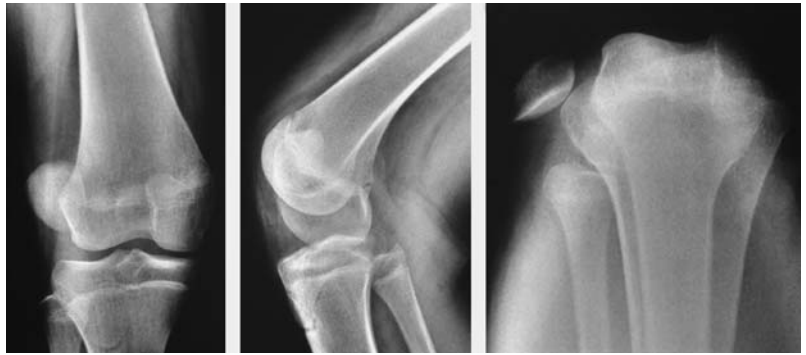
■ Fig. 3.284. Measurement of the height of a patella alta according to Insall & Salvati [8] (LP maximum length of the patella, LT maximum length of the patellar tendon). The normal length ratio is 1.0. A deviation of more than 20% indicates that a pathology is present, whether in the form of a patella alta (position of kneecap too high) or a patella baja (position of kneecap too low)



■ Fig. 3.285. Measurement of subluxation according to Laurin [13]: The line A–A1 links the two highest points of the medial and lateral femoral condyles on the axial x-ray of the patella in  $30^\circ$  flexion. The line B–B1 links both ends of the lateral patellar facet. The angle between the two lines corresponds to the lateral patellofemoral angle according to Laurin



■ Fig. 3.286. CT scan of the knees in extension in a 15-year old boy with subluxation of both patellas



■ Fig. 3.287. AP, lateral and axial x-rays of the right knee with *permanent dislocation of the patella* in a 15-year old girl with Down syndrome



■ Fig. 3.288. Photograph of both knees of an 11-year old girl with *habitual dislocation of the patella*. With the knee in flexion, the kneecaps on both sides are located lateral to the femoral condyle. The left knee has undergone a failed (soft tissue) recentering operation

of being unloaded views and therefore of significance only for the bone-related axes (► Chapter 4.2.2). ■ Fig. 3.287 shows a patella in a position of permanent dislocation, while ■ Fig. 3.288 shows the clinical picture of bilateral dislocation.

### Treatment

A first dislocation of the patella should always be treated conservatively while no major concomitant injuries are present. If any are detected the subsequent course of action is determined by these additional injuries. If hemarthrosis is present, an arthroscopy can occasionally be useful for evaluating the concomitant injury and joint lavage.

### Conservative treatment

The conservative treatment consists of fixation of the knee in extension with a removable splint. Isometric quadriceps exercises must be continued for 6 weeks. These are followed by isokinetic exercises, which can be supported by breaststroke swimming. With consistent conservative treatment, recurrence of the dislocation of the patella can be prevented in the majority of cases [18]. After the first dislocation (provided no osteochondral fracture is pres-

ent), a recurrence rate of just 5% can be expected after conservative treatment [18].

If dislocation occurs more than once in children who are not yet full-grown, we recommend consistent *taping*. The parents and the child are instructed by a physical therapist on how to affix the special adhesive tape in order to pull the patella in a medial direction. If this measure is employed consistently, i.e. on a daily basis, further dislocations can be avoided, while medialization of the patella can promote the formation of a largely normal groove between the femoral condyles, a crucially important factor for the subsequent stability of the patella.

### Surgical treatment

Surgical intervention is indicated in:

- clearly *traumatic dislocations*;
- *recurrent dislocations*, if predisposing factors have been identified, taping has proved unsuccessful and general ligament laxity is not the only factor present;
- *congenital and habitual dislocation*, if there is a chance that a reasonably normal anatomical configuration can be restored;
- *neuromuscular dislocation*, if a corresponding level of suffering is present.

### Osteochondral fragments

These can only be refixed if a sufficient amount of bone is present. If this is not the case, they must be removed since they can otherwise cause further damage in the knee as loose joint bodies (see ► chapter 3.3.4 for methods of refixation and reconstruction).

### Procedures on the retinaculum

The lateral retinacular release can be performed as an arthroscopic or open procedure. In a primary dislocation the retinaculum can be sutured on the medial side. In the Kroggius operation a strip of connective tissue is removed from the medial side and inserted on the lateral side [10]. Procedures on the connective tissue retaining apparatus have a poor success rate. If the patella is poorly guided in respect of the muscle and bone configuration (depth



### 3.3.5 · Dislocation of the patella

of sulcus, axial and rotational positions), such soft tissue interventions are inadequate.

The prevailing forces shape the retaining apparatus, while the general ligament laxity, which is also usually present, ensures that the connective tissue no longer provides any further resistance to the dislocation process. Consequently surgical treatments for recurrent dislocations of the patella involving the retinacula alone are associated with a high rate of recurrence [18]. A particular problem is posed, in our view, by procedures involving concurrent surgery to both the medial and lateral ligamentous apparatus, since the circulation to the patella is impaired (particularly after several repeat operations), with a consequent risk of dystrophy.

#### Corrections involving the distal extensor mechanism

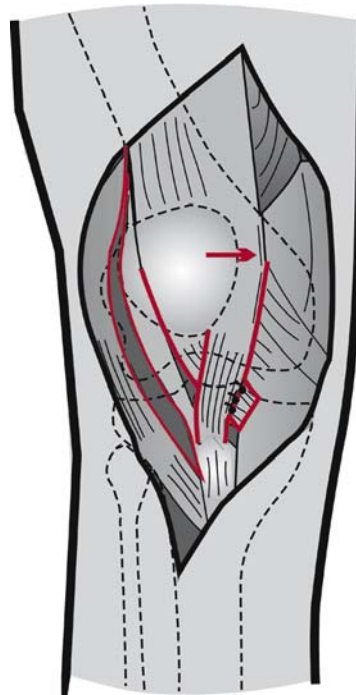
The following procedures can be performed on the distal extensor mechanism:

- *Soft tissue procedures* (medialization of half of the patellar tendon according to Goldthwait [3]),
- *Medialization* of the tibial tuberosity according to Roux [20] and Hauser [5] or Elmslie,
- *Distalization* of the tibial tuberosity according to Roux [20],
- *Ventralization* of the tibial tuberosity according to Maquet [15].

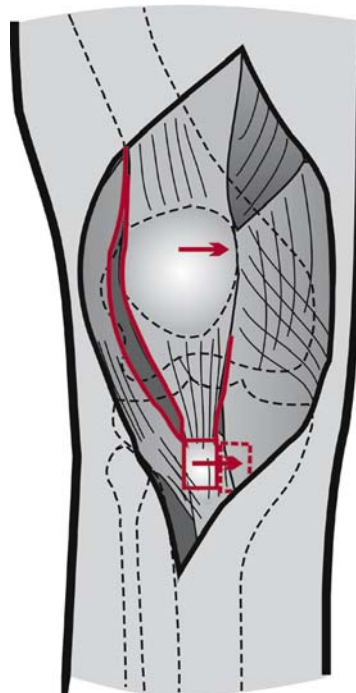
In the *Goldthwait operation* [3], the patellar tendon is split lengthwise and the lateral half is pulled over the medial half and fixed on the medial side (■ Fig. 3.289). This operation medializes the tension exerted by the tendon. The advantage of this method is that it can be performed even if the apophyseal plate has not yet closed. The disadvantage is a much higher recurrence rate compared to bone-based transpositions.

In the procedure of *medialization of the tibial tuberosity* according to Roux [20] and Hauser [5], a bone fragment with the complete patellar tendon attachment is chiseled out of the tibia, relocated laterally and fixed with one or more screws (■ Fig. 3.290). The attachment can also be distalized at the same time. This is particularly indicated for patients with a patella alta (the height of the patella is known to be a common predisposing factor). A disadvantage of distalization of the tendon attachment is a possible increase in pressure in the patellofemoral groove. To avoid this drawback, Maquet proposed the *ventralization of the attachment* [15]. However, we would explicitly discourage this measure since the anterior transfer of the tibial tuberosity frequently produces severe symptoms at this protruding site.

Other *complications* of the transposition of the tuberosity include recurrent lateral dislocations, overcorrections with medial dislocations [16] and a low patella (patella baja) as a result of scarring of the patellar tendon [18]. Consequently, the surgeon must be very cautious in



■ Fig. 3.289. Schematic view of *Goldthwait operation*: The patellar tendon is split lengthwise and the lateral half is pulled over the medial half and fixed on the medial side, while the lateral retinacula are split



■ Fig. 3.290. Schematic view of the *Roux-Hauser operation*: The distal attachment of the patellar ligament is chiseled out with a block of bone and fixed with a screw in a more medial (and possibly distal) position. The lateral retinacula are split

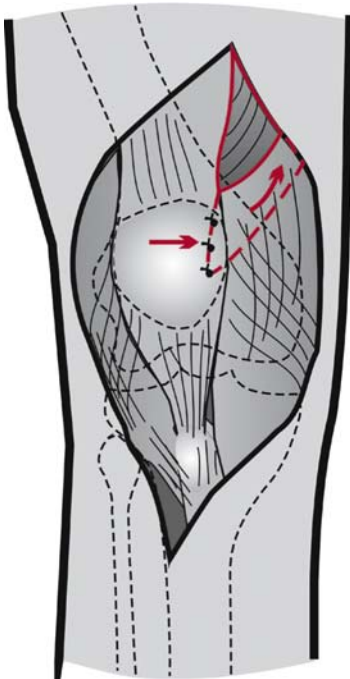
deciding whether such operations are indicated and, if so, implementing the appropriate procedure. Unless a pronounced patella alta is present, the operation should be limited to (graduated) medialization. Ideally, the Elmslie method should be used in which only the proximal part of the tuberosity is transposed, while the distal part is left in place.

### Corrections involving the proximal extensor mechanism

The following procedures can be performed on the proximal extensor mechanism:

- Insall procedure for distalization of the vastus medialis attachment on the patella [9],
- Stanislavjevic procedure for transposition of the complete quadriceps muscle medially [23].

The *Insall procedure for distalization of the vastus medialis attachment on the patella* [9] is indicated if the insertion point for the tendon is too high. This situation is usually linked with a patella alta. In this operation the tendon of the vastus medialis muscle is detached from the patella and anchored to the bone at a more distal point (■ Fig. 3.291). Postoperatively, the extended leg is immobilized in a removable splint for 4 weeks. In order to allow the newly inserted tendon to integrate with the bone, flexion must not exceed 30° during this period. In our experience, this operation, if appropriately indicated, is successful and associated with only low recurrence and complication rates.



■ Fig. 3.291. Principle of the *Insall operation*: The (excessively proximal) vastus medialis muscle is reanchored to the patella in a more distal and lateral position. This procedure causes the muscle to pull

In the *Stanislavjevic operation* [23], the complete quadriceps muscle is detached from the femur and transposed medially (■ Fig. 3.292). This operation may be indicated for *congenital and habitual* dislocations, and occasionally also for neuromuscular dislocations of the patella. We recently followed up 7 patients with 9 congenital dislocations of the patella after an average of 6.7 years [6]. Of the 9 knees, only one was unstable. Before surgery, an extension deficit of 20–50° had been present in all knees. At the latest follow-up, 6 knees could be fully extended, while the other 3 showed a deficit of 10–20°. If indicated, therefore, quadriceps transfer is a useful procedure.

### Correction of the patellofemoral groove

Two options for correcting the patellofemoral groove were recently proposed:

- *Elevation of the lateral femoral condyle by osteotomy* according to W. Müller
- *Deepening of the intercondylar sulcus (trochleaplasty)* by retrograde reaming of the patellofemoral groove.

Although the clinical value of these two operations cannot yet be assessed with any great accuracy, we have had good experiences with trochleaplasty for dysplastic forms. The subchondral bone has to be resected carefully, leaving an intact layer of bone underneath the cartilage. It is a technically difficult but rewarding operation.

### Correction of a rotational defect on the femur and tibia

A major rotational defect of the femur (increased anteversion) or tibia (increased external rotation) can represent an important (co-)factor in the dislocation process. Careful clarification with CT is important (► Chapter 4.2). If a relevant rotational defect is present, the correction should be performed as close to the knee as possible. This procedure may be performed on the tibia only after growth is concluded.

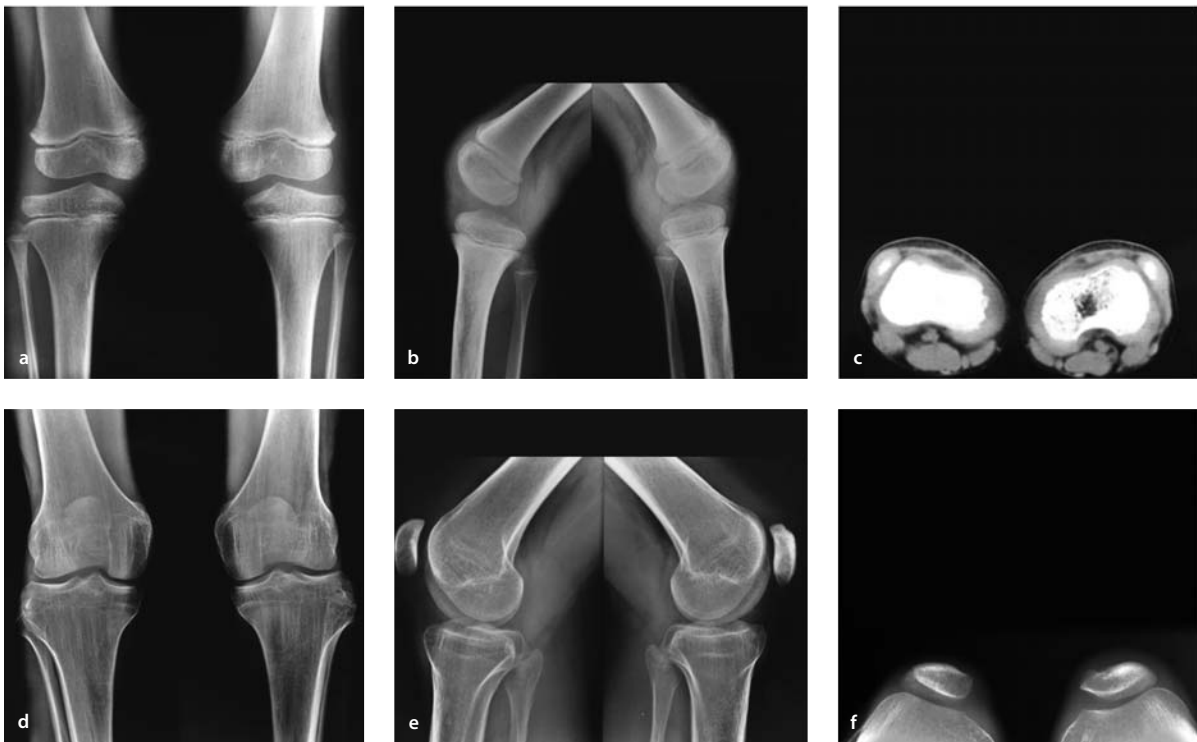
### Evaluation of the indications for the various operations

- ❗ **The surgical treatment of the various forms of dislocation of the patella is difficult. The surgeon must be very cautious in deciding whether an operation is indicated, and the appropriate procedure must be selected after a discriminating analysis of all the factors.**

All operations are associated with recurrence rates of varying degree [18], and the patient and his parents must be informed of these. The surgeon should beware of proceeding with excessively complicated multiple operations after the disappointment of a recurrence.

- ❗ **The circulation in the patella is a critical parameter, and the division of vessels and scar formation on several sides of the patella are only tolerated to a very limited extent.**

## 3.3.5 · Dislocation of the patella



■ Fig. 3.292a–f. 8-year old boy with congenital bilateral patellar dislocation. a–c AP and lateral x-rays and CT scan of both patellas. d–f X-

rays of both knees 15 years after *Stanisavljevic operation*. Both patellas are well centered and the patient is symptom-free

Such multiple operations can result in a protracted ordeal that may ultimately end in a patellectomy, and even this does not mark the end of the patient's suffering. Living without a patella is extremely problematic [14]. Some studies have even shown that the prognosis of patients undergoing surgery is worse than that for patients receiving conservative treatment [1]. Nevertheless, the value of these operations should probably not be viewed so negatively provided all the relevant factors have been taken into consideration, the prevailing pathology is corrected in a targeted manner and surgery is only performed for those cases in which targeted correction really is possible.

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### 3.3.6 Congenital deformities of the knee and lower leg

#### Definition

All anomalies of the knee or lower leg, or combinations of the two, that are present at birth.

#### Classification

A distinction is made between localized disorders and deformities in systemic diseases.

#### Localized disorders

- Fibular deficiency (usually combined with a longitudinal problem of the femur and lateral ray aplasia on the foot)
- Tibial deficiency of the tibia (poss. combined with medial ray aplasia on the foot), incl. tibiofibular diastasis
- Congenital dislocation of the knee
- Congenital absence or hypoplasia of the cruciate ligaments (usually combined with a longitudinal malformation of the femur, fibula and foot)
- Congenital pseudarthrosis of the tibia (usually combined with tibial hypoplasia, often also occurring in connection with neurofibromatosis)
- Discoid meniscus
- Congenital band syndrome

#### Knee deformities in systemic diseases

- Arthrogryposis
- Larsen syndrome
- Multiple epiphyseal dysplasia
- Achondroplasia
- Kniest syndrome
- Dysplasia epiphysealis hemimelica
- etc.

### Localized disorders

#### 3.3.6.1 Fibular deficiency (Fibular hypoplasia / aplasia)

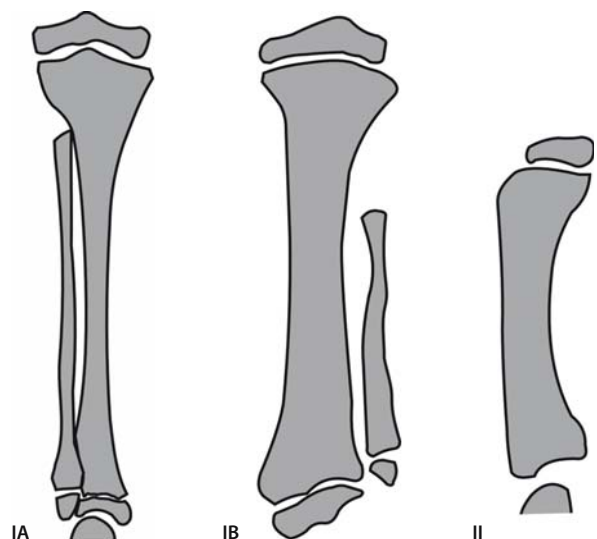
##### Definition

Hypoplasia or aplasia of the fibula can occur in isolation, but is usually accompanied by a malformation of the femur (► Chapter 3.2.7), lateral deformities on the foot and shortening of the whole lower leg of varying degree.

■ **Synonyms:** Congenital longitudinal deficiency of the fibula, congenital fibular hemimelia

##### Classification

The most convincing classification (since it determines the treatment) is that of Achterman and Kalamchi 1979 [1] (■ Fig. 3.293, ■ Table 3.47).



■ Fig. 3.293. Classification of fibular deficiency according to Achterman & Kalamchi [1]: Type IA: Hypoplasia of the fibula in the proximal area, ankle mortise reasonably intact; Type IB: Hypoplasia of the fibula with dysplastic or absent ankle mortise. Type II: Aplasia of the fibula

■ Table 3.47. Classification of fibular deficiencies. (According to Achterman & Kalamchi 1979 [1])

Type	Parameter
IA	Hypoplasia of the fibula in the proximal area, ankle mortise intact
IB	Hypoplasia of the fibula with dysplastic or absent ankle mortise
II	Complete absence of the fibula



### Occurrence

A recent epidemiological study in Germany (involving almost 600,000 neonates in the years 2000/2001) found fibular deficiencies in 0.3 per 10,000 neonates [47].

### Associated anomalies

The foot is almost always affected to a varying extent by the condition. At birth, an equinus, footdrop position is usually observed. One or more lateral rays of the foot are often absent, and the bony structures of the rearfoot may also be incompletely formed, or there may be coalition of the bones of rearfoot [21]. In 15% of cases, a clubfoot position is present [8]. In approximately two-thirds of cases, hypoplasia of the femur or a proximal femoral deficiency is also observed. In addition, the whole lower leg is hypoplastic. There is usually hypoplasia of the lateral femoral condyle and, in 18% of cases, aplasia of the cruciate ligaments as well [34].

### Clinical features, diagnosis

The affected lower leg shows visible shortening even at birth. Tibial anterior bowing is generally present and a varus deformity of the tibia is often observed. A dimple in the skin is apparent at the apex of the curvature (particularly in type II). The lateral malleolus cannot be palpated in aplasia of the fibula (type II) and is abnormally high in hypoplasia (type IB; ■ Fig. 3.294). The foot is usually in an equinovalgus position as a result of contracture of the calf and peroneal muscles. Sometimes the rearfoot is also dislocated laterally and may be at a higher level than the end of the tibia. The rearfoot, and occasionally the metatarsal bones as well, are frequently very rigid as a result of coalition of the talus and calcaneus.

### Treatment

The treatment of congenital anomalies of the fibula and lower leg is very complicated and requires considerable experience. The following therapeutic options are available:

- shoe elevation,
- orthoses,
- prostheses,
- realignment osteotomies,
- surgical leg lengthening,
- rotationplasty,
- amputation.

❗ **Any treatment of patients with an outwardly visible disability should be accompanied by good psychological management.**

The therapeutic strategy should be discussed with the parents at the earliest possible opportunity and subsequently established during the course of early childhood. The main problem to be resolved is whether preservation of the complete extremity and leg length equalization



■ Fig. 3.294. X-rays of the left lower leg of a 1 1/2-year old girl with a type IB fibular deficiency

should be attempted up until the completion of growth, or whether some other solution should be chosen. For very severe deformities, leaving the leg length unchanged with or without amputation of the forefoot with prosthetic management or a rotationplasty with a lower leg prosthesis is usually a better solution in functional respects than surgical leg lengthening. However, parents and patients often find this the more difficult option to accept in psychological respects [32, 33].

The number of existing toes and metatarsal bones in the foot gives a good indication of the prognosis and the severity of the deformity. If 3 or more rays are present, preservation of the limb with a lengthening procedure is recommended. If only 2 toe rays are present, the chances of producing an effectively functioning lower limb with the leg lengthening procedure are very small. In such cases, the parents and child should be carefully guided towards other options and helped to accept the disability.

The *therapeutic strategy* is based not just on the deformity, but also the age of the patient.

### Preschool age (up to 6 years)

Depending on the extent of the shortening in each case, a leg length equalization procedure followed by a shoe



sole wedge or lower leg orthosis is appropriate. If possible, the orthosis should place the foot in a plantigrade position.

### School age (6–10 years)

If the leg is shortened by more than 10 cm at this age, the decision must be taken whether to preserve the whole limb and attempt leg length equalization until the completion of growth, or whether some other solution should be chosen. In this case a shoe wedge will no longer be sufficient as an orthopaedic appliance, but a lower leg orthosis with a foot support and a separate orthotic foot section (■ Fig. 3.295). While this type of orthosis is less attractive cosmetically than if the foot is placed in the orthosis shaft in an equinus position, the plantigrade posture should be attempted if subsequent lengthening of the leg is being considered.

The *indication* for leg lengthening is based primarily on the type of the deformity and, to a lesser extent, on the degree of the leg length discrepancy (although both are interrelated):



■ Fig. 3.295. Lower leg support with separate foot section on a 12-year old patient with a fibular deficiency and leg shortening of 8 cm. The foot is in a plantigrade position

### Good indications for lower leg lengthening

- type I A [1] (hypoplasia of the proximal fibula, ankle mortise intact),
- preserved rays I, II and III on the foot,
- leg length discrepancy at 8 years  $\leq 10$  cm.

### Questionable indications for lower leg lengthening

- type I B [1] (hypoplasia of the fibula with dysplastic or absent ankle mortise),
- preserved rays I and II on the foot,
- leg length discrepancy at 8 years between 8 and 15 cm.

### Poor indications for lower leg lengthening

- type II [1] (fibular aplasia),
- only 1 foot ray preserved,
- leg length discrepancy at 8 years up to 15 cm.

Although lower leg length extensions of up to 25 cm are technically possible, the price for such long extensions is very high as the complication rate rises dramatically for extensions of 8 cm or more.

! We always equalize a lower leg length discrepancy of more than 8 cm in several steps, with a maximum of 8 cm (better: 6 cm) in each case.

Instability of the ankle mortise is not an absolute contraindication for leg lengthening. Ring fixators (of the Ilizarov type or the Taylor Spatial Frame) can be used to incorporate the foot in the extension and thus prevent dislocation of the ankle (see ► chapter 3.3.13 for an illustration of this type of assembly for incorporating the foot). Lower leg lengthening is discussed in greater detail in ► chapter 4.2, while the management of foot problems with the ring fixator are addressed in ► chapters 3.4.3 and 3.4.5.11.

### Procedure if lengthening is not performed

The foot can basically be fitted in the lower leg prosthesis in an equinus position. Cosmetically more satisfying prosthetic management is possible if the forefoot is amputated, although the children and parents find this very difficult to accept. Amputation also has the disadvantages of possible phantom pain and more difficult guiding of the prosthetic foot (shorter lever arm, loss of the important sensory function of the toes).

The decision to have a part of the body cut off is a psychologically painful process, even if the body part in question hinders the patient in functional or cosmetic respects and brings certain disadvantages. The children and their parents must be informed about this option very

carefully, and they must never be placed under pressure. Parents are usually prepared to do everything they can for their children, and some will not be deterred, despite all the warnings, from deciding to opt for an almost hopeless lengthening procedure that is not really indicated. The children will have to face years of an extremely painful treatment characterized by multiple complications and that ultimately ends in both legs being roughly the same length, although the function of the extended limb will be worse than if prosthetic management had been prescribed.

If femoral hypoplasia is also present (as is the case in the majority of patients; ► chapter 3.2.7.2), there is the added problem of the knees being at different heights. This further spoils the cosmetic appearance because the difference in the heights of the knees is clearly visible as soon as the difference exceeds 5 cm. A lower leg prosthesis worn beneath the clothing, on the other hand, is hardly noticed by others. In these cases the possible alternative of just lengthening the thigh rather than the lower leg should be considered. Here, too, the decision will depend on the deformity. Lengthening is hardly ever possible if simultaneously a proximal femoral deficiency is present. The condition of the hip and knee will also influence the decision. Knee problems (e.g. aplasia of the cruciate ligaments) are easier to overcome technically than hip problems, since the knee can be incorporated in the extension apparatus.

#### Amputation methods

Despite the understandable desire to amputate as little as possible, a warning should be given about amputation of the forefoot. The Lisfranc or Chopart amputation can cause numerous problems in prosthetic management. The stump mobility cannot be used functionally but rather leads to constant rubbing in the prosthesis shaft and to repeated pressure sores. Better amputations are the rear-foot amputations according to Boyd or Pirogoff, in which the talus, calcaneus and heel pad are preserved but fused together and with the lower leg, or the lower leg amputation according to Syme (► Chapter 3.4.5.11).

#### Adolescence (10–16 years)

If major discrepancies in length are still present at this age, a second and possibly third extension operation are performed. As mentioned above, the individual steps should not exceed 6–8 cm.

#### Instability of the ankle mortise

If the ankle mortise is very unstable (types IB and II), this can be eliminated by arthrodesis of the upper, and possibly also the lower, part of the ankle when growth is complete. Leg length is preserved in this procedure, and the foot should be fused in a plantigrade rather than an equinus position [23]. The loss of mobility is function-

ally acceptable, and the benefit of the stable foot position more than offsets this drawback.

#### 3.3.6.2 Tibial deficiency (including tibiofibular diastasis)

##### ► Definition

Hypoplasia or aplasia of the tibia, often combined with a varus deformity of the rearfoot and the absence of rays on the medial aspect of the foot, occasionally also with hypoplasia of the distal femur.

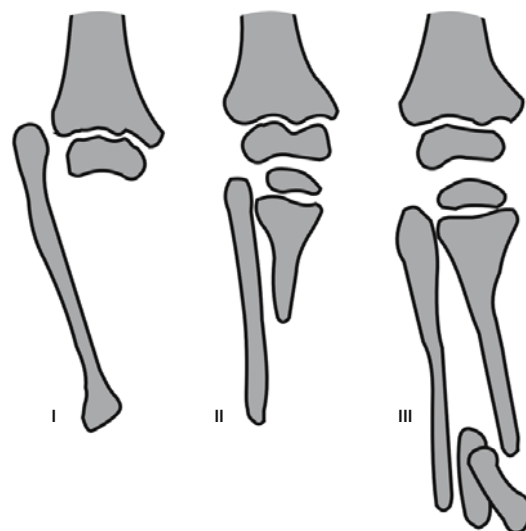
— **Synonyms:** Tibial hemimelia, longitudinal deformity of the tibia.

##### Classification

The best classification was proposed by Kalamchi and Dawe in 1985 [28] (► Fig. 3.296). The various types of deficiency are listed in ► Table 3.48.

► **Table 3.48.** Classification of tibial deficiencies. (According to Kalamchi & Dawe [28])

Type	Parameter
I	Absence of the tibia, foot in inversion and adduction, poss. absence of rays on the medial side of the foot
II	Hypoplasia of the tibia with absence of the distal half, femorotibial joint preserved (rarest type)
III	Dysplasia of the distal part of the tibia with diastasis of the tibiofibular syndesmosis, foot in varus position, prominence of the lateral malleolus (also known as <i>tibiofibular diastasis</i> )



► **Fig. 3.296.** Classification of *tibial deficiency* according to Kalamchi & Dawe. Type I: Complete absence of the tibia; Type II: Hypoplasia of the tibia with absence of the distal half; Type III: Dysplasia of the distal part of the tibia with diastasis of the tibiofibular syndesmosis

### Occurrence

The tibial deficiency is very much rarer than that of the fibula, with an incidence of 0.1 per 100,000 births [7, 47].

### Associated anomalies

The foot is normal in only around half of the patients, and ray defects and coalitions are usually present [15]. Two-thirds of children with longitudinal deficiency of the tibia show associated anomalies [28, 31], including syndactyly, polydactyly, femoral hypoplasia, cryptorchism, cardiac defects, varicocele, etc.

### Clinical features, diagnosis

The shortening and deformity of the lower leg is already clearly visible at birth. If the tibia is absent (type I), the lower leg is usually curved in a valgus position. Radiographic investigation reveals a hypoplastic distal femur but a thickened fibula. The foot is in a clubfoot position. In type II, involving the absence of the distal part of the tibia, the knee is generally normal, whereas the upper part of the ankle is unstable and the foot is inverted and

adducted. Type III (tibiofibular diastasis) is characterized by protrusion of the lateral malleolus with inversion and adduction of the foot (■ Fig. 3.297).

### Treatment

The treatment is based on the type of deformity present.

#### Type I (aplasia of the tibia)

The primary treatment is always *orthotic provision*. Quadriceps function and the condition of the distal femur are crucial to the success of this treatment. The most elegant and functionally best solution is *centralization of the fibula* [10, 14]. Preconditions are a largely normal distal femur and a sufficiently strong quadriceps muscle. If the femur is severely deformed and a pronounced flexion contracture of the knee is present, a *knee disarticulation* should be performed before the patient starts to walk, since it will be almost impossible to fit a prosthesis or orthosis to the flexed knee and the child will not be capable of walking. Occasionally, *arthrodesis of the femur and fibula* can be useful (particularly if the fibula is also deformed [31]), although it should be borne in mind that the growth plates can be adversely affected by an early arthrodesis.

#### Type II (absence of the distal half of the tibia)

The primary objective here is to preserve a stable knee. To this end, a side-to-side fusion of the tibia and fibula is recommended. At the distal end, the arthrodesis of the fibula and talus should be accompanied by amputation of the forefoot as part a modified Boyd procedure. The surgeon should be careful to ensure that the epiphyseal plate of the distal fibula is preserved.

#### Type III (tibiofibular diastasis)

The main problem in this type of deficiency is the instability of the talus beneath the tibia. The talus has a strong tendency to dislocate cranially, causing the Achilles tendon to shorten since it is not stretched. The lateral malleolus protrudes strongly and tends to perforate the skin. An external fixator can be used to reduce the rearfoot back underneath the tibia. The talus and tibia should then be transfixed with a medullary nail and the distal section of the tibia and fibula should be fused. Amputation of the foot is only required in rare cases [5, 15, 35].



■ Fig. 3.297a, b. 8-month old boy with *tibiofibular diastasis* (type III longitudinal deficiency of the tibia). X-rays of the right lower leg: AP (a) and lateral (b)

### 3.3.6.3 Congenital dislocation of the knee

#### ► Definition

Congenital dislocation of the tibia ventrally in relation to the femur with hyperextension of the knee and shortening of the quadriceps muscle. Frequently accompanied by a valgus deformity and rotation. Occurs with or without cruciate ligament aplasia.

### Classification

The classification according to Leveuf [4] is shown in **Table 3.49**.

### Occurrence

A Danish study has calculated an incidence of 1.5 cases per 100,000 neonates [26].

### Etiology

During pregnancy the knee remains in a hyperextended position in some cases (approx. 20%). The lack of cruciate ligaments or fibrosis of the quadriceps can, in particular, lead to dislocation of the knee. However, whether the aplasia of the cruciate ligaments is a triggering factor or a secondary phenomenon is not known. Most cases occur sporadically and are not hereditary.

### Associated anomalies

Congenital dislocation of the knee can occur unilaterally or bilaterally, in isolation or in connection with an arthrogryposis or a Larsen syndrome. It also occurs together with congenital hip dysplasia, clubfoot and other foot anomalies.

### Clinical features, diagnosis

The dislocation of the knee is usually obvious at birth. Since the joint is often in an extreme, hyperextended position, a brief inspection allows an unequivocal diagnosis to be made. Sometimes a hyperextension up to 90° and more is possible. An *x-ray* will confirm the diagnosis, and a lateral view will usually show increased inclination of the tibial plateau towards the back (**Fig. 3.298**).

The differential diagnosis must distinguish between a congenitally recurvated knee and subluxation or dislocation. While the knee is also (slightly or moderately) hyperextended in a recurvated knee, the joint surfaces of the femur and tibia are in regular opposition. If the knee is subluxated or dislocated, an *ultrasound scan* can confirm the presence of the cruciate ligaments at an early stage [38]. The Frick tunnel view shows whether a fossa is present, providing reliable evidence of the presence of the cruciate ligaments [34]. An even more reliable picture of the internal structures can be obtained

by *arthrography*. An *MRI scan* also provides evidence of the condition of the cruciate ligaments. However, since arthrography and MRI are invasive or expensive investigations, these should only be used if possible therapeutic consequences are involved. Since this is not usually the case in the early phase, we restrict ourselves to the ultrasound scan.

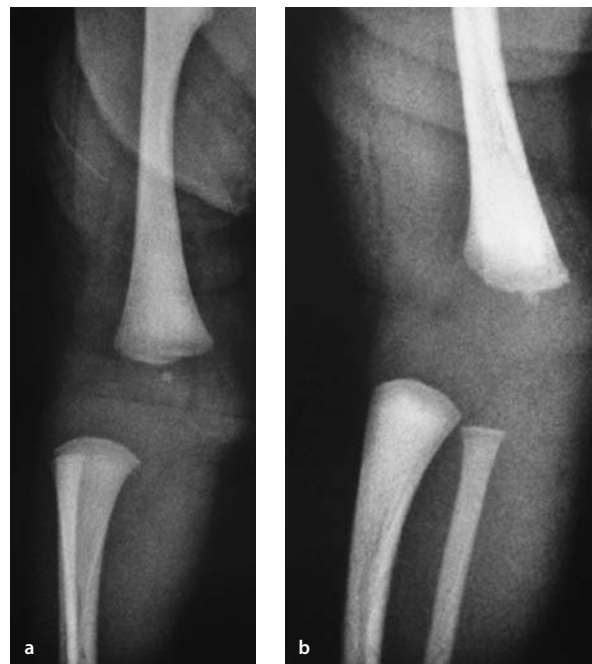
### Treatment

The treatment should start immediately after birth and consists of intensive correction and stretching of the quadriceps. Placing the infant in an appropriate position can also prove helpful. The hip is placed in 90° flexion and the thigh supported down to the knee with a foam block; a weight is secured to the lower leg with bandages and traction is applied in the direction of flexion. When the neutral position has been reached, corrective casts can then be fitted in increasing flexion. This treatment is very successful during the first 3 months [16, 36].

If treatment is not started until after this time, operations often cannot be avoided. By this stage, the quadriceps can be surgically lengthened to permit flexion of the knee. Occasionally, the cruciate ligaments must also be divided. Naturally, the results of this treatment are only moderate [16], whereas patients treated conservatively at an early stage can usually expect largely normal knee function provided the dislocation has not occurred in connection with an arthrogryposis or Larsen syndrome.

**Table 3.49. Classification of congenital dislocation of the knee. (According to Leveuf [4])**

Type	Parameter
I	Hyperextended knee
II	Subluxated knee
III	Completely displaced knee (commonest type)



**Fig. 3.298a, b.** 1-month old infant with congenital *dislocation of the knee*. X-rays of the right knee: AP (a) and lateral (b)



### 3.3.6.4 Congenital absence of cruciate ligaments

#### ► Definition

Congenital absence of – usually both – cruciate ligaments, often associated with congenital dislocation of the knee, also occurs in connection with femoral hypoplasia or a congenital proximal femoral deficiency and fibular or tibial hypoplasia / aplasia.

#### Occurrence, etiology, associated anomalies

Congenital cruciate ligament aplasia very rarely occurs in isolation, but is very common in combination with other anomalies of adjacent structures and is often not diagnosed. The etiology is not known. Congenital cruciate ligament aplasia is particularly observed in connection with congenital femoral hypoplasia, a proximal focal femoral deficiency, fibular deficiency and congenital dislocation of the knee [27, 34]. In tibial deficiency, by definition, the cruciate ligaments are absent. No figures relating to its incidence are available.

#### Clinical features, diagnosis

Absence of cruciate ligament is rarely diagnosed during infancy. In most cases, a careful clinical examination during early childhood, or later, raises the suspicion that these ligaments are missing. Clinical examination reveals an – often pronounced – translation of the lower leg in relation to the femur in the sagittal plane of 15–20 mm with a soft anterior and posterior end point. The pivot shift test (► Chapter 3.3.8) is distinctly positive. In addition to the abnormal mobility in the sagittal plane, valgus and varus rotation at knee level is increased (increased medial and lateral opening). Despite the marked instability, effusions rarely occur, and giving-way phenomena are not often observed. However, the children often have an insecure feeling, particularly during rotational movements. The knee adapts itself to the situation of missing cruciate ligaments and the femoral condyles develop differently from normal as a functional adaptation.

A *plain x-ray* shows a less pronounced intercondylar eminence [34]. This is particularly apparent on the Frick tunnel views. Additionally, the femoral condyles do not show the normal curvature. In the infant, the *ultrasound scan* indicates the absence of the cruciate ligaments. In the older child, the diagnosis can be confirmed by *arthrography*, an *MRI scan* or *arthroscopy*. Since all of these investigations are expensive and/or invasive they should only be performed in the investigation result significantly affects the treatment.

#### Treatment

The treatment of cruciate ligament aplasia should be conservative initially and also based naturally on the underlying disease. If there is no subjective feeling of instability no treatment is required. However, if the child

falls over with excessive frequency, snapping phenomena or pseudolocking can occur. In this case, an *orthosis* will be needed, ideally consisting of a »Heussner spring«, i.e. an elastic support with metal lateral reinforcements and a hinged joint. Even though experiments have shown that such a support hardly has any effect on the sagittal forces (and only reduces valgus and varus rotation slightly), it can greatly improve the subjective feeling of stability in children, in particular, and does not constitute any great handicap. Greater stability (but also constituting more of a handicap) is provided by the Lenox-Hill brace.

Treatment must be continued conservatively for as long as possible. A suitable surgical procedure for replacing the anterior, and particularly the posterior, cruciate ligaments is not possible while considerable growth potential remains and is also extremely difficult towards the end of growth, as the shape of the condyles is not normal. Usually the knee undergoes a certain functional adaptation to the lack of stability over time. Furthermore, since the affected children are not generally very active in sports, the instability-related handicap on completion of growth is not comparable with that produced by a complete traumatic rupture of both cruciate ligaments. After closure of the epiphyseal plates, surgical replacement of the cruciate ligaments with autologous tendon tissue may be considered, but only after careful evaluation of the handicap.

### 3.3.6.5 Congenital pseudarthrosis of the tibia

#### ► Definition

Congenital disorder of bone formation in the distal part of the tibia, with anterior bowing and hamartomatous foreign tissue. A fracture can occur at birth, or also secondarily, and fails to heal because of impaired callus formation, resulting in a pseudarthrosis. The disorder can occur with or without neurofibromatosis.

— *Synonyms:* Crus varum congenitum, infantile pseudarthrosis of the tibia, CPT

#### Classification

Classifications have been proposed by Crawford [12], Boyd [6] and Andersen [3]. The principal types are presented in ■ Table 3.50 and ■ Fig. 3.299–3.302, while ■ Fig. 3.303 provides a schematic presentation of the various types covered by these three known classifications, plus a number of special forms.

#### Occurrence

This is a rare malformation with an incidence of 0.5 per 100,000 births [3]. A concurrent neurofibromatosis exists in around half of the cases [25]. On the other hand, this is one of the commonest hereditary disorders (► Chapter 4.6), and one study found pseudarthrosis of the tibia to be present in 13% of patients with neurofibromatosis [12].



### Etiology

The circulation in the tibia is poorest at the junction between the mid and distal third. This is where delayed healing or even pseudarthroses can occur quite often in (usually adult) patients without any congenital disorder after tibial fractures caused by appropriate trauma but receiving inadequate treatment. This is also the primary site of the pathology in »congenital« pseudarthrosis of the tibia, which is not actually congenital in most cases, since only the predisposition is congenital. Consequently, the term »infantile pseudarthrosis of the tibia« has been proposed, although this has not yet gained widespread

acceptance. »Developmental pseudarthrosis of the tibia« would be another option. Differing illnesses are involved in terms of the etiology.

The dysplastic and cystic types are often associated with a neurofibromatosis, and histological examination reveals neurofibromas in the pseudarthrosis tissue. In patients without neurofibromatosis, the same examination shows a thickened periosteum with proliferating, hamartomatous fibroblastic tissue (resembling the picture of fibrous dysplasia), which disrupts the circulation and callus formation [50]. A typical pseudarthrosis has been produced in rabbit studies by applying a circular, imper-

■ Table 3.50. Classification of congenital pseudarthrosis of the tibia

Type		Parameter
Anterior bowing only	Crawford type I	The tibia is largely normal at birth. Subsequently, anterior bowing and shortening. Fracture in the first years of life after slight trauma. Neurofibromatosis very rare
Anterior bowing, varus curvature and sclerosis	Crawford type II	The tibia is largely normal at birth. Subsequently, anterior bowing and shortening. Fracture in the first years of life after slight trauma. Neurofibromatosis very rare
Cystic	Crawford type III	The tibia is not thinned, but cystic inclusions that histologically resemble fibrous dysplasia are present in the distal third. Usually fractures in the first few months of life. Neurofibromatosis common
Dysplastic	Crawford type IV	The distal third of the tibia and fibula is thinned in an hourglass shape, sclerosed, and the medullary canal is partially or completely obliterated. The tibia shows anterior bowing. The fracture usually occurs when the baby starts walking. Very frequently associated with neurofibromatosis



■ Fig. 3.299a, b. 1-year old boy with *congenital pseudarthrosis of the tibia, Crawford type I* (anterior bowing only). AP (a) and lateral (b) x-rays of the left lower leg

■ Fig. 3.300. 1-year old girl with *congenital pseudarthrosis of the tibia, Crawford type II* (antecurvature with sclerosis). AP (a) and lateral (b) x-rays of the right lower leg



Fig. 3.301. 4-month old boy with congenital pseudarthrosis of the tibia, Crawford type III (cyst formation). AP and lateral x-rays of the of the right lower leg

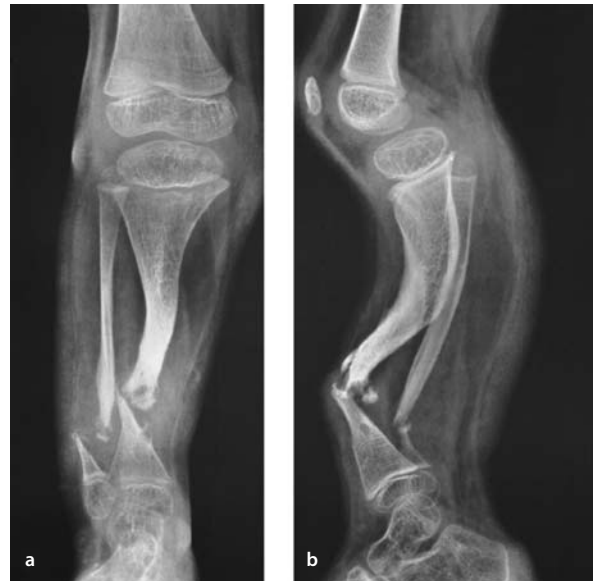


Fig. 3.302a, b. 1-year old boy with congenital pseudarthrosis of the tibia, Crawford type IV (dysplastic type, also with cystic changes). AP (a) and lateral (b) x-rays of the left lower leg

					Pseudarthrosis of fibula without pseudarthrosis of tibia	Intraosseous neurofibromatosis	Club-foot + antecurvature of tibia
<b>Crawford</b>	<b>I</b>	<b>II</b>	<b>III</b>	<b>IV</b>			
<b>Andersen</b>		Sclerotic	Cystic	Dysplastic			Association with clubfoot
<b>Boyd</b>	<b>I</b>	<b>IV</b>	<b>III</b>	<b>II</b>	<b>V</b>	<b>VI</b>	

Fig. 3.303. Classification of congenital pseudarthrosis of the tibia according to Crawford

meable tube around the tibia. Various types of disorders are probably involved and can manifest themselves here at a point of reduced resistance in terms of circulation.

**Clinical features, diagnosis**

The dysplastic type, and usually the cystic type as well, are generally diagnosed at birth. Even if the pseudarthrosis is rarely evident at this point, anterior bowing and possible shortening of the lower leg will nevertheless indicate the existence of a problem. An x-ray should be arranged if such findings are observed. The typical radiographic changes have already been described in the classification section. The late form is usually diagnosed only as the

infant grows during the first few years of life. Here too, anterior bowing and varus curvature, and possibly shortening as well, provide evidence of a disorder. Occasionally, however, the diagnosis is only made when a distal tibial shaft fracture fails to heal despite adequate treatment.

**Treatment**

Provided no fracture is present, splinting with an orthosis should help avoid excessive bowing and possibly also fracturing of the tibia.

When a pseudarthrosis is established, treatment is based partly on the type and partly on the stage of the disease. A multicenter study conducted by the European

### 3.3.6 · Congenital deformities of the knee and lower leg

Paediatric Orthopaedic Society investigated a total of 370 cases across Europe [20]. The following *observations relating to treatment* were made as a result of the study:

- ❗
- **Surgical treatment for children under 3 years of age is almost always unsuccessful.**
- **The best chances of success exist after completion of growth.**
- **Internal fixation with plates and operations with (telescopic) medullary nails are not very successful.**
- **Effective methods include segment transport with the external fixator and the transfer of the vascularized fibula.**
- **A crucial requirement for successful fusion is the complete removal not only of the pseudarthrosis bone, but also the surrounding altered fibrous soft tissues.**

In segment transport, a ring fixator is fitted and the pseudarthrosis and surrounding soft tissues are resected. The bone ends are placed under compression by the fixator, and the tibia and fibula are osteotomied above this point and lengthened [19, 37]. We have used this method successfully on several occasions. The transfer of the vascularized fibula will only prove successful in the hands of an experienced team with a microsurgeon [11, 20, 45]. Here too, complete removal of the fibrous soft tissue surrounding the pseudarthrosis is an important requirement for this method.

Children with congenital pseudarthrosis of the tibia almost invariably face a protracted period of suffering, since the fusion often does not succeed on the first attempt (particularly with the cystic and dysplastic types). An amputation is sometimes the final outcome.

Internal fixation with plates and screws has clearly *not proved effective*. They worsen the already impaired circulation and prevent any increasing compression because of bone resorption. Nor can fixation with a medullary nail produce healing, since this fails to produce adequate compression. The failures can also probably be explained partly by the fact that the pseudarthrosis and surrounding soft tissue areas are usually inadequately resected with this method. Nor is conservative treatment capable of producing any fusion. Bridging with allogeneic bone has likewise

failed to achieve the hoped-for success. Attempts have been made in individual hospitals to produce a cure by electrostimulation [39], although the results to date have not been very convincing.

#### **Our therapeutic strategy for an established pseudarthrosis of the tibia**

Our therapeutic strategy for an established pseudarthrosis of the tibia is shown in ■ Table 3.51 [25].

#### **Prognosis**

The treatment of pseudarthrosis of the tibia, and particularly the dysplastic and cystic types, is extremely problematic. The number of failures is high for all methods and refractures occur repeatedly (■ Fig. 3.304). The risk of further fractures diminishes with time in the older child, particularly after completion of growth. Only the late form has a good prognosis [42]. While the absence or presence of neurofibromatosis does not affect the prognosis [20], it does worsen with the increasing number of repeat operations.

#### **3.3.6.6 Discoid meniscus**

##### ➤ **Definition**

Congenital defective formation of the lateral meniscus, which is consequently shaped like a disc rather than a horseshoe.

##### **Classification**

The most popular *classification* was proposed by Watanabe on the basis of his arthroscopic experience (■ Table 3.52). In the Wrisberg type, the lateral meniscus is not

■ **Table 3.52. Classification of discoid meniscus according to Watanabe**

Type	Name	Frequency [40] (%)
I	Wrisberg type	10
II	Complete type	80
III	Incomplete type	10

■ **Table 3.51. Our therapeutic strategy for an established pseudarthrosis of the tibia**

Age 1–5	Conservative treatment stabilizing lower leg orthosis. Only if this is unable to prevent severe bowing, may the option of internal splinting with telescopic medullary nails inserted through the proximal and distal tibial epiphyseal plates be considered. The goal of treatment is to prevent a fracture. If this has already occurred, however, fusion will not be achievable by this method
From 5 years	Resection of the pseudarthrosis and the surrounding connective tissue, proximal corticotomy in healthy tissue and »segment transport« with the ring fixator (e.g. Ilizarov apparatus). This procedure initially places the pseudarthrosis under compression during shortening, and the leg is subsequently lengthened proximally. If the condition fails to heal, a vascularized fibular graft is inserted



**Fig. 3.304a–f.** Patient with *congenital pseudarthrosis of the tibia, Crawford type III*. AP and lateral x-rays, in each case, of the left lower leg. **a** At the age of 6 years with tibial still intact and cyst formation. **b** At the age of 11 years after fracture and osteolysis, failed healing after conservative treatment. **c** After fitting of the Ilizarov apparatus and insertion of a medullary nail for splinting purposes, with acute

shortening at the site of the pseudarthrosis and lengthening proximally (age: 12 years). **d** At the age of 13 years the proximal lengthening has consolidated well, in contrast with the pseudarthrosis. **e** Fitting of a unilateral external fixator at the age of 14 years. **f** Healed result of the pseudarthrosis at the age of 15 years

anchored to the tibia but is attached only to the lateral meniscofemoral (Wrisberg) ligament, making it hypermobile. It is also hypertrophied. In type II (complete type), the meniscus is very thick and in the shape of a complete disc, while the anchorage is normal. A type III (incomplete type) disc has a more semilunar shape, and here too the meniscus is thicker than usual but with regular anchorage.

#### **Occurrence, etiology, associated knee problems**

While we are not aware of any epidemiological studies, this is a fairly common anomaly. Both sexes are affected with equal frequency. The incidence appears to be higher in East Asia [48] than in Europe. Of the knee problems suffered by children, discoid meniscus is the commonest reason for an investigation or intervention [22]. Medial discoid menisci have also been described in very isolated

cases [9, 46]. While most discoid menisci occur sporadically, there are isolated reports of a familial occurrence [18]. Although other congenital malformations are no more common in children with discoid menisci than in the normal population, osteochondrosis dissecans does occur more frequently in association with a discoid meniscus (in approx. 15% of cases) [2]. A recent study showed an even stronger association between a discoid meniscus and osteochondritis dissecans of the lateral femoral condyle [13].

### Pathogenesis

It was initially assumed that the disc shape developed as a result of incomplete breakdown of the central section of the meniscus. However, embryological studies have shown that the lateral meniscus does not show a discoid appearance at any phase of fetal development [29]. The knee develops from an undifferentiated mesenchymal mass from which the femur, tibia and intra-articular structures form at the end of the second month of pregnancy. By the time the joint space has formed, the central mesenchymal mass has disappeared and mesenchymal tissue is only present at the edges, where the cartilage subsequently differentiates to form the menisci. In discoid meniscus, therefore, fibrocartilage must develop from mesenchymal tissue at a site where this does not normally occur [46].

Some discoid menisci, however, are caused by a different process. In these cases the normal anchorage of the lateral menisci on the tibial condyle is lacking, and the meniscus is only secured to the lateral meniscofemoral (Wrisberg) ligament. Such menisci are hypermobile and become hypertrophied as a result of mechanical loading. The posterior horn, which is not attached to the tibia, subluxates medially on extension [29]. The Wrisberg type is usually symptomatic. MRI studies with adults have shown that meniscal tears occur significantly more frequently in those with discoid menisci compared to those with normal, horseshoe-shaped menisci [44].

### Clinical features, diagnosis

Symptoms are only very rarely present in infancy and do not usually appear until the age of 5–6 years, when a snapping is noted in the lateral part of the knee. On examination, the snapping phenomenon can be elicited on active extension of the flexed knee, usually at approx. 20° flexion. This finding is observed particularly in the hypermobile Wrisberg-type, which is normally symptomatic, whereas the complete and incomplete types do not usually cause any symptoms. *Radiography* regularly shows a slight widening of the lateral knee joint space [30]. Otherwise the plain x-ray is of no help.

A discoid meniscus is readily visualized by an MRI scan. However, in addition to the cost aspect, this investigation involves the drawback of often having to be performed under anesthesia in small children. The clearest

diagnosis, including the type of meniscus, can be established by arthroscopy. Since the treatment can also be administered in the same anesthetic session, we dispense with the MRI scan for a clinically suspected discoid meniscus and, if there is a need for treatment, proceed directly to arthroscopy. Disorders to be considered in the *differential diagnosis* include meniscal cysts, congenital subluxation of the knee, congenital cruciate ligament aplasia, the snapping of tendons and dislocation of the patella.

### Treatment

While discoid menisci cause no, or just a few, symptoms, there is no need for surgical treatment. If frequent, unpleasant snapping occurs, however, arthroscopy and possible treatment are indicated. Many authors continue to recommend a complete lateral meniscectomy, and good results are reported in individual cases [52].

Our own long-term studies, however, have shown that there is a substantial risk of osteoarthritis after total meniscectomy [41]. For this reason (particularly for the complete and incomplete types), only the central section of the meniscus should be removed. This procedure should, if possible, be performed by arthroscopy and is technically more difficult than a resection for a flap or bucket-handle tear. The treatment of the hypermobile Wrisberg type, for which a complete meniscectomy used to be recommended, is even more tricky. We have tried refixing the lateral posterior horn, although we do not have extensive experience with this procedure and we are unaware of corresponding reports in the literature.

### 3.3.6.7 Changes to the knee and lower leg in systemic disorders

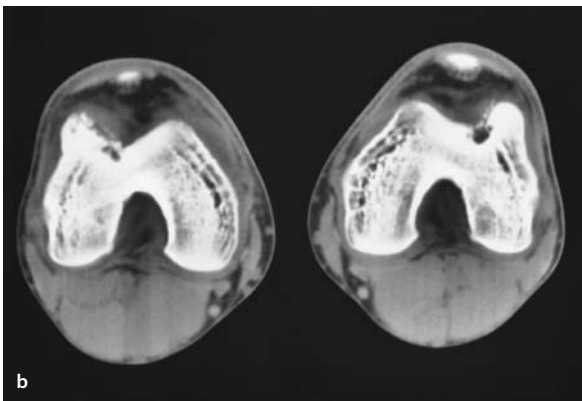
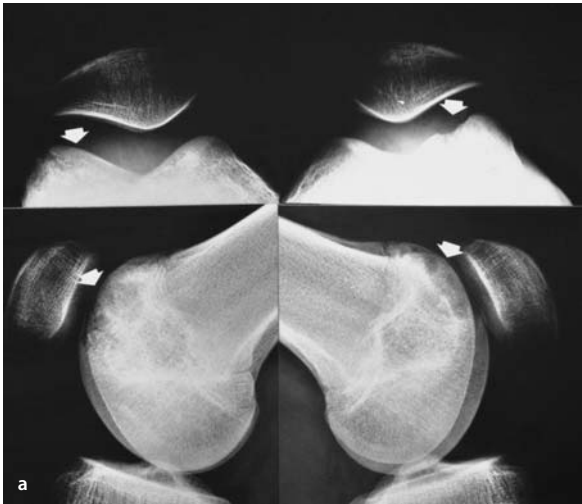
#### Multiple epiphyseal dysplasia

This condition is described in detail in ► chapters 4.6.2.11 and 3.2.7.2. The (rare) more serious form (Fairbank type) invariably also involves changes in the epiphyses in the vicinity of the knee. In some patients, however, only the two femoral condyles are affected. The clinical picture resembles that of osteochondrosis dissecans, except the foci are larger and present on both sides. The foci may also be located in unusual sites, for example on the anterior aspect of the lateral femoral condyles (■ Fig. 3.305).

#### Dysplasia epiphysealis hemimelica

This disorder is described in detail in ► chapter 4.6.2.31. It is a systemic disease involving abnormal, osteocartilaginous formations in the epiphyses and carpal or tarsal bones of the medial or lateral half of a limb. The diagnosis is usually made during early childhood. The commonest sites are the tarsal bones and the distal femoral and proximal tibial epiphyses (■ Fig. 3.306). The changes lead to joint incongruity and deformity, with genu valgum or varum.





**Fig. 3.305a, b.** 16-year old boy with *multiple epiphyseal dysplasia* in the area of the lateral femoral condyle. **a** Axial x-rays of the femoral condyles and patellas of both knees and lateral x-rays of both knees. **b** CT scans (horizontal section through the femoral condyles)



**Fig. 3.306a, b.** 9-year old girl with *dysplasia epiphysealis hemimelica* with osteocartilaginous formations in the area of the lateral femoral condyle. AP (**a**) and lateral (**b**) x-rays of the left knee

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### 3.3.7 Neurogenic disorders of the knee and lower leg

R. Brunner

The commonest problems in the knee from the neurological standpoint concern flexion and extension. Full extension of the knee is required in order to be able to stand without expending large amounts of muscle power. If this is no longer possible, however, the energy expenditure required for standing and walking increases. Either the knee extensors must exert compensatory muscle power or else an external extension moment can be generated by forward inclination of the trunk. In the latter case, the hip extensors are used as compensators for maintaining posture. As a conservative orthopaedic treatment, orthoses can be used to preserve the dynamic stability of the knee joint.

**!** **Functional problems in the sagittal plane – such as hyperextension or a knee flexion contracture – are common in all neurogenic disorders. Since braces that surround the knee or ankle foot orthoses only provide lateral stabilization, it is almost impossible to influence flexion/extension movements, apart from arresting the knee, with such devices. They are only effective in helping avoid knee hyperextension in stance.**

#### 3.3.7.1 Primarily spastic paralyses Functional disorders

##### ➤ **Definition**

Functional changes in the knee with no structural deformity and caused by spastic muscle activity.

■ Table 3.53 provides an overview of the common functional disorders in primarily spastic locomotor disorders (▶ Chapter 4.7.3).

### Knee flexion on foot-strike

#### ➤ Definition

The flexion of the knee at the start of the stance phase is excessively pronounced since the spasticity prevents efficient knee extension in the swing phase.

Young patients, in particular, with spastic diplegia often show a gait pattern with increased knee flexion on foot-



■ Fig. 3.307. Patient with *equinus* gait pattern despite the plantigrade position of the foot as it strikes the ground. The spastic activity of the knee flexors stops the knee from extending sufficiently to place the heel on the ground

strike and in the first half of the stance phase. As a result of the inadequate knee extension at the end of the swing phase the foot strikes the ground on tiptoe despite the plantigrade position of the foot (■ Fig. 3.307). Neither ankle foot orthoses nor corrective casts are capable of placing the heel on the ground. Ankle foot orthoses are nevertheless required to stabilize and support the feet. Typically, the patient continues to walk on tiptoe even when fitted with an ankle foot orthosis designed to produce a plantigrade position of the sole in relation to the lower leg axis. In this case, an incorrect foot position cannot be the cause of the equinus gait pattern.

The therapeutic objective is to reduce the spasticity of the hamstring muscles, although this is difficult in practice. Since this gait pattern will otherwise develop into a crouch gait, regular physical therapy is required to counteract any contracture of the hamstrings. Strength training is important for the knee extensors, which are constantly overstretched in this position.

### Crouch gait

#### ➤ Definition

A crouch gait refers to a gait pattern with permanently flexed hips and knees and typically dorsiflexed ankles.

A crouch gait can result from various causes:

- Foot: talipes calcaneus,
- Knee: knee flexion contracture,
  - Contracture of the hamstring muscles,
  - Spasticity of the hamstring muscles or the rectus femoris muscle,
  - Weakness/excessive length of the knee extensors
- Hip: hip flexion contracture

■ Table 3.53. Functional disorders in primarily spastic locomotor disorders

Deformity	Functional benefit	Functional drawbacks	Treatment
Giving-way of the knee in the early load-bearing phase	–	Increased energy expenditure Risk of contracture	Muscle stretching Reduction of spasticity
Crouch gait	–	Increased energy expenditure Retropatellar pain	Pes calcaneus: ankle foot orthosis Contracture of hamstring muscles: lengthening Hip flexion contracture: lengthening of the flexors, correction of lever arms, shortening of overlong muscles
Stiff knee in the swing phase	–	Small-stepped gait Circumduction	Rectus transfer
Knee hyperextension	Indirect knee stabilization	Overstretching of the posterior elements Pain	Ankle foot orthosis in forward inclination

*Talipes calcaneus* develops as a result of a functional insufficiency of the triceps surae muscle. It may be present as a primary condition or secondarily to excessive or unnecessary lengthening of the Achilles tendon. In functional terms, this produces an uncontrollable – for the patient – forward inclination of the lower leg in relation to that of the foot on the ground during the stance phase. The knee and hip must therefore be flexed in order to keep the center of gravity over the stance area (■ Fig. 3.308). Provided no structural changes have occurred at knee level, the talipes calcaneus must be treated. To prevent the occurrence of a secondary talipes calcaneus, any lengthening of the triceps surae muscle should only be continued until the neutral position is just reached, if the proximal muscles (extensors at the knee and hip) are not completely sufficient.

❗ **Slight shortening of the triceps surae muscle (slight footdrop) prevents the crouch gait, particularly if the knee extensors are insufficient.**

Spastic hyperactivity of the hamstring muscles likewise results in a crouch gait, irrespective of the shape of the foot, although it can also be present in feet with an equinus deformity. The therapeutic objective must be to reduce the spasticity (► Chapter 4.7.2) in order to facilitate looser knee extension without muscle tensing. The primary



■ Fig. 3.308. Patient with *crouch posture*, caused by weakness of the triceps surae muscle. The insufficiency of this muscle produces forward inclination of the lower leg, requiring compensatory flexion at the knee and hip in order to keep upright

emphasis is on physical therapy with the aim of preserving the full length of the hamstrings. Positional splints may also be helpful. If these muscles are not stretched regularly, there is a substantial risk of contractures (see below).

A knee extensor weakness can also lead to deficient knee extension, particularly if the compensatory mechanisms (triceps surae muscle, hip extensors) do not come into play. In order to be able to stand and walk with a *hip flexion contracture*, the knee must secondarily be held in a flexed position. This produces a crouch gait with normally functioning hamstrings. The crouch gait can lead to *complications*. Retropatellar pain occurs not infrequently, irrespective of age. In order to keep upright with flexed knees and hips, the patient constantly has to tense the quadriceps muscles by way of compensation, thereby overstressing the patella (► Chapter 4.7.2, »Walking and standing«) and gradually overstressing the extensor mechanism. This can result in insidious rupture of the patellar ligament, rectus tendon or – occasionally insidious – fracture of the patella. The treatment must not only restore the extensor apparatus (shortening of the extensors) but also the length of the knee flexors [6].

### Stiff-knee gait

#### ► Definition

Constant or asynchronous activity of the rectus femoris muscle prevents knee flexion in the swing phase.

If the *rectus femoris muscle* is out of phase or constantly active, this muscle will prevent adequate flexion during the swing phase despite a crouch gait. Although extension of the knee flexors will then produce a more upright gait, the defective rectus activity prevents forward swinging of the leg because the knee is inadequately flexed [10–12]. The gait becomes less dynamic and the stride length shorter. The range of motion of the knee during walking can be increased by approx. 20°, without any major decline in the strength of the extensor mechanism, by transposing the rectus femoris muscle distally to the medial knee flexors [7, 10, 17, 21]. This procedure will improve the patient's ability to walk.

### Gait with hyperextension of the knee

#### ► Definition

The knee is overstretched in the early stance phase and remains in this position until the end of the stance phase.

The spastic contraction of the *triceps surae muscle* stiffens the ankles and blocks the dorsal extension movement of the foot in the stance leg phase during walking. The thigh then continues its forward motion in relation to the lower leg and the knee is hyperextended (during normal

walking the forward motion of the lower leg is controlled by eccentric activity of triceps surae muscle). If severe spasticity is present, the intrinsic triceps reflex can even move the lower leg in the opposite direction of walking, which likewise produces hyperextension and is inefficient in terms of energy use. In both cases, the treatment must address the functional or structural equinus foot (► Chapter 3.4.10).

### Structural changes

#### ► Definition

Structural deformity of the knee caused by spastic muscle activity

■ Table 3.54 provides an overview of common structural deformities in primarily spastic locomotor disorders.

### Contracture of the hamstring muscles

#### ► Definition

Structural contracture of the hamstrings is present even at rest, thereby preventing extension of the knee.

Extension of the knee with the hip extended is the crucial factor in evaluating the functional significance of a contracture of the hamstring muscles. The degree of knee extension with the hip flexed, on the other hand, provides information about the length of the knee flexors and their contribution to the extension deficit of the knee. The severity of the contractures of the hamstring muscles with the hip flexed is irrelevant, however, for as long as the knee can be fully extended (i.e. slightly overstretched) with the hip extended when the patient is examined on the couch. Only with a knee flexion contracture of approx. 15–20° and above with the hip extended does the knee flexion contracture increase further over the course of time as result of gravity. This also increases the load on the extensor mechanism, which always has to perform the necessary postural work by way of compensation. As a result, the extensor mechanism runs the risk of rupturing in the long term. If posture can no longer be controlled, the patient's ability to walk and stand is jeopardized.

The *therapeutic strategy* is aimed at extending the knee. If full extension is achieved, the knee flexors are regularly extended sufficiently by standing – and possibly also by walking – thereby improving the gait [2, 3, 9, 12, 20]. The treatment for contractures between 10° and 15° involves intensive physical therapy with stretching exercises, backed up in individual cases by knee extension splints. If the knee flexion contractures increase, lengthening of the knee flexors is indicated – regardless of the patient's age – if these muscles are contributing to the contractures.

As well as knee flexors, the hamstrings also act as hip extensors. Before this muscle group is lengthened, other possible causes of the crouch gait must be ruled out in order to avoid any adverse effects on hip extension. Temporary hip extensor weakness has been reported after the lengthening of the hamstring muscles. Hence hamstring lengthening needs to be done very cautiously. Preoperative gait analysis is also needed to establish whether any additional deformities of other joints also require correction and the extent to which any defective activity of the rectus femoris muscle contributes to the contracture (see relevant section). Walking function will be improved [12, 20] and energy expenditure reduced [13] only if the contractures of all the affected leg joints are corrected. If contractures that have developed by way of compensation are not addressed at the same time, the lengthening of the hamstring muscles will not prove very effective.

The most appropriate *follow-up treatment* after knee flexor lengthening involves the fitting of a knee extension splint (instead of the previously used casts; ■ Fig. 3.309). The splint is used to increase the stretching of both the muscles and the other soft tissues (particularly the nerves) in a continuous and gradual manner. The extent of the stretching can be adapted to the patient's symptoms, and the splint can be removed for nursing care procedures. Since the latter allow regular inspection of the skin pressure sores can be avoided. To ensure that the splint can be used in the immediate postoperative period it must be prepared before the operation. The previously used cast treatments not only resulted in

■ Table 3.54. Structural deformities in spastic locomotor disorders

Deformity	Functional benefit	Functional drawbacks	Treatment
Contracture of hamstring muscles	(Hip extension)	Energy use increases during walking and standing	Lengthening
Patellar dislocation	–	Pain Instability	Recentering of the patella (Green, Stanislavljevic, Elmslie)
Rotational deformity	Compensation of rotational deformities in the hip and foot	Entanglement of feet, feet not in the direction of walking	Correction osteotomy



pressure sores, but also long-term nerve damage from overstretching [1].

If the contractures had been slight, the follow-up treatment phase is relatively short, particularly because the quadriceps will not have adapted by lengthening excessively in performing its postural work. If the operation is not carried out until the knee flexor contractures are very pronounced ( $80^\circ$ – $90^\circ$ ), the follow-up treatment and rehabilitation will last for years because of the insufficiency of the quadriceps femoris muscle. It is more useful, therefore, to shorten any excessively long knee extensors in the affected segment and thus restore its proper tension. Otherwise a relapse will occur because the patients are often unable to cope with such a long rehabilitation phase and the muscle can no longer compensate for its overlength.

If the contractures have already been present for several years, the joint capsule and ligaments will also have shortened, in which case a simple muscle-tendon lengthening procedure will no longer be sufficient. The dorsal capsule of the knee can also be released in the same procedure (we do not have any experience with this method). Over time, however, all the other soft tissues, including the subcutaneous fascia and the skin, will become con-

tracted, and surgical lengthening of all these structures is not feasible. If the contractures have been present for a prolonged period we recommend lengthening of the knee flexors and follow-up treatment until no further progress can be made. A supracondylar extension osteotomy is added in these cases in order to restore full extension. In either case, the goal of treatment must be full extension at the knee. The more residual flexion remains, the greater the likelihood of a recurrence.

We use the extension splint as follows: Directly after the operation, the splint is worn at all times (except for nursing care procedures). Once substantial stretching has been achieved, the splint may be worn for shorter periods. It has proved to be more beneficial to wear the splint for several hours during the day rather than at night, because sleep is disturbed by the uncomfortable sensations produced by muscle stretching. When full extension has been restored, a recurrence can be delayed, or even prevented, by wearing the splint for approx. 15–30 min every 2–3 days. If severe contractures are present it may prove necessary to use the knee extension splint as a functional orthosis in order to prevent the knee extensors from being permanently overstretched.

**! The decision to proceed with surgical lengthening, and particularly the timing of the operation, must be based on the functional handicap and the extent of the deformity rather than the patient's age.**

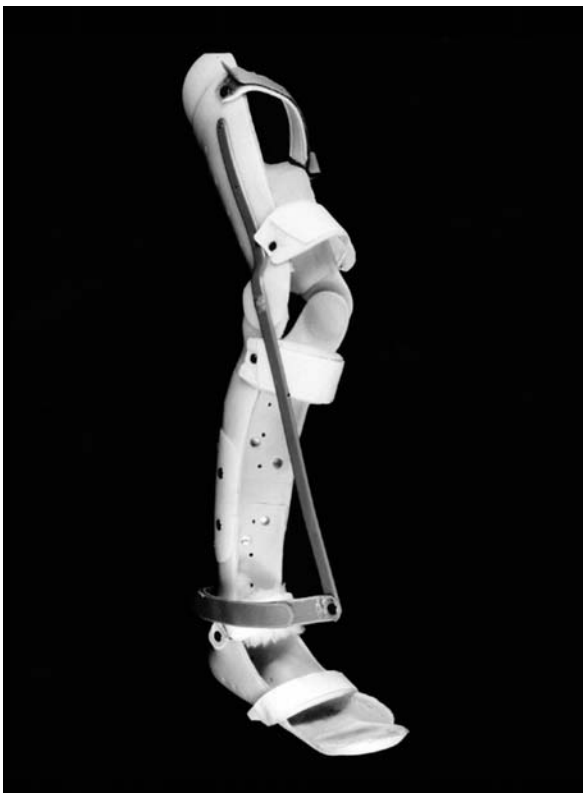
In addition to knee extension, spasticity can also block knee flexion during the swing phase. The result is delayed flexion, after which there is insufficient time for the extension and the knee remains in the flexed position during foot-strike. Moreover, the knee is stiff in the swing phase. This abnormal gait can be documented during gait analyses, and the EMG shows a prolonged, out-of-phase activity of the rectus femoris muscle. In such cases, the rectus femoris muscle can be transposed to the knee flexors (gracilis or semitendinosus muscles) [10, 11, 22]. Less than 20% of knee extension force is lost as a result of this procedure, whereas knee flexion is improved by  $10$ – $20^\circ$  in the swing phase [15]. By contrast, injections of botulinum toxin into the rectus femoris muscle produce disappointing results in our experience.

### Habitual dislocation of the patella

#### ➤ Definition

Repeated, and in some cases very frequent, dislocations occurring as a result of poor dynamic control of the patella.

Habitual dislocation can occur as a result of poor coordination of the muscular control of the patella, although it is much more common in patients with primarily dystonic and slightly atactic disorders than in severely spastic patients. The habitual dislocation of the patella causes



**■ Fig. 3.309.** Knee extension splint as follow-up treatment after lengthening of the knee flexors. The knee flexion position can quickly and simply be adjusted via the strap on the extension rod

rupturing of the medial structures and overstretching of the resulting scar tissue. The lateral ligaments are shortened.

One *treatment* that has proved effective is distalization of the vastus medialis muscle in combination with an extensive lateral release (according to Green), particularly in the cranial direction [18]. As a general rule, the vastus medialis muscle is distalized sufficiently to produce 90° flexion of the knee during the operation, while still ensuring a centered patella. Transfer of the tibial tuberosity is only required in exceptional cases. The *follow-up treatment* requires a cylinder cast for 6 weeks. Functional follow-up treatment is difficult in patients with coordination problems since they tend to lose their footing and can thus tear apart the sutured medial muscles.

### Rotational deformities

#### ➤ Definition

Rotational deformities of the lower leg lead to malpositioning of the foot in relation to the leg axis. Both external and internal rotational deformities can occur.

Rotational deformities of the lower leg, usually increased internal rotation, produce an intoeing gait, although the underlying mechanism of this development is not fully understood. It may be assumed, however, that the increased abnormal loading of the foot, usually in a varus position, regularly produces an internal rotation moment as the adducted foot strikes the ground with the toes and rolls forward on its outer edge. Since the rotational defects are additive (inward rotation of the hip, internal rotation of the lower leg and varus adduction deformity of the foot), this defect often requires correction. As the foot, which is the lever arm for the triceps surae muscle, goes out of alignment with the direction of movement, this essential muscle for posture control becomes insufficient. Consequently, rotational abnormalities are of functional, not just cosmetic, relevance.

The occurrence of rotational deformities can be avoided by keeping the feet facing forwards during walking. Twister cables, elastic strands fitted between a pelvic ring and ankle foot orthoses, can provide functional support. If the twister cables are pretensioned before the ankle footorthoses are fitted (outward rotation for

toeing-in, inward rotation for toeing-out), a torque force acts to correct the foot position. However, these cables cannot correct the underlying skeletal deformity and may exert a problematic rotational force on the knee, which restricts the options for their use. They may help, however, in bridging the period till the surgical correction.

In all cases, the foot skeleton must be placed in orthoses in an anatomically correct position. Any rotational deformities must be accepted or surgically treated. A supramalleolar osteotomy will be sufficient. We prefer fixation with the AO low contact plate (LCP) with screws which provide angular stability, since the patients can start weight-bearing immediately and muscle power and training level are not greatly affected. An abduction flat-foot cannot be left untreated in order to compensate for any internal rotation but must also be corrected.

### 3.3.7.2 Primarily flaccid paralyses of the knee Functional disorders

#### ➤ Definition

Functional changes in the knee with no structural deformity and caused by reduced or absent muscle activity. The knee extensor and flexor mechanisms are particularly affected.

■ Table 3.55 provides an overview of common functional deformities in primarily flaccid locomotor disorders (► Chapter 4.7.4).

A deficit of the knee flexors does not cause significant problems with walking. The swing movement at the knee is elicited with the help of the upper body. Even a deficit of the knee extensors is compatible with minimally restricted walking. By way of compensation, the knee has to be locked in slight hyperextension throughout the stance phase. Consequently, the lower leg should not be allowed to move forward in relation to the foot standing on the ground, which presupposes controlled activity of the triceps surae muscle. If this muscle is also weak, a contracture of this muscle (equinus foot) will fulfill the same purpose. Otherwise, the ankles must be fixed in a slight equinus foot position (backward lean) with a stiff lower leg brace.

■ Table 3.55. Functional deformities in primarily flaccid locomotor disorders

Deformity	Functional benefit	Functional drawbacks	Treatment
Knee extensor insufficiency	–	Standing with flexed knees not possible	Full knee extension
Knee flexor insufficiency	–	Deficient momentum (knee extension contracture)	Passive swinging of the leg during walking

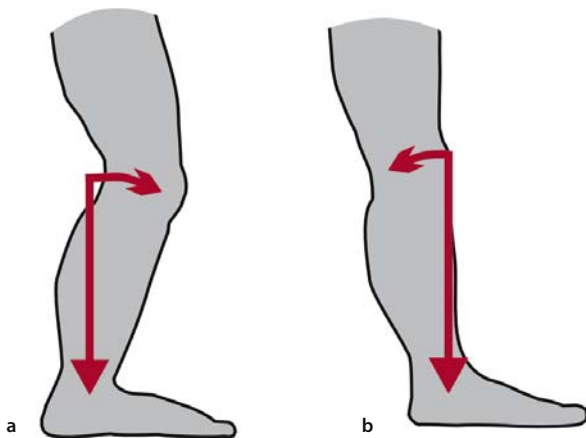
If full knee extension is not possible, the patients have to lean forward with their upper body in order to bring the ground reaction force in front of the knee and thus produce an external extension moment in order to extend the knee indirectly (■ Fig. 3.310). This maneuver transfers at least part of the postural work to the hip extensors. If the upper body has to lean far forward the patients push their arm against the knee to support themselves while walking. Any knee flexion contracture will prevent this compensatory mechanism from coming into play and thus restrict walking ability.

### Structural changes

#### Definition

Structural deformity of the knee caused by reduced or absent muscle activity.

■ Table 3.56 shows the common structural deformities in primarily flaccid locomotor disorders.



■ Fig. 3.310. Schematic view of flexion and extension moments involved in knee extension: a in forward inclination, b in backward inclination

### Knee flexion contractures

#### Definition

Full knee extension is rendered impossible by dorsal capsule and/or muscle shortening.

In flaccid paralyses, full extension (or even slight hyperextension) of the knee is required to compensate for insufficiency of the extensor mechanism (► Chapter 3.3.7.1, »Gait with hyperextension of the knee«). A 5° hyperextension is harmless. If the hyperextension is more pronounced, however, the ligamentous apparatus will be overstretched and eventually give way as a result of lever forces. If extension is lost, the patients have to support themselves by placing a hand above the knee to extend the knee indirectly. Since this maneuver is only possible with a bent upper body, the patients can no longer walk upright or look straight ahead while walking, their hands are no longer free and they develop severe flexion contractures of the hip and knees. Higher neurological levels are more likely to predispose to knee flexion contractures.

Conservative *treatment* is possible with a supporting and stabilizing thigh orthosis with a foot section and designed to lock the knee during the walking. A more attractive and promising alternative for patients is *surgical* lengthening at knee level. Various techniques are available: As any further muscle weakening in these conditions should be avoided the procedure of choice is the supracondylar extension osteotomy. We fix it with an AO LCP-plate with screws with angular stability which enables immediate weight bearing. Muscle or tendon lengthening procedures are also possible but bear the risk of additional weakening.

Full extension can be achieved gradually by using an external fixator attached above the knee (► Chapter 3.3.13). The amount of daily correction can be adapted to match the individual patient's symptoms. One possible complication of such soft tissue lengthening procedures is a temporary painful loosening of the ligamentous ap-

■ Table 3.56. Structural deformities in primarily flaccid locomotor disorders

Deformity	Functional benefit	Functional drawbacks	Treatment
Knee flexion contractures, non-osseous	–	Increased energy required to walk and stand	Soft tissue lengthening procedures, supracondylar extension osteotomy, Ilizarov apparatus in special cases
Knee flexion contractures, osseous	–	Increased energy required to walk and stand	Correction osteotomy
Knee extension contractures	–	Sitting aggravated	VY-plasty of the quadriceps tendon
External rotational deformity of the lower leg	Compensation of increased femoral anteversion	Feet not in direction of leg axis	Correction osteotomy
Internal rotational deformity of the lower leg	(Compensation of abducted pes planovalgus)	Feet not in direction of leg axis	Correction osteotomy

paratus. In some cases, therefore, it may prove necessary to fit a stabilizing support for some time after removal of the fixator to give the ligaments time to restabilize themselves.

### Knee extension contractures

#### ➤ Definition

In this rare deformity the knee extensor mechanism is too short.

This deformity prevents walking and sitting. A VY-plasty of the quadriceps tendon produces good results if physiotherapeutic stretching exercises fail. There is, however, a risk of an active extension lag as a consequence of weakening, which may finally result in a crouch gait.

### Rotational deformities in the lower leg

#### ➤ Definition

The loads produced when rolling from heel to toe and unbalanced muscle forces lead to rotational defects in the lower leg. Both external and internal rotational deformities can occur.

Rotational deformities are common in cases of flaccid paralysis. Patients with flaccid paralyse often require an orthosis to stabilize the feet. Flexible orthoses are of no use since they are unable to prevent talipes calcaneus. Stiff orthoses, on the other hand, make the heel-toe rolling action more difficult, and therefore require the fitting of a rocker to the shoe. If the feet deviate only slightly from the direction of walking, every step produces a torsion moment which, even after the age of 5, can lead to a gradual change in rotation. An external or internal rotational error can require correction if the feet can no longer be positioned in the direction of walking, the efficiency of the gait is impaired or the patients trip over their feet. Occasionally, cosmetic reasons can play a decisive role.

We prefer a supramalleolar correction and use the AO LCP-plate for fixation. Immediate weight bearing is possible after this procedure, and the patients retain their muscle strength. Although orthoses cannot correct the rotational deformities, if functional orthoses are used for other reasons, major deviations in the foot position from the direction of walking must be avoided as much as possible.

### 3.3.7.3 Muscular dystrophy

#### ➤ Definition

Functional and structural problems of the knee associated with myopathies are limited to flexion deformities and contractures that restrict the ability to stand and walk.

■ Tables 3.57 and 3.58 provide an overview of common functional and structural deformities that can occur in muscular dystrophies (► Chapter 4.7.6). Muscular dystrophy patients tend to suffer flexion contractures as they approach the end of their ability to walk. To prolong the ability to stand and walk, some authors have proposed soft tissue lengthening operations as early as 6–8 years of age [18, 19]. Although, in purely statistical terms, such operations can postpone the loss of walking and standing functions [9], in our view patients are not particularly motivated to undergo surgery. We therefore consider the onset of contractures as a compromise for the timing of surgical procedures, even if this potentially involves the loss of the ability to stand and walk for several months. Muscular dystrophy patients sit for longer periods and their muscles undergo fibrous and fatty changes, thus making stretching more difficult. However, since our treatment regimen already ensures a minimal loss of extension at the hips and knees, almost no functional surgery has been carried out in recent years in our patients.

■ Table 3.57. Functional deformities in muscular dystrophies

Deformity	Functional benefit	Functional drawbacks	Treatment
Knee extensor insufficiency	–	Standing with flexed knees not possible	Full knee extension, prevention of pes calcaneus
Knee flexor insufficiency	–	Deficient momentum (knee extension contracture)	Passive swinging of the leg during walking

■ Table 3.58. Structural deformities in muscular dystrophies

Deformity	Functional benefit	Functional drawbacks	Treatment
Flexion contracture at the knee	–	Crouch position (loss of walking and standing)	Lengthening of knee flexors

Since the increasingly restricted muscle power requires passive stabilization of the ligaments for standing and walking, these functions are only possible if the joints can be fully extended. Surgery is therefore required even for fairly small contractions. Purely from the technical standpoint, the knee flexor lengthening procedure must be performed towards the end of the ability to walk, when the muscles are increasingly degenerated, and in such a way that sufficient length can be obtained directly by the operation without the need for protracted follow-up treatment. For this reason, tendon lengthening procedures are preferable to those for the aponeurosis. A loss of muscle power no longer matters much at this stage. Immediate, full mobilization after the operation is part of the follow-up management.

In muscular dystrophy patients in particular, the *equinus foot* represents a key stabilizing component in standing and walking. The slight equinus foot position locks the upper part of the ankle and prevents dorsiflexion. This indirectly extends the knee and the patient is able to hold himself upright passively. Neither orthotic treatment nor surgery is indicated to correct this equinus deformity. On the contrary, the ankles must be stabilized with an orthosis if any free dorsiflexion is present. This brace must be prepared with a slight backward lean in order to stabilize the knee indirectly and thus achieve the same effect as a slight equinus position. In the backward lean position, an extension moment acts on the knee to stabilize the joint in extension. A powerful triceps muscle or a marginal or slight equinus foot is invariably helpful as these can lock the lower leg in relation to the foot and prevent forward movement, thereby helping to produce the indirect extension moment. Severe equinus deformities, however, require correction but only to  $-5^\circ$  to  $0^\circ$  of dorsiflexion.

A slight *hyperextension* of the knee of up to  $5^\circ$  is acceptable, whereas a more pronounced hyperextension can overstretch the knee capsule and lead to later problems in this joint with pain anteriorly. The hyperextension is best prevented indirectly by corresponding orthotic provision for the lower leg and foot, and the forward or backward lean can be adjusted via the heel height. Supports for stabilizing the knee are difficult to produce and hardly ever produce the desired result. Since patients feel uncomfortable during the day with a high-fitting orthosis, a low-fitting orthosis should always be used if possible. They often have unjustified and unproven concerns about possible deformities, e.g. axial deviations in the frontal plane as a result of trunk-swinging movements. The incidence of such problems is not known. For these reasons, it is not useful to provide excessive treatments, but rather to solve problems as and when they occur.

**!** A knee ankle foot orthosis (KAFO) with a freely moveable knee can protect the knee from hyperextension in the stance phase only during knee movement in the sagittal plane.

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### 3.3.8 Meniscal and ligamentous lesions

#### ► Definition

Lesions of the menisci or the ligamentous apparatus of the knee in children and adolescents.

#### Occurrence

Compared to adults, children rarely suffer from lesions of the menisci and/or ligamentous apparatus of the knee. In fact, it used to be thought that such structures could not be injured at all in children. For a long time, the only known internal knee injury in children was tearing of the ligaments at the cartilaginous attachment (particularly the medial collateral ligament and the anterior cruciate as an eminence avulsion) or a meniscal lesion in an existing (lateral) discoid meniscus (► Chapter 3.3.6.6).

The growing popularity, in recent years, of sport and training during childhood and early adolescence has been matched by an increasing number of internal knee lesions that did not used to be seen in this age group. Improved diagnostic techniques have also contributed to this trend: arthroscopy and die MRI scan can now provide a much more accurate diagnosis of the lesion than had been possible in the past. An epidemiological study in Sweden calculated annual incidences of meniscal lesions in children of 7 per 100,000 at the start of the 1960's and 25 per 100,000 children at the start of the 1980's [1]. This is still a low incidence compared to that during the 3rd and 4th decades of life of 90 per 100,000 inhabitants [9].

If an arthroscopy is routinely performed for a hemiarthrosis in a child, a rupture of the anterior cruciate ligament and/or a meniscal lesion will be found, depending on the age group, in 30–40% of cases (children) or 50–60% of cases (adolescents) [16, 27, 28]. Since internal knee lesions in children are difficult to diagnose purely by clinical examination, arthroscopy is often required to correct the clinical diagnosis [3, 16, 27, 28]. The surgical treatment of intraligamentous ruptures of the anterior cruciate ligament while the epiphyseal plates are still open has become a routine treatment in specialist centers over the past 15 years [3, 8, 19]. In our hospital we have observed and treated around 45 such cases over the past 15 years.

#### Diagnosis

##### Recently traumatized knee

##### History

The possibility of a lesion of the internal structures of the knee must always be considered if there is a history of adequate trauma and a subsequent joint effusion. The precise accident mechanism can rarely be reconstructed. Everything usually happens very quickly and children are often unable to recall the precise circumstances. Typical mechanisms are injuries in a flexion/external rotation/valgus position or with a hyperextended knee.

#### Clinical examination

The clinical examination of a recently traumatized knee is painful and should therefore be performed gently. A visual inspection will identify any abrasions or skin injuries. A recent effusion is also usually very readily visible. Although we do not test for the meniscus signs in the acutely injured painful knee, the status of the ligamentous apparatus can be clinically assessed to a certain extent even in a painful knee.

Translation in the AP direction can be investigated by the *Lachman test*, i.e. drawer test with almost full extension: The examiner grasps the thigh with one hand and the lower leg with the other with the knee in 10–20° flexion and tests for the drawer sign anteriorly and posteriorly (► Chapter 3.3.1). As well as assessing the extent of movement, the examiner should also note the quality of the end point (whether »firm« or »soft«). This test must always be performed on the other side for the purposes of comparison. It should be borne in mind that the knee ligaments are more lax generally in children than in adults and that an anterior drawer of up to 10 mm can still be normal [5]. *Medial* and *lateral opening* can also be checked without causing the child too much pain. Here too, the lower leg and thigh are each grasped with one hand and a valgus or varus stress is applied (► Chapter 3.3.1). Note that a slight degree of lateral (but not medial) opening is normal.

However, the pivot shift phenomenon should not be tested in a patient with recent trauma (► Chapter 3.3.1). This test is painful and would not have any consequences at this time. After the knee has been aspirated, we do not perform any more diagnostic procedures, but apply a dorsal plaster cast in approx. 20° flexion. We re-examine the child after two weeks.

#### Radiographic findings

If a hemiarthrosis is present, an *x-ray* should always be recorded to rule out a fracture. Intra-articular fractures most commonly appear as bony avulsions of the anterior cruciate ligament at the intercondylar eminence and, rarely, of the proximal attachment in the intercondylar notch. The other possible intra-articular fractures are described in ► chapter 3.3.9. On the x-ray, the effusion is usually visible as an enlarged soft tissue shadow, otherwise the x-ray does not reveal any specific signs of a lesion of the internal structures of the knee. Meniscal lesions and ligament ruptures cannot be diagnosed on a plain x-ray.

#### Aspirate

A major *joint effusion* should generally be *aspirated*. The following conclusions can be drawn on the basis of the aspirate:

- *Serous effusion*: Not a consequence of a recent internal knee lesion. Either chronic irritation is present (e.g.

### 3.3.8 · Meniscal and ligamentous lesions

due to osteochondritis dissecans, a loose joint body or chronic knee instability), or the patient has juvenile rheumatoid arthritis (► Chapter 3.3.11).

- **Hemarthrosis with fat globules:** The fat globules are an indication of a fracture. The most likely fracture is a bony avulsion of the anterior cruciate ligament at the intercondylar eminence, although other intra-articular fractures are also possible (► Chapter 3.3.9).
- **Hemarthrosis without fat globules:** This finding very probably suggests the presence of a lesion of the internal structures, i.e. a meniscal lesion, a rupture of the medial collateral ligament or an intraligamentous rupture of the anterior cruciate ligament. Another possibility is a recent, traumatic dislocation of the patella with tearing of the retinacula.

#### Knee traumatized 2 weeks or more in the past

##### History

If the trauma occurred slightly further back in the past, the following questions are relevant:

- Does *genuine locking* occur (the knee can neither be flexed nor extended from a particular position)?
- Does *pseudolocking* occur (in a particular position the knee has to overcome an occasionally painful snapping)?
- Does a *giving-way phenomenon* occur (the knee gives way in a particular position during walking)?

##### Clinical examination

After 2 weeks, the acute pain has subsided and the effusion has also usually regressed. The knee can now be examined thoroughly. We proceed according to the following examination protocol (the examination technique is described in detail in ► chapter 3.3.1):

##### Inspection

- knee axis (morphotype) in the standing patient (normal axis, valgus / varus axis with intermalleolar / intercondylar distance),
- knee contours (normal, flattened out, effusion?),
- anterior contours of the knee in the lying patient at approx. 90° flexion (check whether a posterior drawer is present; ► chapter 3.3.1),
- asymmetry of the quadriceps muscles (atrophy on the affected side),
- swelling, redness?

##### Palpation

- palpate the effusion by pressing the suprapatellar pouch and checking for »dancing« of the patella (► Chapter 3.3.1).
- palpate the knee joint space medially and laterally, check whether any painful point migrates posteriorly during increasing flexion (evidence of a meniscal lesion).

- palpate the patellar facets, push the patella in the lateral direction and check whether this causes pain medially (evidence of tearing of the medial retinacula),
- check the temperature, excessive warmth?

##### Stability testing

- Lachman test, i.e. drawer test with almost full extension,
- drawer test in approx. 60° flexion,
- test for lateral opening,
- test for the pivot shift phenomenon [7].

##### Testing for the meniscus signs

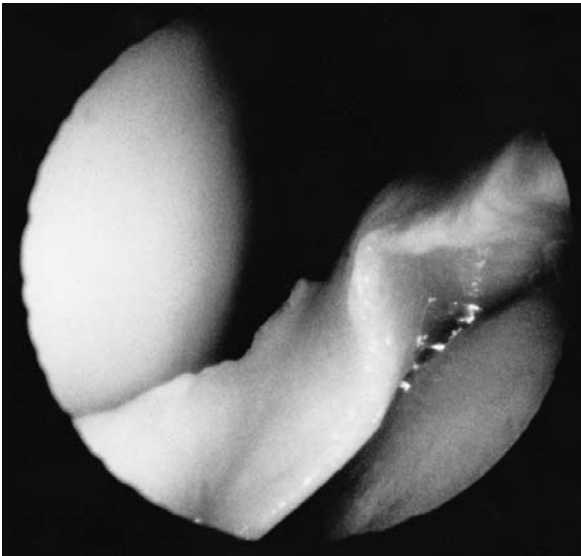
- test whether pain is elicited during internal and external rotation in various flexion positions.

##### Other imaging procedures

The prospect of being able to diagnose internal knee lesions by *ultrasound* raised great hopes, particularly since it is a cheap, painless and non-invasive investigation method. Periarticular structures (collateral ligaments, tendons, bursae) can readily be evaluated [6]. Greater difficulties are posed, however, by the internal structures (particularly the anterior cruciate ligament), although certain experienced investigators appear to be able to evaluate these with a high degree of reliability [25]. An *MRI* scan can provide information about lesions of the menisci and cruciate ligaments in children and adolescents, although it is not as reliable as arthroscopy. Even though certain authors have reported a close correlation between MRI and arthroscopic findings [20], the sensitivity of the MRI scan is generally poor, particularly in children under 12 years of age, and the MRI is no more reliable in terms of diagnostic accuracy than the clinical examination [17, 22]. Furthermore, it is difficult to assess the need for treatment on the basis of MRI findings.

**! The MRI scan frequently shows structural changes within the menisci that are of no clinical significance and yet are readily overrated. Since the MRI scan is an expensive method, the doctor should be extremely cautious in deciding whether this imaging procedure is appropriate. The indication for any need for treatment must be provided by the hospital.**

*Arthroscopy* is much better than MRI for checking the diagnosis and establishing whether surgical treatment is indicated, particularly since any treatment can be administered during the same anesthetic session – whether as an arthroscopic or open procedure. In the hands of an experienced investigator, arthroscopy is by far the most reliable method for diagnosing internal knee lesions (► Fig. 3.311), and it should always be planned with a view to any resulting treatment being provided in the same anesthetic session. The arthroscopic investigation should



■ Fig. 3.311. Arthroscopic photograph of a *bucket-handle lesion* of the medial meniscus in a 14-year old boy with still open epiphyseal plates

always be implemented with the aid of hook probes introduced via a separate surgical approach. Only then can the condition of the posterior horns of the menisci and a partially ruptured anterior cruciate ligament be properly assessed.

### Treatment

The treatment of internal lesions of the knee in children and adolescents with open epiphyseal plates differs to some extent from that for adults. Certain therapeutic methods are unsuitable for children because of the open growth plates, and different factors sometimes need to be taken into account with children in deciding whether a treatment is indicated.

### Medial meniscal lesions

The – fortunately rare – lesions of the medial meniscus in children and adolescents are always very serious. They never occur as flap tears of the inner edge of the meniscal posterior horn as is common in (particularly elderly) adults. Rather, the meniscus either suffers a bucket-handle tear (■ Fig. 3.311) or tearing at the edge. If a bucket-handle lesion occurs, the inner portion of the bucket-handle usually has to be resected, ideally in an arthroscopic procedure. If the lesion is at the edge of the meniscus, the orthopaedist can either await the spontaneous course of events or (for a fairly extensive lesion) perform (arthroscopic) suturing of the meniscus.

### Lateral meniscal lesions

Lateral meniscal lesions in children and adolescents almost always occur in connection with discoid menisci

(see ► chapter 3.3.6 for further details). Only the inner part of the meniscus should be removed, leaving the outer part in place [23]. The lateral meniscus has a very important stabilizing function, and a particularly problematic situation in this context is widening of the popliteal hiatus during resection of the posterior horn. This results in the loss of an important posterolateral stabilizer. Severe instability can result if this occurs in connection with a lesion of the anterior cruciate ligament, posing one of the most difficult therapeutic problems for the knee.

### Medial collateral ligament lesions

Lesions of the medial collateral ligament are likewise not especially rare in children. In many cases, avulsion occurs at the cartilaginous proximal attachment. This cartilaginous portion subsequently ossifies and is visible on the x-ray as a »Stieda-Pellegrini shadow«. Since isolated lesions of the medial collateral ligament have a good prognosis, the treatment should be conservative. It should be noted, however, that a medial opening of more than + (= 5 mm) indicates that the central pillar must also be damaged, i.e. that a concurrent lesion of the anterior cruciate ligament must be present. If these two lesions occur in combination with a medial meniscal lesion this produces the classical »unhappy triad«, which occurs during external rotation-flexion-valgus trauma. Since the same mechanism can also lead to dislocation of the patella, the possibility of this concomitant injury should also be considered (► Chapter 3.3.5). If a combination of such injuries has been diagnosed, the cruciate ligament and possibly the meniscal lesion and dislocation of the patella will require surgical correction, but not the medial collateral ligament rupture.

! **N.B.:** The medial collateral ligament lesion always heals with conservative treatment provided the central pillar is intact. If this is not the case, the central pillar (i.e. the anterior cruciate ligament) must be restored so that the medial collateral ligament can then heal spontaneously. If the anterior cruciate ligament is left in an unstable position, the medial ligamentous apparatus will also be unstable. It will then prove very difficult to stabilize at a later stage.

### Avulsion of the intercondylar eminence

The avulsion of the intercondylar eminence, i.e. the cartilaginous/bony avulsion of the distal attachment of the anterior cruciate ligament is one of the commoner knee injuries in children and adolescents (► Chapter 3.3.9). The condition can be diagnosed on the basis of a plain x-ray, and conservative treatment is sufficient in most cases. If the fragment reduces in hyperextension, a cylinder cast is fitted in this position for 6 weeks. Only if the fragment

cannot be replaced in its original position is (arthroscopic) reduction and refixation required.

### Anterior cruciate ligament lesions

Although, as already mentioned, the intraligamentous rupture of the anterior cruciate ligament is a rare event in patients with open epiphyseal plates, it is occurring with increasing frequency. The following forms are distinguished:

- isolated incomplete rupture (commonest form in the age group under discussion, usually overlooked),
- isolated complete rupture,
- complete rupture with concurrent injuries.

The *isolated incomplete rupture* has the potential to regenerate, as confirmed by our own experiments [10]. When young adults are arthroscopied, changes in the consistency and length of the anterior cruciate ligament are very frequently found without any corresponding rupture. Such patients often cannot recall any corresponding trauma, even though the condition of the cruciate ligament clearly indicates that »something must have happened«. It is assumed that most of these patients had once suffered a partial rupture of the anterior cruciate ligament at some point in their childhood. Many do not subsequently experience any problems with the knee, while others do so only in connection with a further trauma. Symptomatic instability during adolescence, however is rarely observed with this injury. Treatment should therefore be conservative while the Lachman test does not show any significant difference between the two knees and while the pivot shift phenomenon cannot be elicited [18]: cast until the swelling subsides (usually 2 weeks), muscle training.

The *isolated complete rupture* does not have the potential to regenerate [10]. The anterior cruciate ligament is either resorbed or else it sticks to the posterior cruciate ligament. In the latter case, clinical examination reveals an enlarged anterior drawer but with a firm end point. Clinically evident instability is not usually present, however, while the secondary stabilizers are intact. However, the risk of further injuries at a later date is very high [3]. Generally-speaking, the prognosis for rupture of the anterior cruciate ligament in children and adolescents is worse than in adult patients [3], probably because of the increased activity levels of young patients combined with the general ligament laxity and the associated greater »joint play«. Since primary suturing of the anterior cruciate ligament has a poor prognosis we recommend the following course of action: After the primary trauma, the effusion is aspirated and a dorsal cast applied for 2 weeks. The knee is then clinically investigated.

If a rupture of the anterior cruciate ligament is suspected, the bone age is determined and an arthroscopy

performed. If an isolated rupture of the anterior cruciate ligament is diagnosed, the procedure will depend on the bone age. If this is older than 12 years in boys or 10 1/2 years in girls, the anterior cruciate ligament is reconstructed using the technique for adults.

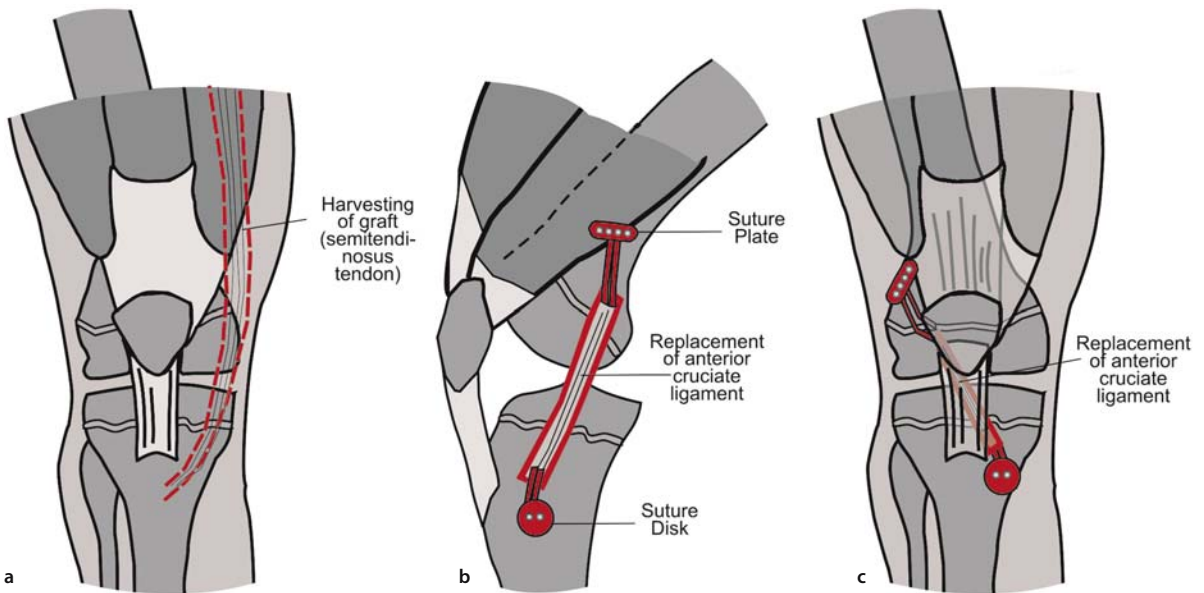
For this we use the middle section of the quadriceps tendon with a bone fragment from the proximal patella. In younger patients we use the semitendinosus tendon. The graft is sutured and pretensioned 3 times using a special device and then pulled from the medial side of the tibial tuberosity centrally through the epiphyseal plate to the eminence and fixed to the lateral aspect of the intercondylar notch with transosseous sutures. To this end, a Position Suture Plate is used on the femur and a Position Suture Disk on the tibia. The operation is performed as an arthroscopically assisted procedure (■ Fig. 3.312, 3.313).

To avoid the subsequent occurrence of any growth disorder, the drilled channel should not be any wider than 10 mm nor positioned eccentrically in the tibia, no bone fragments should be present in the plates and the fixation components should not bridge the plates [19]. Provided these requirements are satisfied, no significant growth disorder will occur. The middle third of the patellar tendon can be used only if the plates are fully closed, since the apophyseal plate of the tibial tuberosity must not be damaged. The semitendinosus tendon may be used as an alternative to the quadriceps tendon, although it produces a slightly weaker graft.

If a *complete rupture with concurrent injuries* occurs, the conservative approach to cruciate ligament ruptures cannot be maintained. Fortunately, such injuries are extremely rare in children. As already mentioned in the section on medial collateral ligament lesion, the concurrent injuries can only heal if the central pillar remains intact. Top priority should therefore be given to restoring the stability of the anterior cruciate ligament. A conventional reconstruction of the anterior cruciate ligament should not be considered if the bone age is less than 13 years in boys or 11 1/2 years in girls. Suturing of the anterior cruciate ligament is also inappropriate, since the remnants of this ligament become necrotic and cannot usually form vital bridges. We therefore perform an anterior cruciate ligament reconstruction according to the technique described above [8].

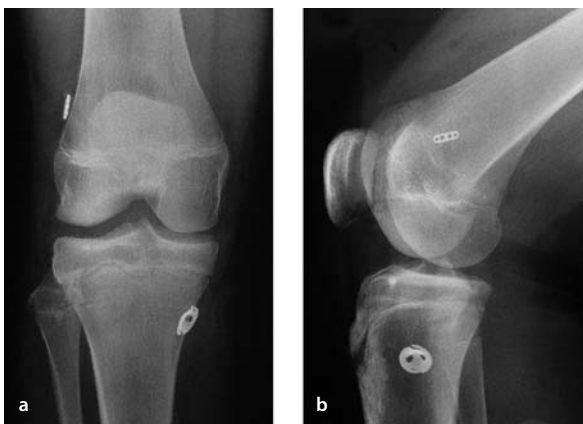
Recent investigations with relatively large sample groups of 60 children and adolescents in each case showed that no growth disorders occurred after a conventional surgical technique with the drilling of holes through the epiphyseal plate, and that the results were significantly better than after conservative treatment [2, 21]. As regards concurrent injuries, meniscal lesions must always be repaired and ruptures of the lateral structures sutured, whereas lesions on the medial side do not require surgical treatment after stabilization of the central pillar.





**Fig. 3.312a–c.** Technique of *anterior cruciate ligament reconstruction* for patients with open epiphyseal plates using the semitendinosus tendon. **a** The tendon is removed using a »tendon stripper« and folded into 3 layers, pretensioned with a tensioning device and sutured with anchoring sutures. **b** Anchoring channels are drilled in the tibia and femur, and the graft is pulled through in a distal to proximal direction.

The sutures are brought out transcutaneously onto the surface of the lower leg and the »suture plate« can be tilted by pulling on the sutures, causing it to cling to the femur. The surgeon now pulls distally on the anchoring sutures, adjusting the precise degree of tension under arthroscopic control, and secures the sutures to the tibia with the »suture disk« (c)



**Fig. 3.313 a,b.** Right knee of a 12-year old girl after *anterior cruciate ligament reconstruction* with open epiphyseal plates using the semitendinosus tendon. AP (a) and lateral (b) x-rays

**!** The following should be noted in respect of the timing of the surgical treatment of knee ligament injuries: The operation should not be performed during the phase of acute trauma reaction, i.e. during the first 2 weeks after the trauma. The risk of arthrofibrosis is much higher during this early phase [24].

### Our therapeutic strategy for anterior cruciate ligament lesions

Our therapeutic strategy for anterior cruciate ligament lesions is shown in **Table 3.59**.

#### Postoperative management

In the immediate postoperative period we use a dynamic splint. Full weight-bearing is permitted from the start. No restrictions are imposed on passive flexion and extension. Intensive isometric muscle training exercises are performed for 6 weeks. Active extension between 20° and 0° is only permitted after 6 weeks.

#### Posterior cruciate ligament lesions

Lesions of the posterior cruciate ligament have even more serious consequences than those of the anterior cruciate ligament. While anterior instability jeopardizes sporting ability, posterior instability can adversely affect the patient's ability to walk, since posterior subluxation of the tibia occurs with every step. Fortunately, ruptures of the posterior cruciate ligament are very rare in children and adolescents with open epiphyseal plates.

From bone ages of 14 years in boys and 12 1/2 years in girls, the approach adopted for adults may be employed, although the prognosis for treatment of an injured posterior cruciate ligament is not good in adults either [4, 13]. The surgical treatment is very difficult



■ Table 3.59. Therapeutic strategy for lesions of the anterior cruciate ligament

Type of injury	Bone age <11 ½ years for a girl or 13 years for a boy	Bone age >11 ½ years for a girl and 13 years for a boy
Partial	Conservative (cast for 2 weeks, muscle training)	Conservative (cast for 2 weeks, muscle training)
Complete, isolated	Conservative (cast for 2 weeks, muscle training), re-evaluation on completion of growth	Cast for 2 weeks, followed by conventional cruciate ligament reconstruction with patellar tendon
Complete, with concomitant injuries	Cruciate ligament reconstruction or strengthening of the ligament residues with semitendinosus tendon	Cast for 2 weeks, followed by conventional cruciate ligament reconstruction with patellar tendon

and requires considerable experience. It is also doubtful whether surgery produces better results than conservative treatment when the injury occurs in isolation [25]. If (for a complex injury) an operation is indicated for these adolescents, we recommend referral to an experienced knee surgeon who is regularly confronted with these problems.

### Prognosis

While countless published articles have addressed the prognosis of internal knee injuries, only a few have concerned themselves with these injuries in children and adolescents [2, 3, 18]. While the prevailing view in the 1970's and 80's favored surgical treatment as a matter of course for adults with cruciate ligament and meniscus injuries, the approach nowadays tends to be more discriminating. Surgery is indicated particularly for injuries in the group of younger, active patients under 40 years of age, especially if complex injuries are involved. Isolated ruptures of the anterior cruciate ligament are, as a rule, treated by surgery only in patients who are very actively

involved in sports. Long-term studies have not been able to show, convincingly, that surgery prevents the development of osteoarthritis.

Although the joint will spontaneously stabilize with the onset of osteoarthritis, there is no doubt that young patients who are actively involved in sports will benefit from surgical stabilization. Investigations have shown that patients who suffer their injury in childhood or adolescence subsequently experience instability problems [2, 3, 14, 15]. This is probably attributable to the fact that adolescents tend to subject their knees to excessive loads, resulting in lesions of the secondary stabilizers at an early stage. They also have greater »joint play«. While a very cautious approach to surgery is appropriate while the patient is still growing and an operation should only be performed for complex injuries, the patient's situation and the possibility of surgery should always be re-evaluated on completion of growth.

The evaluation system proposed by the »International Knee Documentation Committee« (IKDC), which was developed by 20 knee surgeons in Europe and America, should be used for assessing the therapeutic results of knee ligament-stabilizing procedures [11, 12]. This system ensures that the evaluation is conducted according to uniform criteria.

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The prognosis for knee ligament lesions is worse in children and adolescents than in adults because of the greater joint play

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### 3.3.9 Fractures of the knee and lower leg

C. Hasler

#### 3.3.9.1 Fractures of the distal femur

##### Occurrence

Fractures of the distal femur account for approx. 0.5% of all fractures during growth.

##### Diagnosis

##### Clinical features

Epiphysiolyse and epiphyseal fractures usually result in hemarthrosis. Concomitant neurovascular lesions occur primarily in epiphysiolyse after hyperextension trauma. They must also be ruled out in apparently harmless fractures with slight displacement. The same applies to rotation deformities. Concurrent ligament lesions must also be sought specifically at operation or after conservative treatment (after consolidation) as these can be expected in just under 10% of cases [8].

##### Imaging investigations

Standard AP and lateral x-rays. If epiphyseal fractures are suspected, views recorded in 45° internal and external rotation are helpful, particularly for identifying posterior condylar fractures. For the rare complex fracture patterns (e.g. for transitional, type II triplane fractures), a CT scan with 3D reconstruction will facilitate the surgical planning.

##### Fracture types

*Compression fractures* involve stable bulging of both cortices in the supracondylar area.

*Complete metaphyseal fractures* occur increasingly in connection with osteoporosis and/or restricted movement of the hip and knee, e.g. in children with cerebral palsies [2, 17] (■ Fig. 3.314).

*Epiphyseal separations* (Salter types I and II) represent the commonest distal femoral fractures, predominantly



## 3.3.9 · Fractures of the knee and lower leg

in the form of detachments with a metaphyseal, usually lateral, wedge (type II; ■ Fig. 3.314). They occur particularly during puberty [8]. In undisplaced type I separations, the key diagnostic pointers are local swelling, pain, slight physeal widening on the x-ray and the periosteal reaction on the consolidation x-ray. Even if the fractures only appear slightly displaced on the x-ray, the possibility of substantial displacement of the fragments at the time of the trauma with correspondingly underestimated soft tissue damage should be borne in mind. Displacements occur primarily in the varus or valgus direction. Although epiphyseal separations more commonly occur in adolescents, isolated cases are also seen in connection with birth trauma or in toddlers in connection with child abuse.

*Epiphyseal fractures* (Salter types III and IV) primarily occur as a result of valgus traumas during sport (■ Fig. 3.315). The epiphyseal fracture line tends to pass through the intercondylar notch rather than the condyles themselves.

*Transitional fractures* of the distal femur are far rarer than those of the distal tibia. These are usually in the form

of triplane fractures with the fracture lines running vertically through the epiphysis, horizontally through the still open part of the growth plate and from the distal anterior to the proximal posterior part of the metaphysis.

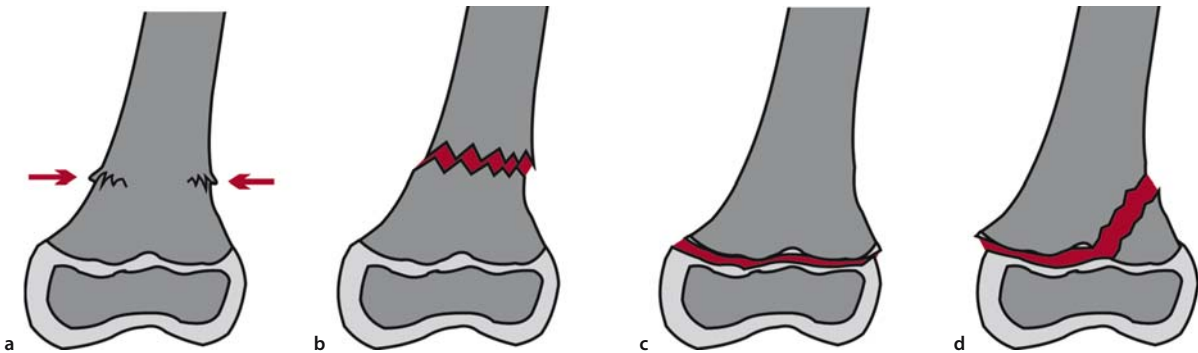
The very rare bony *avulsions of the lateral collateral ligament* are only discernible on x-rays by a fine bone flake at the level of the growth plate on the lateral side. Small, posttraumatic peripheral physeal bridges rapidly lead to a valgus deformity [10].

### Treatment

We *aspirate the hemarthrosis* for analgesic purposes only if the knee is painfully swollen.

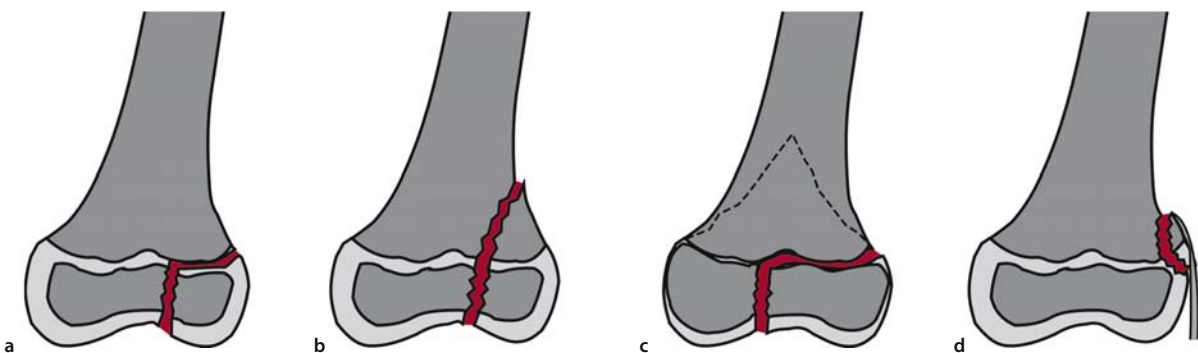
### Spontaneous corrections

As with shaft fractures, axial deviations should not be left uncorrected. *Limit of tolerance for anterior bowing, posterior bowing*: 15–20° in under 10-year olds, decreasing thereafter. Also as with shaft fractures, *valgus and varus deformities* can correct themselves spontaneously to a slight extent in younger children.



■ Fig. 3.314a–d. Metaphyseal fractures of the distal femur: Metaphyseal compression fractures (a), complete metaphyseal fracture (b),

epiphyseal separations without (Salter I; c) and with (d) metaphyseal wedge (Salter II)



■ Fig. 3.315a–d. Epiphyseal fractures of the distal femur: Epiphyseal fracture without (Salter III; a) and with metaphyseal wedge (Salter IV;

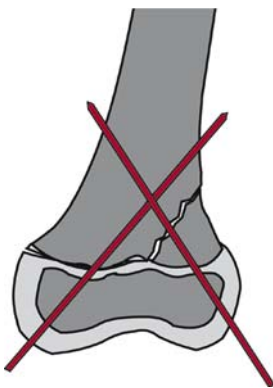
b), Transitional fracture (»triplane fracture«; c) and bony avulsion of the collateral ligament (d)

### Conservative treatment

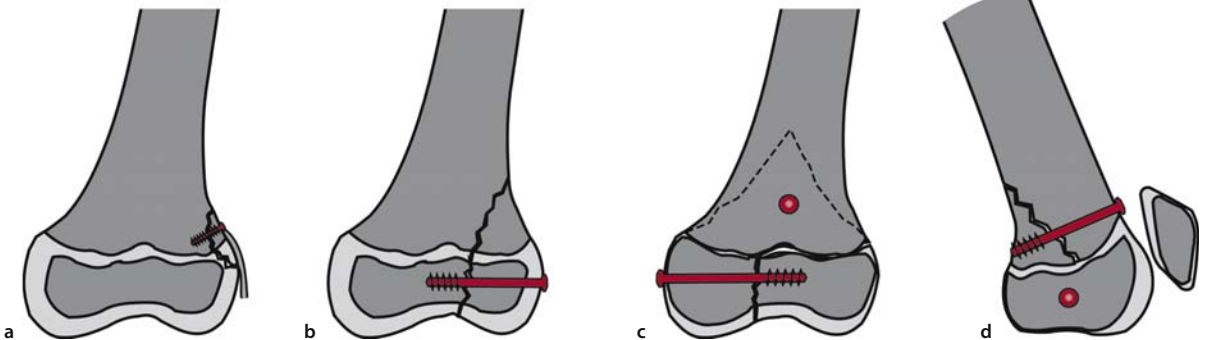
*Non-displaced fractures* are immobilized in a plaster slab, which is replaced by an encircling cast after the swelling has subsided. The position of fractures at risk of displacement are checked one week after the trauma. Primary or secondary axial deviations up to 10° are wedged after 7–10 days.

### Surgical treatment

*Supracondylar fractures* and *epiphysiolyse*s with axial and/or rotational deformities are reduced under anesthesia. Hyperextension deformities are ideally reduced with the hip and knee in flexion. After each reduction, the result is fixed with percutaneous, crossed Kirschner wires because of the significant risk of secondary displacement (■ Fig. 3.316) and immobilized in a cast with cut-outs around the wires. If a very large metaphyseal wedge is present, the internal fixation can also be performed using only percutaneously inserted metaphyseal lag screws (■ Fig. 3.317a) and, depending on the stability in the individual case, subsequently treated without a cast.



■ Fig. 3.316. Treatment of displaced metaphyseal fractures of the distal femur: After closed reduction, these fractures can be secured definitively with percutaneous crossed Kirschner wires. The wires should be trimmed above skin level so that they can then be removed at a later date without anesthesia



■ Fig. 3.317a–d. Treatment of displaced epiphyseal fractures of the distal femur: Since these are joint fractures, they must be reduced openly in the exact anatomical position if dehiscence or a step is present. The

Open reduction to overcome a soft tissue interposition is rarely indicated.

*Intra-articular fractures* with steps or dehiscences exceeding 2 mm are openly reduced, stabilized with internal fixation and functionally treated postoperatively (■ Fig. 3.317).

### Duration of immobilization and consolidation

Depending on the patient's age and the fracture, this period ranges from 2 weeks for compression fractures to 5 weeks in adolescents.

### Follow-up management and controls

The displacement risk of unstable fractures without internal fixation justifies a visual radiological control after 7–10 days.

Patients with epiphyseal separations/fractures are followed up for at least 2 years after the trauma because of the risk of growth disorders. Since the risk of a clinically relevant growth disorder is negligible after transitional fractures given the imminence of physeal closure, the completion of treatment is based on the restoration of free function and freedom from symptoms.

### Complications

*Growth disturbances and posttraumatic deformities* are described in up to 50% of cases after fractures involving the distal femoral growth plate, whether epiphyseal separations or actual fractures. Persisting or incompletely remodeled malpositions or partially inhibiting growth disturbance resulting from physeal bridges with secondary deviations are possible causes. Their clinical impact depends on the amount of residual growth, the location and the extent of posttraumatic ossification of the growth plate.

The probable vascular origin of the bridge formation after epiphyseal separations or after local physeal destruction in epiphyseal fractures explains why an immediate anatomical and gentle reduction, possibly supplemented by compression osteosynthesis, cannot reliably prevent

result is stabilized by small-fragment screws running parallel to the growth plate

### 3.3.9 · Fractures of the knee and lower leg

bridge formation, hence the importance of early briefing of the parents and follow-up controls until the completion of growth.

#### 3.3.9.2 Patellar fractures

##### Occurrence

A high proportion of shock-absorbing cartilage, the substantial mobility of the patella and the relatively weak extensor muscles explain the rareness of patella fractures during childhood and adolescence.

##### Diagnosis

##### Clinical features


Pain, swelling, inability to weight-bear, possibly high-riding patella, hemarthrosis.

##### Imaging investigations

AP, lateral and axial x-rays.

Ossification of the cartilaginous structures occurs between the ages of 3 and 6, frequently initiating from several, irregularly defined centers and continuing up until around 10–12 years of age. Differentiating between acute traumatic changes and ossification variants or a bipartite patella can pose problems. The *bipartite patella* shows rounded edges on the non-fused, superolateral center.

##### Fracture types

The fracture types are presented in  Fig. 3.318.

*Fractures of the tip of the patella* are commonest and typically the result of sudden, forced contractions of the

quadriceps, e.g. on landing. The extent of the injury is usually underestimated on x-rays as substantial parts of the fragment consist of cartilage and periosteum. Typically, small tip fragments and a high-riding patella are present. In younger children, the periosteum anteriorly and the cartilage posteriorly are pulled from the tip of the patella like a glove (*periosteal sleeve fracture*). *Differential diagnosis*: Sinding-Larsen-Johansson disease, chronic tendinitis due to repetitive stress, usually in patients who are very actively involved in sports. Similar radiographic findings are observed in children with a permanently high degree of quadriceps tension, e.g. in cerebral palsy patients.

*Transverse fractures*, the consequences of direct trauma, are best viewed on the lateral x-ray. Complete fractures show dehiscence as a result of the quadriceps tension.

*Vertical fractures*, detected on an axial x-ray. Incongruity is generally present.

*Comminuted fractures*.

*Avulsions of the superior pole* are very rare events.

*Medial retinacular tears, osteochondral fractures* after lateral dislocations of the patella.

*Stress fractures* are usually transverse and occur in athletes or patients with cerebral palsy or spina bifida [2].

##### Treatment

##### Conservative treatment

All undisplaced fractures. Aspiration of the hemarthrosis if this is very pronounced and thus painful. Cylinder cast and partial weight-bearing on crutches for 4–5 weeks.

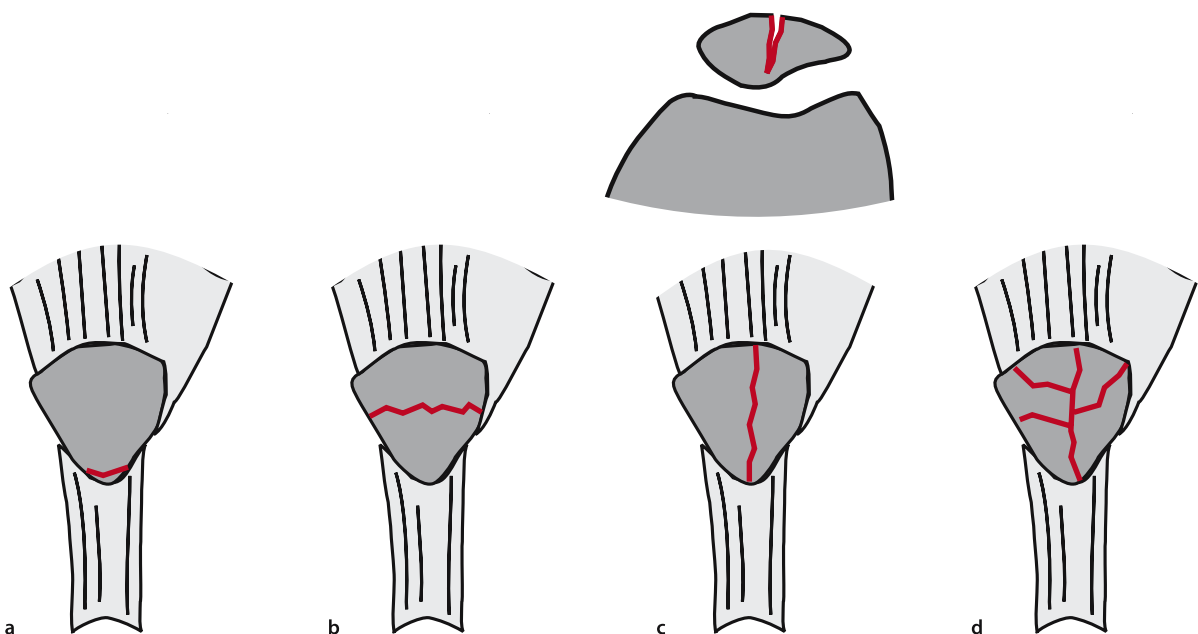


 Fig. 3.318a–d. Patellar fractures. a Fracture of the tip of the patella. b Transverse fracture. c Vertical fracture. d Comminuted fracture



### Surgical treatment

All displaced fractures (>2 mm dehiscence and/or step). Depending on the type of fracture, the classical procedure of tension-band wiring, miniscrew fixation or bone sutures can be used. A total patellectomy should be avoided where possible, since a significant loss of power can be expected from the subsequent loss of the fulcrum. Early postoperative functional treatment on the passive motorized splint (Kinetek) with a gradual, fracture-related build-up of the range of motion, e.g. 0–30°, 30–60° and 60–90° in 2-week intervals. Isometric quadriceps training starts in the immediate postoperative period.

### Follow-up controls

Consolidation x-ray after 5 weeks. Metal removal after around six months.

### Complications

*Growth disturbances* are not known. *Patella alta and calcification of the patellar ligament* can occur after overlooked or inadequately refixed inferior pole avulsions [1]. *Femoropatellar arthrosis* after fractures with incongruent consolidation or after partial patellectomy are possible [11]. *Pseudarthrosis* rarely occurs.

#### 3.3.9.3 Fractures of the proximal tibia

##### Occurrence

Fractures in the area of the proximal tibial epiphysis and metaphysis are rare, accounting for less than 1% of all childhood fractures. This is primarily attributable to the fact that this region is mechanically well protected: laterally by the fibula, ventrally by the tuberosity and patellar ligament, medially by the growth plate-bridging medial collateral ligament and posteromedially by the attachment of the semimembranosus muscle.

The growth plate of the proximal tibia projects anteriorly like a tongue into that below the tibial tuberosity. This plate section is exposed to traction forces produced by the patellar ligament, which is inserted at this point, and can thus be considered as an apophysis from the functional standpoint. In physiological respects, these anterior sections are the last to undergo physal closure towards the end of growth.

##### Diagnosis

##### Clinical features

The tibial head (and thus the proximal epiphysis and metaphysis) is readily inspected and palpated, at least in its anterior sections, thanks to the thin soft tissue covering. Epiphyseal fractures usually lead to hemarthrosis, while metaphyseal fractures, in contrast with the corresponding lesions of the distal femur, occur outside the joint and therefore do not produce any joint effusion,

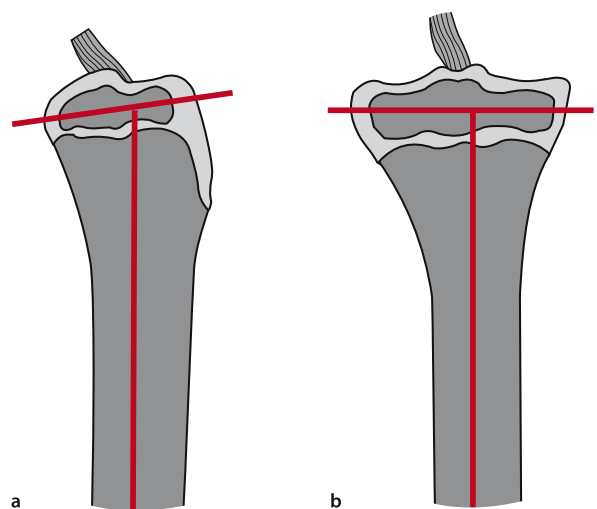
although pronounced local swelling is usually present. A high-standing patella indicates a displaced avulsion of the tibial tuberosity.

After an epiphyseal separation, the doctor in charge must check for any compression of the popliteal neurovascular bundle (peroneal nerve). Moreover, fractures that appear only slightly displaced on the x-ray may induce a false sense of security since substantial displacement with spontaneous reduction may have occurred at the time of the trauma.

### Imaging investigations

Standard AP and lateral x-rays should be supplemented by views in 45° external and internal rotation if epiphyseal fractures are suspected. More complex fracture patterns, e.g. in transitional fractures, are preferably visualized by 3D-reconstructed CT scans, which facilitate more accurate planning of the surgical approach and technique. One essential requirement is the radiological recording of deformities, particularly varus or valgus deformities, as experience has shown that these are poorly corrected by growth. The requirement that the shaft of the lower leg should be perpendicular to the tibial plateau can easily be checked by the angle of the epiphyseal axis (■ Fig. 3.319). On the lateral view, the tibial plateau normally slopes in the posterior direction at an angle of approx. 10–15° (tibial slope).

In the early stages, fatigue fractures often show only minimal radiological signs in the area of the postero-medial cortex of the proximal tibia and local cortical thickening and lamellar periosteal reactions. A striking finding on the bone scan is local hyperactivity, while the CT scan shows perifocal edema in the soft tissue



■ Fig. 3.319a, b. Tibial axes. a On the lateral view the tibial plateau slopes backward by approx. 10°. b On the AP view, the epiphysis-axis angle should be 90°

## 3.3.9 · Fractures of the knee and lower leg

window and periosteal and endosteal callus in the bone window [5].

**Fracture types**

Fractures of the *intercondylar eminence* correspond to bony avulsions of the distal anterior cruciate ligament insertion, and typically occur in the 8 to 12 year age group, while the growth plates are still wide open [18]. The accident mechanism in such cases corresponds to that of an anterior cruciate ligament lesion in adults, with hemarthrosis and increased anterior tibial translation in the Lachman test. The latter is the most sensitive test for anterior cruciate ligament lesions: The anterior translation of the tibia is tested in the supine patient at approx. 20° knee flexion. The ligaments should not be tested, however, after recent trauma, partly because this is a painful procedure and partly because guarding in children will usually produce an inconclusive outcome. The following degrees of displacement are differentiated [14]:

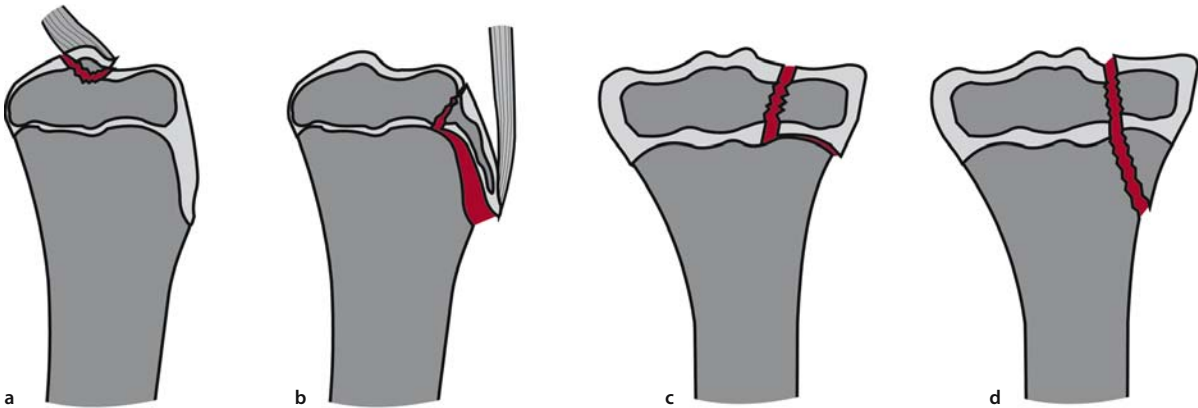
- *type I*: No displacement.
- *type II*: The fragment is elevated anteriorly like a tongue the posterior hinged part is still in contact with the tibial plateau.

— *type III*: Complete displacement. Often the avulsed fragment appears comminuted, a situation described as a type IV fracture, as a supplement to the original classification of Meyers and McKeever. The size is frequently underestimated on the x-ray because of the cartilaginous section.

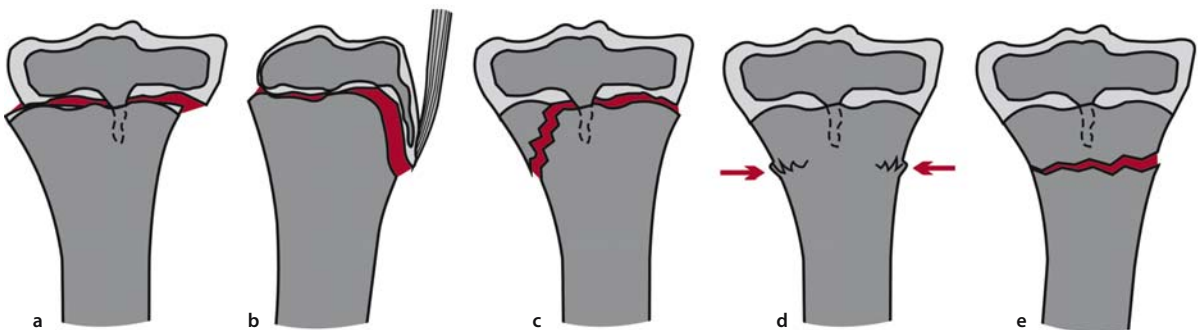
*Epiphyseal fractures (Salter-Harris types III and IV – chapter 4.1 – and transitional fractures)* are even rarer than those of the distal femur [3] (■ Fig. 3.320). They occur predominantly during adolescence. Concomitant ligament lesions or menisci trapped in the fracture gap are often identified only secondarily or during surgical management of a fracture [7].

*Compression fractures* are stable, are not associated with any misalignment and heal without complications. They primarily affect younger children.

*Epiphyseal separations (Salter-Harris types I and II)* are the result of indirect valgus forces or hyperextension traumas. The latter produce anterior displacement of the epiphysis, including the tuberosity. As a consequence, the now prominent metaphysis may compromise the popliteal artery on the posterior side (■ Fig. 3.321).



■ Fig. 3.320a–d. *Epiphyseal fractures of the proximal tibia*: Avulsion of the intercondylar eminence (a), avulsion of the tibial tuberosity (b), epiphyseal fracture without (Salter III; c) and with (Salter IV; d) metaphyseal wedge



■ Fig. 3.321a–e. *Metaphyseal fractures of the proximal tibia*: epiphyseal separation without (Salter I; a – lateral view in b) and with (Salter II; c) metaphyseal wedge. Metaphyseal compression (d) and bowing fracture (e). The tibial tuberosity is part of the epiphysis and is also detached during epiphyseal separations

*Bowing fractures* after valgus traumas without physal involvement can essentially be divided into two fracture types:

- complete fracture of the proximal tibia and fibula,
- metaphyseal greenstick fracture with curvature of the lateral cortex and fracture of the medial cortex, with or without fibular involvement.

*Metaphyseal bowing fractures of the proximal tibia:*

The initial valgus deformity is usually so slight that it is easily overlooked if there is no consistent check for split fractured sections on the medial side and the axial relationships on the x-ray are not measured. If the initial deformity is left untreated there is a high risk of a progressive valgus deformity (see below for complications; ■ Fig. 3.322)

*Avulsions of the tibial tuberosity* typically affect adolescent athletes after a sudden, strong quadriceps contraction or forced knee flexion while the quadriceps is activated, e.g. on landing [13]. Extra-articular avulsions are considered to be displaced if there is more than 5 mm of elevation, whereas the corresponding threshold for intra-articular (Salter-Harris type III) fractures is 2 mm. In some cases, a soft tissue injury with distal, limited bony avulsion of the patellar ligament predominates.

*Fatigue fractures* should be considered in the differential diagnosis if there is no history of trauma and chronic activity-related pain is typically present. They can usually be differentiated from malignant tumors even on the basis of this history (rest or night pain). Unaccustomed loads in untrained individuals or frequent, excessive training sessions in those with high sporting ambitions are triggering factors. Young female endurance athletes should be questioned specifically about a possible female athlete triad: anorexia, osteoporosis and amenorrhea are the key ele-

ments, and these may be accompanied by anemia, fatigue, depression, cold intolerance and a lack of concentration.

### Treatment

*Spontaneous corrections* of valgus and varus deformities are unreliable and should therefore not be considered as part of the primary treatment [19]. Usually there is no genuine correction, rather the proximal and distal epiphysis realign themselves horizontally, while the deformity in the shaft grows, resulting overall in an S-shaped deformity. Top priority is accorded therefore to the elimination of all primary valgus and varus deviations. In patients younger than 10 years old, any (rare) deformities of up to around 20° in the sagittal plane can be left to correct themselves spontaneously.

### Conservative treatment

*Non-displaced fractures:* Initial immobilization in a plaster slab, replaced by an encircling cast after the swelling has subsided, usually after a few days. Stable fractures (e.g. compression fractures) do not require a positional check. If a positional check of unstable fractures after approx. 7 days reveals a primary or secondary varus or, more frequently, valgus deformity of up to 10°, then cast wedging can be tried.

*Eminence fractures:* Aspiration of the hemarthrosis if this is very pronounced and painful. In all cases a cylinder cast is applied for 4 weeks.

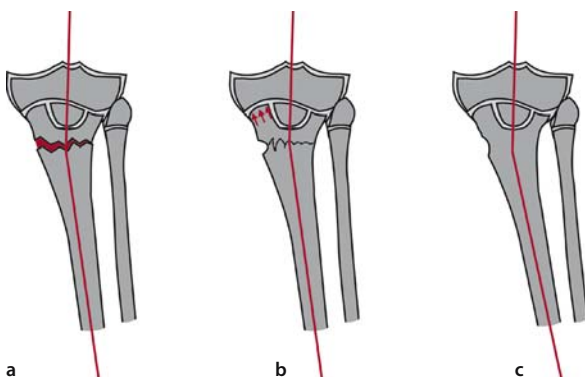
- *Type I:* Cast immobilization in extension [9].

### Surgical treatment

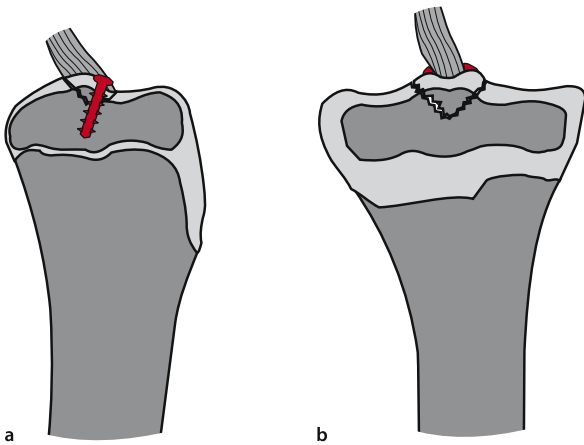
*Eminence fractures:*

- *Type II:* The fragment can usually be reduced by closed manipulation with knee extension under image intensifier control. Failure to produce a complete reduction may be due to interposition of the anterior horn of the lateral meniscus or, more commonly, the transverse genicular ligament, which can be freed by arthroscopy [9] (■ Fig. 3.323).
- *Type III:* Limited anteromedial arthrotomy or arthroscopically assisted refixation, e.g. with antero-grade, cannulated screws that do not cross the growth plates [6]. For multifragment avulsions, sutures may be inserted in the distal part of the cruciate ligament, which are then passed distally through 2 small holes drilled in the tibia and knotted over the proximal tibia. A slight lowering of the avulsed fragment below the level of the surrounding cartilage compensates for the plastic elongation of the ligament.

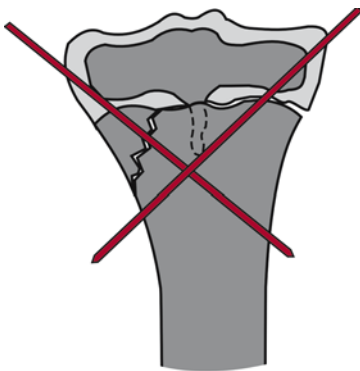
*Metaphyseal fractures:* If the deformity is initially more than 10°, or if the lower leg axis cannot be anatomically corrected after cast wedging, closed reduction is tried and the result fixed percutaneously with crossed Kirschner wires (■ Fig. 3.324). If the fracture cannot be reduced



■ Fig. 3.322a–c. Growth disorder after metaphyseal bowing fracture of the proximal tibia: By definition, bowing fractures of the proximal tibia involve an axial deviation of varying severity, usually a valgus deformity, which is reflected in the gaping fracture gap on the medial side. If this is left untreated, medial consolidation will be delayed, causing increased stimulation of the medial plate and, in turn, increased medial growth. The already existing valgus deformity will be exacerbated, resulting in a unilateral genu valgum



■ Fig. 3.323a, b. Treatment of displaced eminence fractures: All patients with a displaced eminence fracture should be investigated arthroscopically for concurrent injuries. The eminence itself should be reduced arthroscopically and, wherever possible fixed by an epiphyseal screw (a). If this cannot be performed by arthroscopy, the fragment is resecured with a sturdy intraepiphyseal Dexon suture via an arthrotomy (b)



■ Fig. 3.324. Treatment of displaced epiphyseal separations of the proximal tibia: These fractures are managed by closed reduction and stabilized with percutaneously inserted, crossed Kirschner wires

manually, a soft tissue interposition is usually present and can either be freed openly or compressed using a small external fixator (■ Fig. 3.325, 3.326).

For *epiphyseal fractures*, the guidelines for the management of joint fractures apply (■ Fig. 3.327).

*Displaced tuberosity avulsions* frequently involve an interposed periosteal flap. After the flap is freed, the fracture is reduced, with the knee extended, and the result fixed by lag screw osteosynthesis. Predominantly periosteal avulsions of the patellar ligament can be managed by bone sutures, secured if necessary by tension-band wiring [4] (■ Fig. 3.328).

#### Duration of immobilization

Three weeks for compression fractures, 4–5 weeks for the other fractures. After stable internal fixation, a splint is worn until the swelling subsides and the wound has healed. Mobilization can then begin immediately on the motorized splint.

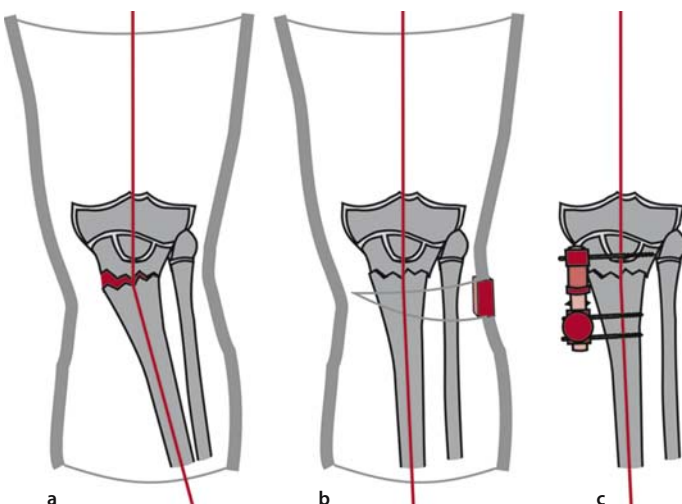
#### Follow-up controls

Positional controls after 7–10 days for fractures at risk of displacement. Because of the potential risk of growth disturbances, subsequent controls are justified for at least 2 years following trauma while the growth plates are still open, excluding compression fractures. If movement is restricted and/or axial asymmetry is present, the patient is monitored until physal closure occurs.

#### Complications

##### Growth disturbances and posttraumatic deformities

*Partial growth plate closure* is a possible complication of an epiphyseal fracture, but can also occur after epiphyseal separations that often appear trivial on the x-ray, even after correct primary treatment. Parents and patients should be informed of this possibility even at the time of fracture treatment if more than 1–2 years of residual

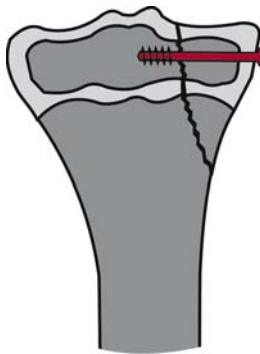


■ Fig. 3.325a–c. Treatment of metaphyseal bowing fractures of the proximal tibia: By definition, every metaphyseal bowing fracture involves an axial deviation, usually a valgus deformity, which is reflected in the gaping fracture gap on the medial side (a). If the primary valgus can be eliminated and the medial fracture gap compressed by cast wedging (b), the consequences of the increased medial growth will not be clinically significant. If the medial fracture cannot be closed by cast wedging, the fracture must be managed by closed reduction and the resulting position secured ideally with a medially fitted external fixator (c). The surgeon must be careful to avoid injury to the apophyseal plate

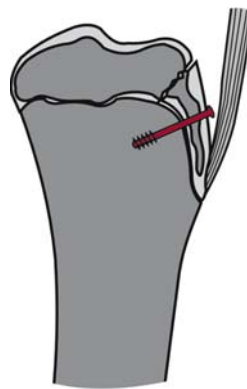


**Fig. 3.326.** Treatment of a compression fracture of the proximal tibial metaphysis: 4-year old girl with metaphyseal bowing fracture which, by definition, shows a moderate valgus deformity (reflected in the medially gaping fracture gap). The valgus deformity was eliminated by cast

wedging, but the medial fracture gap does not initially appear to be compressed. During consolidation, however, the medial fracture gap is bridged by callus. The subsequent increased medial growth only resulted in a moderate, clinically insignificant increase in the valgus position



**Fig. 3.327.** Treatment of displaced »typical« epiphyseal fractures, Saltzer types III and IV, of the proximal tibia: Since these are joint fractures, they must be reduced openly in the exact anatomical position and stabilized with a small-fragment screw running parallel to the growth plate



**Fig. 3.328.** Treatment of displaced tuberosity avulsion: Intra- and extra-articular tuberosity avulsions are reduced in the exact anatomical position and refixed with a large-fragment screw

growth is expected. Depending on the size and location of the closure, this can lead to abnormal axial growth, e.g. with a recurvated knee after tuberosity avulsions or eminence fractures with screw placement through the anterior growth plate section of the proximal tibia.

*Partial medial physal stimulation* with a subsequent valgus deformity may be observed after greenstick fractures of the proximal tibia. The imbalance between rapid consolidation of the lateral bending component and the delayed healing of the completely fractured tension side

results in increased medial vascularity and thus asymmetrical plate growth (as proven by bone scans) [20]. The interposition of periosteum, the collateral medial ligament or the pes anserinus can maintain this process. A growth of up to 1 cm in tibial length may be observed.

A tibia valga as the result of medial physal stimulation after proximal bowing fractures in children under 6 years of age should be corrected surgically, preferably towards the end of growth, in view of the high risk of recurrence. In children over 6 years with a difference



### 3.3.9 · Fractures of the knee and lower leg

between sides of over 10–15°, a correction osteotomy should be performed in the first year after trauma. If this is delayed, it may subsequently be possible to correct the deformity only by means of a double osteotomy, since the valgus position moves to the center of the shaft as growth progresses and the proximal and distal tibial joint surfaces spontaneously align themselves horizontally resulting in an S-shaped double deformity.

#### 3.3.9.4 Fractures of the tibial diaphysis

##### Occurrence

Fractures in the area of the tibial diaphysis are common, accounting for around 15% of all fractures during childhood and adolescence. A crucial distinction for treatment and prognosis is between isolated tibial fractures and complete lower leg fractures, which occur in a ratio of 2:1.

##### Diagnosis

##### Clinical features

*Local pain and swelling.* Deformities can be detected on clinical examination thanks to the thin anteromedial soft tissue covering.

Even if the axes are in the anatomical position, a malrotation may be present and can be quantified by comparing the angle between the transverse axis of the tibial head and the malleolar axis on both legs. Essential requirements include the recording of the pulses of the dorsalis pedis and posterior tibial arteries by clinical means, and if necessary by doppler ultrasound, and a well documented peripheral neurological status. Taut swelling of a muscle compartment, usually the anterior tibial compartment, and severe pain indicate a threatened or established compartment syndrome.

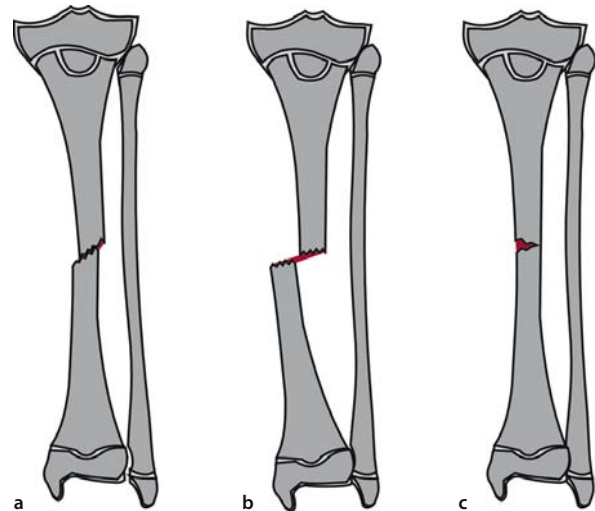
##### Imaging investigations

Standard AP and lateral x-rays, including the adjacent joints. If there is an obvious deformity on clinical examination and reduction is clearly indicated, one x-ray in the position that is least distressing for the patient is sufficient.

##### Fracture types

*Isolated tibial fractures* (■ Fig. 3.329) represent the commonest fractures of the lower limb during childhood and adolescence, typically occurring in children under 10 years of age after indirect trauma. The fractures are usually oblique or spiral and originate from the transition between the distal and middle third of the bone.

- *Compression and greenstick fractures* are rare and generally show only slight angulation.
- *Complete tibial fractures* rarely produce shortening thanks to the splinting effect of the intact fibula, and initially involve a varus deformity of less than 10°. Further varization is observed during the first 2–3 weeks in around 50% of patients.



■ Fig. 3.329a–c. *Isolated tibial shaft fractures:* The classical oblique fracture represents a stable situation, even though it can result in a varus deformity (a). Rare, completely displaced transverse fractures (b) are unstable, in contrast with greenstick fractures (c)

*Complete lower leg fractures* (■ Fig. 3.330) are usually the result of direct trauma, particularly in road traffic accidents. The instability of the fibula and the consequent shortening action of the anterolateral muscle groups produces valgus angulation.

##### Treatment

##### Spontaneous corrections

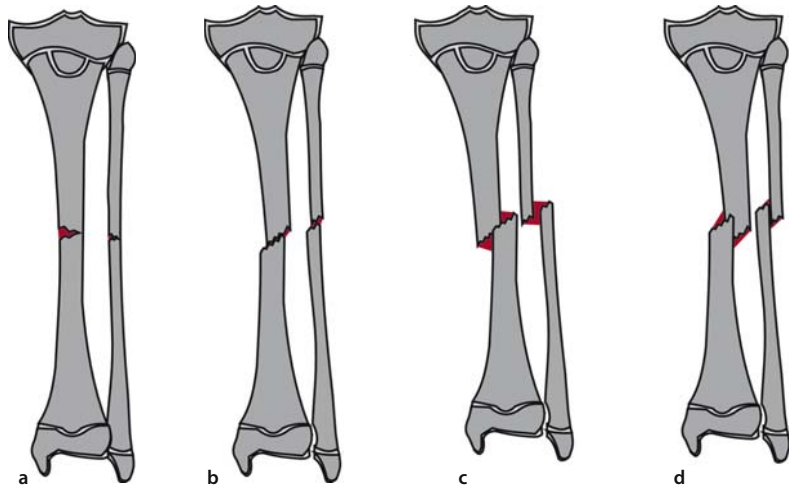
Varus deviations of 10–15° and recurvations of approx. 20° are spontaneously corrected in children under 10. Anterior bowing deformities are rare. The remodeling of malrotations, usually external rotational deformities, is unreliable before the age of 5 and cannot be expected to occur at all at a later age [16]. Although most deformities are within the remodeling tolerance range, they should nevertheless be correct initially in order to ward off post-traumatic leg length discrepancies. Valgus errors have a negligible correction potential.

##### Conservative treatment

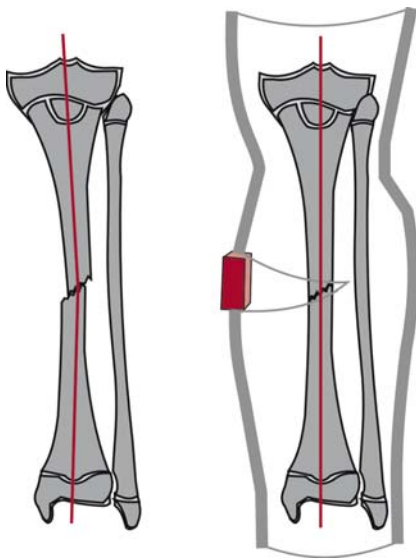
Non-displaced fractures and those with axial deviations of up to 10° are immobilized in a plaster slab, which is replaced by an encircling cast after the swelling has subsided. Primary or secondary axial deviations of up to 20° are wedged after 7–10 days (■ Fig. 3.331).

##### Surgical treatment

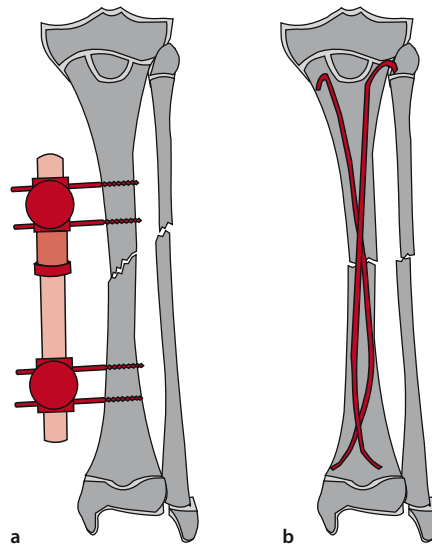
Axial deviations of over 20° and rotational deformities of over 10° are reduced by closed manipulation and secured in a plaster slab, which is closed up after the swelling has subsided. The position and the need for plaster wedging is checked after 7–10 days.



**Fig. 3.330a–d. Lower leg shaft fractures:** The rare greenstick fracture can be classed as a stable fracture (a). All transverse fractures with good fragment contact are considered to be relatively unstable (b). All completely displaced fractures and oblique fractures are considered to be unstable (c, d)



**Fig. 3.331. Treatment of stable diaphyseal fractures of the lower leg:** These fractures are treated conservatively as a rule. Cast wedging is the most reliable method of eliminating any occurring, or primarily present, axial deviations



**Fig. 3.332a, b. Treatment of unstable diaphyseal fractures of the lower leg:** To ensure the earliest possible weight-bearing and mobilization of the patient, unstable spiral, comminuted or long oblique fractures are managed with an external fixator (a), while unstable transverse and short oblique fractures are managed with descending flexible intramedullary nails (b)

Surgical fixation is indicated for unstable fractures:

- Completely displaced transverse fractures.
- Complete lower leg fractures with more than 1 cm of shortening.
- Open fractures.
- Compartment syndrome (fasciotomy).

We use descending intramedullary flexible nails for transverse and short oblique fractures and a monolateral external fixator for long oblique, spiral and multifragment fractures (Fig. 3.332).

The *immobilization period* ranges from 2–3 weeks for compression fractures in under 5-year olds to 8–10 weeks for comminuted fractures in adolescents.

**Follow-up management**

Patients with a cylinder cast are mobilized, without weight-bearing, on crutches, while children with surgically stabilized fractures are mobilized as permitted by the severity of symptoms. Clinical controls are continued for 2 years after the trauma in order to check for any post-traumatic leg length discrepancies.

## Complications

### Growth disturbances

The more pronounced the deformity on consolidation, the more active and the more protracted are the remodeling processes and the greater the stimulation of the tibial growth plates. A posttraumatic increase in length of 0.5–1 cm at most can be expected in children under 10 years. Correction of the length discrepancy may be required and will depend in each case on the extent of any pre-existing leg length discrepancy. Shortening may persist in older patients with no further potential for sufficient growth or length alteration can occur as a result of posttraumatic premature physal closure [15].

### Posttraumatic deformities

Rotational deformities of over 10° are cosmetically conspicuous and cannot be offset in functional respects by the adjacent joints. Accordingly, the need for a supramalleolar derotation osteotomy should be decided at an early stage. The same applies to valgus deformities in all age groups and to varus deformities from the age of 10.

### Pseudarthroses

Pseudarthroses are extremely rare and only observed after high-energy traumas, comminuted fractures, open fractures, osteomyelitis and unstable internal fixations [12].

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## 3.3.10 Infections of the knee and lower leg

### 3.3.10.1 Septic arthritis of the knee

#### Preliminary anatomical observation

Since the knee capsule is attached to the metaphyseal sections of the femur and tibia, i.e. beyond the physis, metaphyseal osteomyelitic foci in the distal femur or proximal tibia can primarily lead to symptomatic, reactive, sterile effusions in the knee. If the infection persists for a prolonged period, it can penetrate into the joint and produce secondary infection of the knee effusion. This can also occur in association with isolated hematogenous osteomyelitis of the patella.

#### Etiology

#### Hematogenous septic arthritis as a primary condition

Up to the age of 3, a septic arthritis of the knee generally occurs as a primary condition in the joint (▶ Chapter 4.3.2). In a recent study 204 children (137 boys and 67 girls) aged 12 years and under with septic arthritis were examined [5]. Their mean age was 2.6 years. The majority of cases therefore occurred in early childhood. The most common joints affected were the knees and shoulders. The most common organisms cultured (in the Malawian children of this study) were species of Salmonella.

#### Hematogenous osteomyelitis

After the age of 3, the infection normally originates in the metaphysis of the distal femur or proximal tibia. Rare sites include the proximal fibular metaphysis or the patella [7]. Any type of hematogenous osteomyelitis may be involved. Whereas a common pathogen in infancy used to be Haemophilus influenzae, this type of osteomyelitis has almost disappeared following the systematic vaccination against this organism [1, 2].

### Posttraumatic infections

The knee is particularly exposed to all kinds of injuries, including the penetration of foreign bodies, as a result of falls or direct trauma. Penetrating and perforating infections of the knee can occur via (inadequately treated) superinfected abrasions of the patella, septic bursitis or intra-articular foreign bodies. The pathogens most commonly involved are staphylococci, although an infection can be caused by any other organism.

### Frequency and location

Precise frequency data about direct and indirect, primary and secondary, hematogenous and posttraumatic infections of the knee are not available.

### Growth prognosis

Epiphyseal separation of the affected bone can occur in infants in connection with septic arthritis. This usually results in a central partial closure of the growth plate with the formation of a cone epiphysis (■ Fig. 3.333, 3.334). Occasionally however, the growth disturbance only occurs years later, hence the importance of follow-up checks until the completion of growth [6].

### Diagnosis and treatment

Further details can also be found in ► chapter 4.3.2.

! Any local pain and any swelling and inflammation in a patient with a fever must continue to be handled as a case of septic arthritis or acute hematogenous osteomyelitis until the contrary is proven.

The septic arthritis is diagnosed by the detection of pathogens in the joint aspirate, while the acute hematogenous osteomyelitis is diagnosed by the detection of pathogens in the blood, local aspirate or – if no germ can be found – by a bone scan or MRI. The possibility of a primarily chronic osteomyelitis should be considered if an osteolytic focus is observed in the primary or postprimary period in

a patient without fever but with local pain, swelling and inflammation. The most important diagnosis to be differentiated is a tumor (► Chapter 3.3.12). The diagnosis must be confirmed by a biopsy, which also serves as treatment at the same time.

! Conservative treatment for a suspected case of primarily chronic osteomyelitis is obsolete.

The possibility of an infection should be considered if any growing pain or any increasing swelling and inflammation are observed after an operation or some other soft tissue injury. The infection may originate from a deep-seated focus or still be superficial. If an initial conservative treatment does not produce a prompt improvement within 5 days at most, surgical revision, or at least opening of the wound, is indicated. The usual *laboratory sample* is taken if an infection is suspected. If, exceptionally, an effusion cannot be clinically evaluated with any



■ Fig. 3.334a, b. Post-infection (partial) physeal closure. a Closure of the distal femoral epiphyseal plate in a 4-year old boy. b Partial closure of proximal tibial epiphyseal plate in a 4-year old girl with consecutive genu varum



■ Fig. 3.333a–f. Course of septic arthritis of the knee. a 18-day old infant; admitted with a swollen, painful right knee. The check x-ray shows epiphyseal separation of the distal femur. Aspiration produced a creamy pus. b After irrigation and intravenous antibiotic sepsis treatment for 14 days. c Healed infection (6-week check). d, e, f Situation at

1, 5 and 9 years. The initial periosteal reaction subsequently regressed spontaneously. The residual central portion of the distal femoral epiphysis is suggestive of cone epiphysis formation (5-year check-up), although the situation had returned to normal by the 9-year check-up (f). The leg is shortened by 2 cm and the knee is freely mobile



certainly, an *ultrasound scan* must be arranged, as well as *x-rays* in two planes.

Any *effusion in a patient with fever* must be aspirated as a matter of urgency. Administration of antibiotics before aspiration should be avoided at all costs in order to allow cultivation of micro-organisms. If the exudate is cloudy or purulent, arthroscopic joint lavage is performed in the same session. Even much more than for the hip, the arthroscopic lavage of the infected knee has also become a widely accepted procedure in children [4]. We dispense with the period of immobilization that used to be standard practice and now mobilize the patient immediately with the dynamic splint. The subsequent management is the same as that for all other types of septic arthritis (► Chapter 4.3).

#### Follow-up management

Follow-up management essentially involves functional treatment on the dynamic splint. The knee is not immobilized, rather the patient is mobilized from the outset with crutches without weight-bearing.

#### Follow-up controls

On completion of the antibiotic treatment, the C-reactive protein (CRP) is rechecked 8 days later. If this has returned to normal, knee function is checked 4 weeks later. If the knee is fully mobile by this time, check-ups at 3- to 6-monthly intervals, possibly for up to 2-years after the onset of the illness (depending on the underlying illness in each case) will suffice. Thereafter, possible leg length discrepancies and growth problems should be monitored, in addition to joint function, until the completion of growth.

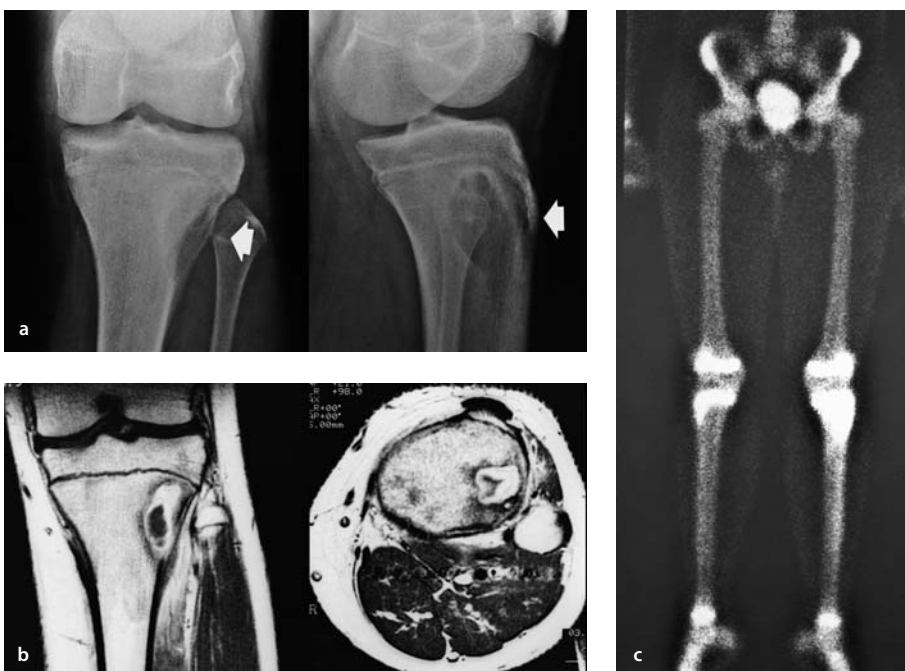
#### Post-infection deformities

Partial or complete destruction of the nearest growth plate – and in some cases even a whole condyle [8] – with corresponding abnormal growth can occur in connection with protracted infections. Only rarely are the epiphyseal or metaphyseal bridges small enough to permit the resection of same. They are usually so extensive that symptomatic corrections, lengthening osteotomies, growth plate obliteration, etc. are the only options for treating the deformity.

#### 3.3.10.2 Infections in the lower leg

The shaft of the lower leg is affected by an existing osteomyelitis almost exclusively in the chronic stage. Incipient osteomyelitis always affects the metaphyses (this is discussed in detail in ► chapter 4.3.1). Secondarily chronic stages with sequestrum formation, infected pseudarthroses etc. have fortunately become rare events in the developed world. Their treatment should always be based on the individual situation in each case, and all the available surgical methods and conservative measures must be employed to this end. See ► chapter 4.3.1 for details of diagnosis and treatment.

In the lower leg osteomyelitis behaves as it does in other long bones, although defective healing occurs more frequently here than in the femur [3], probably because of the poorer circulatory situation. The proximal medial tibial metaphysis tends to be particularly susceptible to primary chronic osteomyelitis. This is the commonest site for the »Brodie abscess« (■ Fig. 3.335). In a study of 21 cases of Brodie abscesses, 13 occurred in the second de-



■ Fig. 3.335a–c. 14-year old boy with primarily chronic osteomyelitis in the proximal tibial metaphysis (*Brodie abscess*). a X-rays. b MRI. c Technetium bone scan



cade of life, 11 were located in the tibia and 7 exclusively affected the proximal medial tibial metaphysis [9]. The treatment consists of careful surgical removal.

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### 3.3.11 Juvenile rheumatoid arthritis of the knee

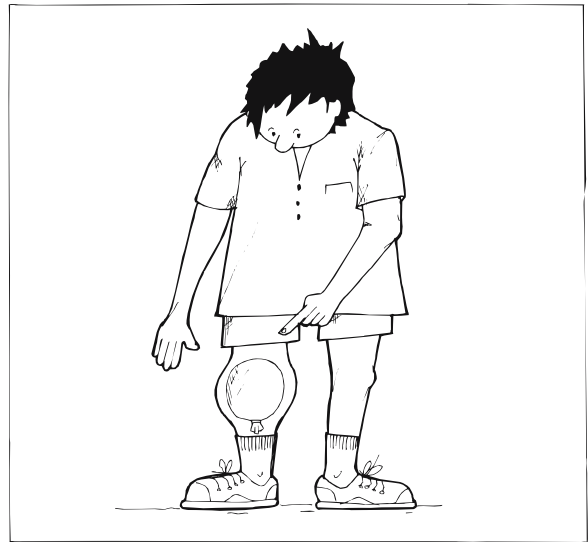
#### Occurrence

The knee is less frequently affected than the hip in connection with juvenile rheumatoid arthritis, which is discussed in detail in ► chapter 4.4. The hip is involved in around 9% of cases [5], but precise figures are not available for the knee.

#### Clinical features

Juvenile rheumatoid arthritis of the knee is relatively easy to diagnose. If a chronic effusion occurs without a history of trauma and persists for more than a month, this condition is likely to be present. The effusion can readily be detected on clinical palpation (► Chapter 3.3.1) and can be confirmed by an *ultrasound scan*. In addition to the effusion, knee movement is restricted (only slightly so in the initial stages).

If a knee effusion without a traumatic cause is diagnosed, the other large joints must always be examined as well. Around half of the cases of juvenile arthritis occurring in the knee are mono- or pauciarticular, while the



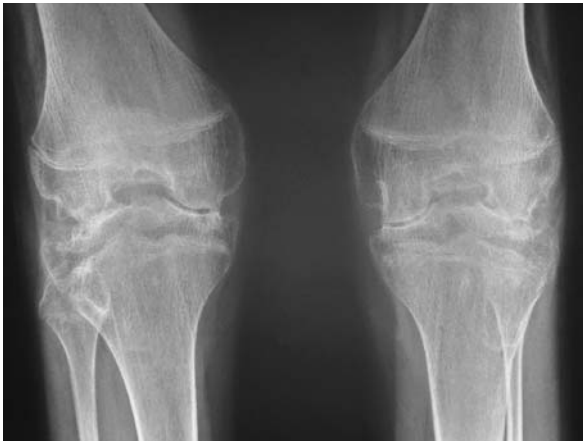
other half are polyarticular or systemic. The other knee and the hips, ankles, shoulders and elbows must also be examined, with palpation of the contours and measurement of the range of motion. If the range of motion of the hip is restricted, an ultrasound scan is useful for identifying an effusion. The *course* of the disease can be regressive, recurrent or progressive (► Chapter 4.4).

The progressive form often involves the formation of joint contractures. No changes are initially apparent on the *x-ray*. Signs of osteoarthritis may appear at a later stage in the chronic form. In contrast with osteoarthritis-caused by overexertion, rheumatoid arthritis starts not with unilateral but symmetrical narrowing of the joint space. The sclerosis near the joint is less prominent than the cyst formation and osteoporosis (► Fig. 3.336). A *laboratory test* is always indicated, although the rheumatoid factors are only positive in rare cases (► Chapter 4.4). The rheumatoid factors have greater prognostic rather than diagnostic value.

The most important *differential diagnosis* is an effusion or hematoma after trauma. It is not always possible to establish whether a child has actually suffered an injury, and sometimes a trauma will be reported that is of no relevance to the formation of the effusion. *Spontaneous effusions* (hematomas) also occur (without relevant trauma) in:

- osteochondrosis dissecans (► Chapter 3.3.4),
- chronic instability (► Chapter 3.3.8),
- infectious arthritis (► Chapter 3.3.10),
- hemophilia (► Chapter 4.6.9),
- synovial chondromatosis (► Chapter 4.5.4.2),
- pigmented villonodular synovitis (► Chapter 4.5.4.2).

The first four conditions on this list can be diagnosed by careful history-taking, clinical examination, an x-ray and possibly routine laboratory tests. Synovial chondromato-



■ **Fig. 3.336.** AP x-rays of both knees of 14-year old female patient with severe juvenile rheumatoid polyarthritis and widespread destruction of the knees

sis and villonodular synovitis are not visible on the x-ray in the initial stages. If one of these diagnoses is seriously suspected, an MRI scan is indicated. A spontaneous effusion similar to that in transient synovitis of the hip has not been observed in the knee.

### Treatment

#### Conservative treatment

The conservative treatment of juvenile rheumatoid arthritis of the knee is no different from that for arthritis in the other joints. See ► chapter 4.4. for the drug treatment.

#### Surgical treatment

The following options are available:

- soft tissue procedures,
- correction of contractures with an external fixator,
- knee implants.

#### Soft tissue procedures

We distinguish between the following operations:

- Hydraulic mobilization under anesthesia,
- Synovectomies.

#### Hydraulic mobilization under anesthesia

If an incipient joint contracture only is present and there is no major arthrosis in the joint, hydraulic mobilization of the knee can prove worthwhile. We use the arthroscope for this procedure in which the affected joint is filled several times with fluid under high pressure, causing the capsule to distend. The surgeon can also take this opportunity to assess the state of the cartilage and synovium at the same time. Postoperatively, the child is placed on a dynamic splint. The epidural anesthesia can be continued via an epidural catheter for several days while intensive physical therapy is administered. Associated with minimal morbidity, this is a relatively simple way of

improving the range of motion, and particularly walking ability, for several months, or even years. No long-term effect, however, is achievable for the progressive form of the condition.

#### Correction of the flexion contracture (with the ring fixator Ilizarov apparatus or Taylor Spatial Frame)

The principle of this treatment is discussed in ► chapter 3.3.13. This treatment can be successfully employed even for severe flexion contractures and very advanced arthroses [1, 2]. The permanent flexion position of the knee hinders walking so much that the actual ability to walk at all is jeopardized even if the hips and ankles are only slightly affected. While the Ilizarov apparatus can produce permanent extension, one should not expect the full range of movement to be restored by this treatment. What is gained in extension is lost in flexion. For practical everyday purposes, however, extension is much more important than flexion.

#### Synovectomy

The once common procedure of synovectomy has proved a disappointment in juvenile rheumatoid arthritis. Although it reduces the pain temporarily, it does not improve the range of motion, and thus everyday knee function. Radiological follow-up has even shown a faster progression of the arthrosis [3]. Synovectomy cannot be recommended, therefore, as a treatment for juvenile rheumatoid arthritis [5].

#### Knee implants

If the patient loses the ability to walk as a result of the progressive arthrosis, only a total knee replacement will be able to restore mobility. Knee implants are not as common as artificial hips in young patients, but can prove useful in some cases [3]. As with hip implants, a relatively high perioperative complication rate can be expected with this patient group (because of the long-term treatment with cortisone and cytotoxic drugs).

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### 3.3.12 Tumors in the knee area

#### ➤ Definition

Primary bone tumors originating in the distal femur, patella, proximal tibia or fibula, and soft tissue tumors arising from the muscles, connective tissues, blood vessels or nerve tissues in the immediate vicinity of the knee.

#### Occurrence

##### Bone tumors

Almost 40% of all bone tumors in children and adolescents occur in the knee area. The distal femur is the commonest site for bone tumors, accounting for 21% of cases, followed by the proximal tibia in second place (18%). There is a logical explanation for this situation: the epiphyseal plates in this area are the most active in terms of growth. The distal femoral epiphyseal plate alone accounts for approx. 20% of the total growth in a person's

height (i.e. approx. 25 cm for a final height of 175 cm, involving a growth of 125 cm from birth = 50 cm to completion of growth).

The corresponding proportion contributed by the proximal tibial epiphyseal plate is approx. 13%. The distribution of the various tumor types around the knee is shown in ■ Table 3.60. The individual tumor types are described in detail in ► chapter 4.5.2–4.5.4.

##### Benign tumors

##### Distal end of the femur

The tumor that most commonly occurs in the area of the distal femoral metaphysis is the osteochondroma (cartilaginous exostosis; ► chapter 5.5.2.2). Since this tumor often interferes with the knee mechanics it occasionally has to be resected. This region is also a very typical site for non-ossifying bone fibromas. In absolute terms this tumor is much more common than would be suggested

■ Table 3.60. Primary bone tumors of the distal femur, proximal lower leg and lower leg shaft in children and adolescents (n=805) compared to adults (n=849). (Basel Bone Tumor Reference Center)

	Children and adolescents						Adults					
	Distal femur		Proximal tibia + tibia		Lower leg shaft		Distal femur		Proximal tibia + tibia		Lower leg shaft	
Osteochondroma	144	36.7%	<80	24.1%	7	8.6%	70	15.2%	31	11.6%	9	7.4%
Chondromyxoid fibroma	2	0.5%	4	1.2%			2	0.4%				
Chondroblastoma	16	4.1%	7	2.1%			9	2.0%	3	1.1%		
Enchondroma	3	0.8%	1	0.3%	5	6.2%	62	13.5%	33	12.3%	18	14.9%
Aneurysmal bone cyst	14	3.6%	15	4.5%	3	3.7%	6	1.3%	5	1.9%	5	4.1%
Non-ossifying bone fibroma	58	14.8%	89	26.8%	3	3.7%	17	3.7%	7	2.6%	3	2.5%
Giant cell tumor	8	2.0%	8	2.4%			50	10.9%	34	12.7%	1	0.8%
Fibrous dysplasia	1	0.3%	10	3.0%	1	1.2%	4	0.9%	13	4.9%	4	3.3%
Osteofibrous dysplasia		0.0%	4	1.2%	5	6.2%		0.0%	1	0.4%	4	3.3%
Osteoid osteoma/osteoblastoma	6	1.5%	8	2.4%	9	11.1%	5	1.1%	7	2.6%	16	13.2%
Other (benign) tumors	16	4.1%	16	4.8%	18	22.2%	20	4.3%	18	6.7%	17	14.0%
<i>Tumor-like lesions</i>	9	2.3%	8	2.4%	5	6.2%	14	3.0%	11	4.1%	13	10.7%
<b>Osteosarcoma</b>	<b>77</b>	<b>19.6%</b>	<b>54</b>	<b>16.3%</b>	<b>9</b>	<b>11.1%</b>	<b>48</b>	<b>10.4%</b>	<b>24</b>	<b>9.0%</b>	<b>8</b>	<b>6.6%</b>
<b>Parosteal osteosarcoma</b>											<b>6</b>	<b>5.0%</b>
<b>Ewing sarcoma</b>	<b>7</b>	<b>1.8%</b>	<b>9</b>	<b>2.7%</b>	<b>7</b>	<b>8.6%</b>	<b>2</b>	<b>0.4%</b>				
<b>Chondrosarcoma</b>	<b>5</b>	<b>1.3%</b>	<b>1</b>	<b>0.3%</b>			<b>39</b>	<b>8.5%</b>	<b>25</b>	<b>9.3%</b>	<b>8</b>	<b>6.6%</b>
<b>Adamantinoma</b>				<b>0.0%</b>	<b>6</b>	<b>7.4%</b>	<b>56</b>	<b>12.2%</b>	<b>28</b>	<b>10.4%</b>	<b>3</b>	<b>2.5%</b>
<b>Other malignant tumors</b>	<b>26</b>	<b>6.6%</b>	<b>18</b>	<b>5.4%</b>	<b>3</b>	<b>3.7%</b>	<b>56</b>	<b>12.2%</b>	<b>28</b>	<b>10.4%</b>	<b>6</b>	<b>5.0%</b>
<b>Total</b>	<b>392</b>	<b>100%</b>	<b>332</b>	<b>100%</b>	<b>81</b>	<b>100%</b>	<b>460</b>	<b>100%</b>	<b>268</b>	<b>100%</b>	<b>121</b>	<b>100%</b>

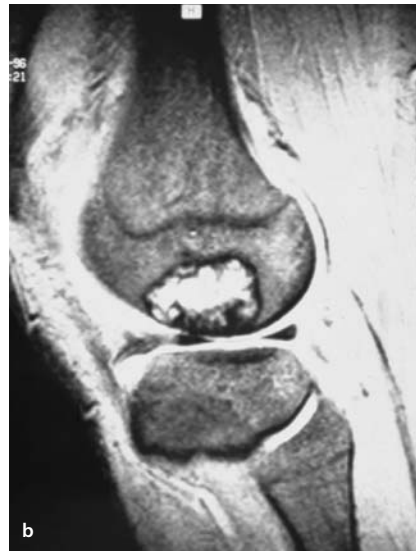
## 3.3.12 · Tumors in the knee area



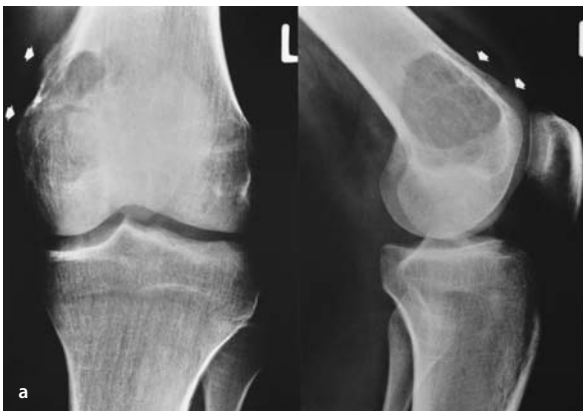
■ **Fig. 3.337.** Lateral x-ray of the left knee of a 15-year old girl with a *non-ossifying bone fibroma*. The clear demarcation with marginal sclerosis and the lobular structure are typical

by the statistical data of our bone tumor register, since such a tumor is hardly ever biopsied these days. A reliable diagnosis can usually be made on the basis of a plain x-ray, and treatment is unnecessary since non-ossifying bone fibromas either disappear spontaneously or ossify after completion of growth (■ Fig. 3.337). Moreover, the patients are almost always asymptomatic and the fibroma is almost invariably diagnosed by chance. Non-ossifying bone fibromas form at the sites where the tendons and ligaments radiate out in the vicinity of the epiphyseal plates [35] and the great majority are encountered around the knee.

A very typical tumor in adolescents at this site is the chondroblastoma (■ Fig. 3.338). In contrast with the two aforementioned tumors, which almost always occur in the metaphyses, the chondroblastoma is primarily always observed in the epiphyseal area. Chondroblastomas are



■ **Fig. 3.338a, b.** AP x-ray (a) and sagittal MRI (b) of the left knee of a 15-year old girl with *chondroblastoma* in the lateral femoral condyle. Such a finding should not be confused with a case of osteochondrosis dissecans (▶ Chapter 3.3.4)



■ **Fig. 3.339a, b.** AP and lateral x-rays (a) of the left knee of a 16-year old boy with a metaphyseal/epiphyseal *giant cell tumor* with second-

ary *aneurysmal bone cyst*. **b** Situation 6 months after curettage and reconstruction with allogeneic bone graft (allograft)

painful and are not diagnosed as a chance finding but always on the basis of the signs and symptoms.

Another relatively common tumor is the aneurysmal bone cyst (■ Fig. 3.339), which are also located primarily in the metaphyses. All other tumor types only occur rarely. By comparison with adults, the giant cell tumors and enchondromas in particular are underrepresented in children and adolescents (■ Table 3.60). In contrast with the situation for the proximal femur, solitary bone cysts hardly ever affect this area. Similarly, fibrous dysplasia rarely occurs here. In general, the ratio of benign to malignant tumors in the statistical data for our register tends to favor the malignant type, since many benign tumors neither need to be biopsied nor treated and therefore do not appear in the statistics. All malignant tumors, on the other hand, are recorded in the register.

### Proximal tibia and fibula

For the most part, the same tumors form on the proximal lower leg as on the distal femur (■ Table 3.60). Osteochondromas are very typical and frequently irksome. They can almost always be diagnosed reliably on the basis of a plain x-ray. Non-ossifying bone fibromas are even more frequently encountered in the proximal tibial metaphysis than in the distal femur. Giant cell tumors and fibrous dysplasia are also slightly more common here. This is a particularly typical site for the rare chondromyxoid fibroma [47]. On the other hand, giant cell tumors are rare compared to their frequency of occurrence in adults [24, 29]; this also applies to the enchondroma. Here, too, many more benign tumors occur in this part of the body, in absolute terms, than would be suggested by the statistical records.

### Tibial shaft

Tumors in the tibial shaft are fairly rare (as generally applies for diaphyses). In addition to osteoid osteomas, enchondromas, aneurysmal bone cysts (■ Fig. 3.340) and fibrous dysplasia, osteochondromas can also grow from the metaphysis into the shaft area. A condition that particularly affects the tibial shaft is *osteofibrous dysplasia according to Campanacci* (■ Fig. 3.341), a tumor that almost always occurs on the tibia, predominantly in the shaft area [5, 32, 42] (▶ Chapter 4.5.2.7). This condition can sometimes be confused with the malignant *adamtinoma*, which also occurs almost exclusively at this site (▶ Chapter 4.5.3.6).

### Patella

Tumors on the patella are very rare. We have only encountered 11 cases. Only the osteoma occurred twice. The other 9 patients had 9 different (benign) tumors. No malignant tumors were observed. The literature primarily describes chondroblastomas and giant cell tumors, while malignant neoplasms are extremely rare [27].



■ Fig. 3.340. AP and lateral x-rays of the left knee of a 11-year old girl with an *aneurysmal bone cyst* in the tibial shaft



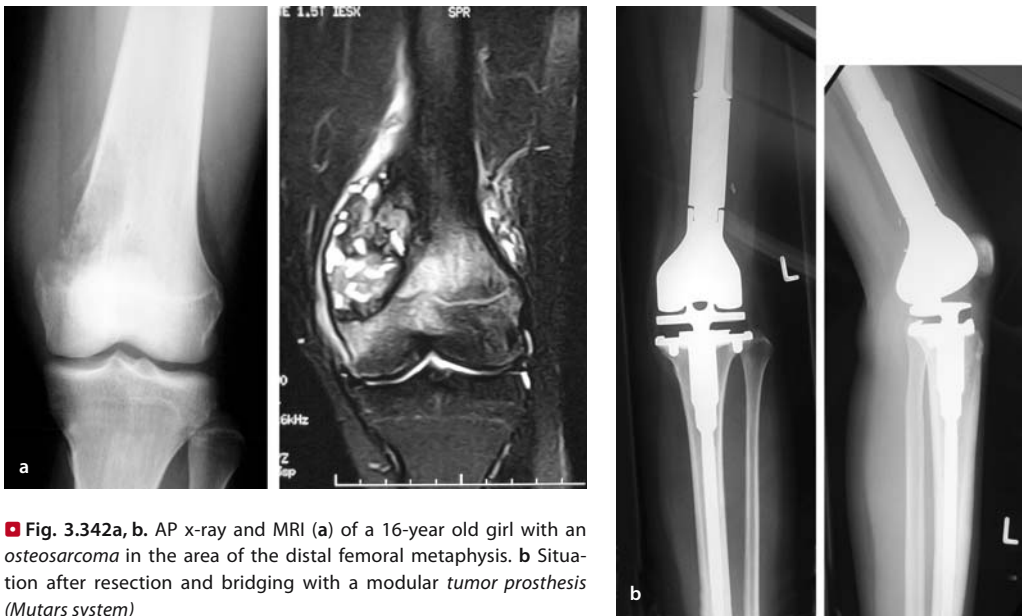
■ Fig. 3.341. AP and lateral x-rays of the left knee of a 7-year old girl with an *osteofibrous dysplasia (Campanacci)* of the proximal tibia

### Malignant tumors

#### Distal end of the femur

The distal femoral metaphysis is the classical site of the *osteosarcoma* (■ Fig. 3.342), accounting for around 1/4 to 1/3 of all osteosarcomas. Although any other malignant primary bone tumor can also occur here, they are fairly rare in children and adolescents. This also applies to Ewing sarcoma, which tends to occur in the diaphysis





**Fig. 3.342a, b.** AP x-ray and MRI (a) of a 16-year old girl with an osteosarcoma in the area of the distal femoral metaphysis. b Situation after resection and bridging with a modular tumor prosthesis (Mutars system)

and is only exceptionally located at the end of the femur. Similar frequencies are reported in the literature [14]. Tumors that are particularly common in adults include parosteal or periosteal osteosarcoma (in contrast with the classical high-grade osteosarcoma, these are weakly malignant tumors), chondrosarcoma and malignant fibrous histiocytoma.

#### Proximal tibia and fibula

The distribution of malignant tumors in the proximal lower leg is also similar to that in the distal femur. The classical osteosarcoma dominates in children and adolescents, while all of the other malignant tumors rarely occur.

#### Tibial shaft

Malignant tumors occur less commonly in the shaft area compared to the metaphysis. As a medullary cavity tumor, however, Ewing sarcoma usually forms in the diaphysis or metadiaphysis. One tumor that particularly affects the tibia is the *adamantinoma*, a low-grade malignant tumor that occurs only in the tibia (► Chapter 4.5.3.6). It can be confused with a benign osteofibrous dysplasia [42].

#### Soft tissue tumors

In contrast with the pelvis and upper thigh, soft tissue tumors are easier to diagnose in the knee area since they are not covered by a thick soft tissue layer. Soft tissue tumors are rare in children and adolescents. Lipomas are observed, as are desmoids. The knee itself can be affected

by *synovial chondromatosis* [7] and *pigmented villonodular synovitis* [1]. The knee is the second most frequent site for the latter condition after the finger joints. Although these involve benign changes they can cause major treatment problems. *Popliteal cysts* are very typical and common tumor-like lesions in children. These are cysts filled with a gelatinous fluid that form from the tendon attachment of the gastrocnemius muscle, or more rarely the popliteal, semimembranosus or biceps muscles. Popliteal cysts should not be confused with Baker cysts, which form as an excrescence of the joint capsule in internal knee lesions associated with degenerative changes in the knee. Baker cysts hardly ever occur in children and adolescents. Large popliteal cysts can sometimes prove slightly irksome, but disappear spontaneously, at the latest by the completion of growth.

Of the malignant soft tissue tumors, the *rhabdomyosarcoma* is the commonest in children and adolescents.

#### Diagnosis

Since the knee only has a thin soft tissue covering, tumors in this area are usually diagnosed at a relatively early stage. The following principle, in particular, should be observed:

- ❗ **The knee should always be x-rayed if pain of unclear origin that is not clearly load-related persists for longer than 4 weeks. Particular attention is indicated if the pain occurs at night.**

It is important to distinguish between the pain associated with tumors and »growing pains«. These also occur at

night, typically around the knee. Further investigations can be dispensed with if clinical examination shows no abnormalities in the toddler and the pains alternate or occur simultaneously on the right and left sides. If either of these criteria is not fulfilled, an x-ray must always be recorded. This even applies if an adolescent rather than a toddler is involved.

The primary imaging procedure is always the *plain x-ray*. Two common tumors that occur around the knee can be reliably diagnosed on the basis of the plain x-ray: an osteochondroma (cartilaginous exostosis) and a non-ossifying bone fibroma. No further investigations are required to confirm these diagnoses. Note however that the diagnosis of »non-ossifying bone fibroma« is unlikely to explain the symptoms. Very rarely, a break in the cortical bone in the tumor area can cause symptoms, particularly in the proximal fibula. The subsequent diagnostic procedure in uncertain cases is detailed in ► chapter 4.5.1.

### Treatment

#### Benign tumors

*Osteochondromas* in the area of the knee should be removed only if they interfere with the knee mechanics. Although widely differing opinions have been expressed in the literature on the risk of malignant degeneration this is probably fairly small [39]. If surgical treatment is chosen, the excision must include the base of the tumor as this is where malignant degeneration starts. While growth disturbances are less likely to occur in the lower extremity compared to the forearm in connection with multiple osteochondromas, they are still encountered fairly often [34]. Leg shortenings that require correction only occur sporadically, whereas axial deviations are more common. As mentioned above, *non-ossifying bone fibromas* do not require treatment. Growth disturbances arise particularly in connection with *enchondromatosis* (Ollier disease, Maffucci syndrome) [8] and often require axial corrections and leg lengthening procedures. Malignant degeneration is less likely to occur in enchondromas around the knee than in those close to the torso, for example in the pelvis or the torso itself [4, 18]. *Chondroblastomas*, *chondromyxoid fibromas*, *giant cell tumors*, *osteoblastomas* and *aneurysmal bone cysts* should be completely resected with a margin of healthy tissue if possible. Since they usually grow in the vicinity of the joint cartilage or epiphyseal plate en-bloc resection is rarely possible. Careful curettage is required in such cases (see ► chapter 4.5.5 for the procedure). The recurrence rate of these tumors can be reduced from over 50% to less than 10% with effective curettage [15, 26]. If the fibular head needs to be resected because of a tumor, a corresponding replacement must be constructed in order to provide an anchoring point for the lateral collateral ligament and the biceps femoris muscle (■ Fig. 3.343).

### Malignant tumors

#### Therapeutic strategies

The strategies for dealing with malignant tumors are described comprehensively in ► chapter 4.5.5. Only a few special features of tumors in the knee region will be mentioned at this point.

All malignant tumors in the knee area must be *surgically removed*. The surgeon should always try to perform a wide resection with a margin of healthy tissue. A radical (extracompartmental) resection is usually equivalent to an amputation and nowadays is indicated only in extremely rare cases.

Resection into healthy tissue means that the resected tissue must be surrounded by a layer of healthy cells. The surgeon should not be tempted to leave any tumor tissue in the patient because of the proximity of blood vessels. If necessary, vascular surgical bridging of vessel sections must be planned in advance. More problematic is resection in the vicinity of nerves. Whereas the lesion resulting from the concurrent resection of the femoral nerve in the distal femoral area (for example in the adductor canal) is acceptable, the limits of limb-preserving tumor treatment are exceeded with the concurrent resection of the sciatic nerve in the popliteal fossa. On the other hand, concurrent resection of the peroneal nerve with tumors of the fibula or proximal tibia is usually acceptable. The resulting foot drop can be offset by the use of a posterior leaf spring ankle foot orthosis.

The *epiphyseal plate* poses a particular problem. We do not take the growth plate into consideration in children of 10 years or older as the distal femoral epiphyseal plate contributes to subsequent growth by no more than 5 cm at this stage. Depending on the patient's expected final height we perform surgical closure of the contralateral epiphyseal plate concurrently with the resection or we schedule a lengthening procedure at a later date.

In children under 10 years of age, the subsequent leg length discrepancy can become considerable, but here too the resection must naturally include a margin of healthy tissue. One alternative to amputation is rotationplasty, as first described by Borggreve [3] and later by Van Nes [43] for the treatment of congenital defects. In this procedure the knee is resected and the lower leg with the foot – rotated through 180° – is anchored to the femur. As a consequence of this operation the upper part of the ankle functions as the knee (► Chapter 4.5.6). Although the rotated foot is difficult for the parents and child to accept psychologically [19], the functional advantages over amputation are so great that the esthetic disadvantages usually become well tolerated with time [17, 18, 23, 46] (► Chapter 4.5.5). A rotationplasty is also a suitable »salvage operation« in cases of infection or the failure of prostheses or allografts [44].



■ Fig. 3.343a, b. AP MRI and lateral x-ray (a) of the left knee of a 15-year old boy with a *giant cell tumor* in the area of the fibular head. b Situation 2 years after resection of the fibular head, division of the fibular shaft, replacement of the fibular head with half of the shaft, internal fixation and anchorage of the collateral ligament and iliotibial tract on the lengthened fibula. The patient's knee is very stable

### Reconstruction options

The treatment of malignant tumors of the distal femur or proximal tibia is usually associated with the loss of all or part of the joint surface. Only those tumors located in the proximal fibula can generally be removed without substantially impairing the joint (■ Fig. 3.343). The reconstructive measures in this case are limited to the anchoring of the lateral ligamentous apparatus of the knee. In many cases, however, the peroneal nerve also needs to be resected, which will need to be offset by the use of a posterior leaf spring ankle foot orthosis. If part of the joint surface must be removed as well then *reconstruction* will be required (► Chapter 4.5.5).

In our experience, the use of *allografts* in the knee area has not proved effective particularly in those cases in which only a part of the joint surface of the femur or tibia has to be removed. By contrast, the use of allografts

for the complete distal femur or proximal tibia is feasible (■ Fig. 3.344 and ■ 3.345). The advantage of the allograft over a joint prosthesis is the possibility of preserving the part of the joint opposite the tumor. At the proximal tibia this provides a better anchoring option, compared to a prosthesis, for the patellar tendon (and thus the complete extensor apparatus). Although considerable experience – up to 36 years – has been accumulated with the use of such large allografts, certain disadvantages should be mentioned: for example, joint function is not usually very good, the mechanical strength is inferior to that of metal implants, and the complication rate is very high (40% fractures, 15% infections) [9, 12, 19, 30, 36].

The use of metal and plastic *tumor prostheses* has become a standard method of treating malignant tumors in the knee area. These are modular prosthesis with resection and anchoring sections in any desired sizes. The femoral and tibial sections are firmly linked by a hinge joint. More recent models also permit rotation in the horizontal plane (rotating hinge; ■ Fig. 3.346). The most widely-used prostheses in Europe are the implant developed by Kotz [22] and the MUTARS prosthesis from Münster [6]. We routinely use such prostheses particularly for tumors of the distal femur (■ Fig. 3.342). Since the anchoring point of the extensor apparatus can be preserved, the functional results are very satisfactory.

Tumors of the proximal tibia are more problematic, as the anchorage for the patellar tendon is inadequate on the tumor prosthesis. With an allograft however, the fixed tendon can integrate with the allograft, which is not possible with a metal implant (■ Fig. 3.345).

The soft tissue covering occasionally represents a critical problem, hence the frequent use of gastrocn-

mius flaps with a vascular pedicle. The short- and medium-term results of treatment with tumor prostheses are very good [28], although long-term results, for example over a period of 50 years and more after the implantation of prostheses in adolescence, are awaited with interest. We have not had much experience with the combination of a homogenous osteocartilaginous graft (allograft)



■ Fig. 3.344a, b. AP and lateral x-rays (a top row) and frontal and sagittal MRI scans (a bottom row) of the right knee of a 17-year old girl with a *chondrosarcoma* of the distal femur in the area of the medial femoral condyle. b Situation 1 year after resection and replacement with allogeneic medial femoral condyle (allograft) and internal fixation



■ Fig. 3.345a, b. AP and lateral x-rays (a) of a 14-year old girl with an *osteosarcoma* in the metaphyseal/diaphyseal area of the proximal tibia. b Situation 1 year after resection and bridging with allogeneic bone (allograft)

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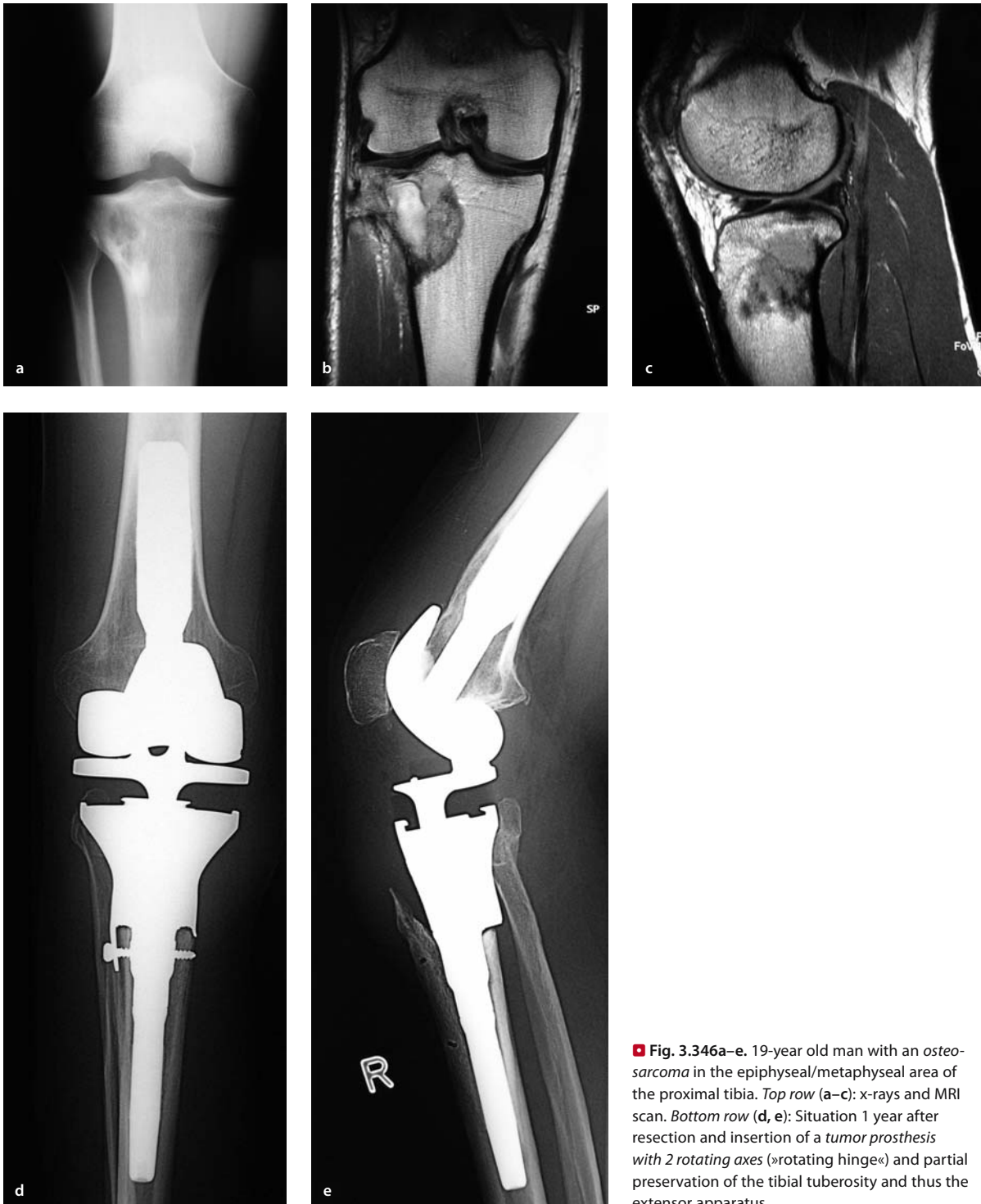


## 3.3.12 · Tumors in the knee area

and knee prosthesis. Such combinations are usual and useful at the proximal femur, but less so in the knee area. The literature only offers a few reports on such combinations [45]. There are only isolated reports on the procedure involving removal of the tumor with the joint section, extracorporeal irradiation of the bone and reinsertion at the site of removal [21]. An arthrodesis

is occasionally required if the use of a prosthesis is not possible.

Another alternative in *children under 10 years of age* is the extendable prosthesis [10, 11, 13]. Some centers now have over 20 years' experience with such prostheses. More recent models permit extension via a mechanism triggered by forced knee flexion, thereby minimizing the



**Fig. 3.346a–e.** 19-year old man with an *osteosarcoma* in the epiphyseal/metaphyseal area of the proximal tibia. *Top row (a–c):* x-rays and MRI scan. *Bottom row (d, e):* Situation 1 year after resection and insertion of a *tumor prosthesis with 2 rotating axes* (»rotating hinge«) and partial preservation of the tibial tuberosity and thus the extensor apparatus



number of revision operations. Nevertheless, the problems associated with this method should not be ignored. Since the bone grows not only in length but also in width, the anchorage of the prosthesis deteriorates with advancing growth. The lengthening also means a loss of power, and the epiphyseal plate on the other, unaffected and healthy side of the knee (where the prosthesis must also be anchored) shows reduced growth [10]. The risk of infection is relatively high because of the size of the prosthesis and the necessary number of operations.

### Soft tissue tumors

*Popliteal cysts* almost never require treatment as they always disappear spontaneously. Recurrences frequently occur after resections and can be avoided only if a part of the tendon from which the cyst originates is resected as well. Resection only needs to be considered if the popliteal cyst is large enough to cause symptoms (extremely rare). In patients with *synovial chondromatosis* the cartilage fragments must be carefully removed from the joint. A complete synovectomy (from the ventral and dorsal sides) is required in cases of *pigmented villonodular synovitis*. If it is not possible to bring the tumor under control by this method, a chemical synovectomy with osmic acid or radiocolloids may be required, although this treatment can be administered only after the patient has stopped growing (for further details see ► chapter 4.5.4).

### Prognosis

The survival rate after the treatment of malignant bone tumors in the knee area in children and adolescents has improved considerably over the last 30 years. Whereas the five-year survival rate for both osteosarcoma and Ewing sarcoma was below 15% in the 1970's, a survival rate of 90% can be expected nowadays if the osteosarcoma responds well to chemotherapy and the tumor is adequately resected [16]. The average five-year survival rate (including poor responders) is approx. 80% [25, 40]. The prognosis is not quite as good for Ewing sarcoma as micrometastases form at a very early stage with this tumor. Nevertheless, survival rates of over 50% are still achievable [33, 41]. It is important that the treatment should be administered in a center involved in a multi-center-evaluated tumor protocol.

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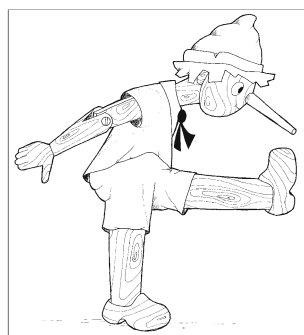
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## 3.3.13 · Knee contractures

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## 3.3.13 Knee contractures

» *Once upon a time there was a piece of wood. Master Cherry gave the piece of wood to his friend Geppetto, who took it to make himself a wonderful marionette that would dance, fence, and turn somersaults. »What name shall I give him?«, he said to himself. »I think I shall call him Pinocchio. This name will bring him luck...« ... and from the rigid wood he fashioned a living, moving marionette... « (Carlo Collodi)*



➤ **Definition**

Permanent restriction of the range of motion of the knee, usually in the form of incomplete extension (flexion contracture) or, more rarely, the loss of the ability to flex (extension contracture).

**Etiology, differential diagnosis**

The knee contracture is a symptom rather than a pathology and can be caused by a wide variety of factors. In the differential diagnosis we make a distinction between two situations:

- contractures already present at birth or which develop slowly in connection with a (known) systemic disorder;
- acutely occurring contractures, with or without trauma, that occur during growth unaccompanied by any known systemic disorder.

Typical systemic disorders in which contractures of the knees occur include:

- spastic cerebral palsy (▶ Chapters 3.3.7, 4.7.1 and 4.7.3),
- flaccid paralyzes (poliomyelitis, myelomeningocele; ▶ chapters 3.3.7, 4.7.1 and ▶ 4.7.4),
- severe chronic juvenile rheumatoid oligo- or polyarthrititis (▶ Chapters 3.3.11 and 4.4),
- arthrogryposis,
- pterygium syndrome,
- Larsen syndrome,

- diastrophic dwarfism,
- sporadically in many other hereditary disorders (incl. achondroplasia, osteogenesis imperfecta, nail-patella syndrome).

A slowly progressing flexion contracture of the knees can occur in these illnesses. Both knees are almost invariably affected to varying degrees. A severe flexion contracture on one side inevitably produces the same situation on the other leg as this cannot then be extended otherwise the patient's upper body will become imbalanced. Extension contractures also occur but are extremely rare.

The actual causes of the contracture can be:

- neuromuscular (e.g. in cerebral palsy, arthrogryposis),
- arthrogenic (e.g. in rheumatoid polyarthritis),
- the result of connective tissue changes (e.g. in pterygium or Larsen syndromes).

■ Table 3.61 provides information on the differential diagnosis in acutely occurring contractures without any underlying systemic disorder.

### Treatment

While the treatment of acute knee contractures should always be based on the underlying cause (and is addressed in the corresponding chapters), we shall confine ourselves at this point to the treatment of chronic, fixed, severe contractures in connection with systemic disorders. Such treatments are most commonly required in arthrogryposis, for which various conservative and surgical solutions are available. *Conservative treatment* options include the Quengel cast or splint. This is a leg splint with a hinged joint at knee level and an extension rod to which the lower leg section can be attached with increasing pressure (▶ Chapter 3.3.7).

■ Table 3.61. Differential diagnosis of acquired knee contractures

History	Clinical features	Affected structured	Additional investigations	Differential diagnosis
<b>Locking</b>				
Recent trauma	Effusion, instability	Capsular ligamentous apparatus	poss. aspiration, radiography	Ligament lesion (▶ chap. 3.3.8)
	Giving way	Menisci		Meniscal lesion (▶ chap. 3.3.8)
	Inability to walk	Retinacula Bone		Patellar dislocation (▶ chap. 3.3.5) Fracture (▶ chap. 3.3.9)
No recent trauma	Effusion (poss.)	Menisci	Arthroscopy	Meniscal lesion (bucket-handle) (▶ chap. 3.3.8), Discoid meniscus (▶ chap. 3.3.6)
	Instability, effusion, giving way	Trapped cruciate ligament remnants	Arthroscopy	Cruciate ligament lesion (chap. 3.3.8)
	Pain on external rotation	Femoral condyles	Radiography (tunnel view)	Osteochondrosis dissecans (▶ chap. 3.3.4)
<b>Contracture of gradual onset</b>				
	Effusion with/without fever	Synovial membrane	CRP, ESR, blood count, serology, bacteriology	Rheumatoid arthritis (▶ chap. 3.3.11)
		Bone/cartilage	Joint aspiration	Infectious arthritis
			Radiography, ultrasound	Osteomyelitis near the joint (▶ chap. 3.3.10)
	Effusion, capsule swelling/thickening	Synovial membrane	MRI	Tumor in the joint (joint chondromatosis, pigmented villonodular synovitis; chap. 3.3.12)
Hardening, bulging, thickening	Quadriceps, hamstring muscles, calf muscles	MRI	Soft tissue tumor (hemangioma, desmoid, sarcoma etc.; ▶ chap. 3.3.12, 4.5.4)	
Shortening	Hamstring muscles Gastrocnemius muscle Quadriceps femoris muscle	Neurological investigations	Neuromuscular contracture (occurs both in primarily spastic and primarily flaccid paralyzes (chap. 3.3.7)	

### 3.3.13 · Knee contractures

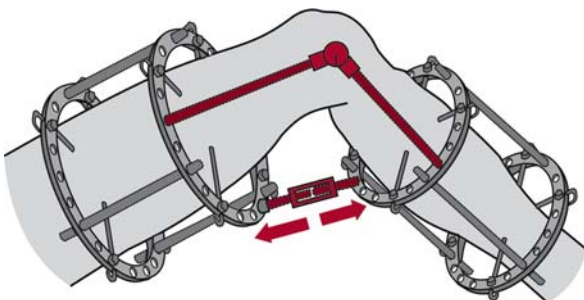
Physiotherapy is an important conservative treatment option, although it must be borne in mind that, if the dorsal structures (muscles and soft tissues) are contracted, forced extension will always produce excessively high pressures on the ventral side of the knee. This limits the options for conservative treatment.

Various *surgical treatments* have been proposed [2, 9, 10]: lengthening of the hamstring muscles, division of the shortened, dorsal soft tissue structures [5], epiphysiodesis of the anterior part of the distal femoral epiphyseal plate and a femoral or tibial extension osteotomy. While soft tissue operations cannot achieve any lasting effect in cases of severe contractures (particularly in arthrogryposis), extending osteotomies are effective [3], albeit at the expense of a permanent alteration in joint anatomy.

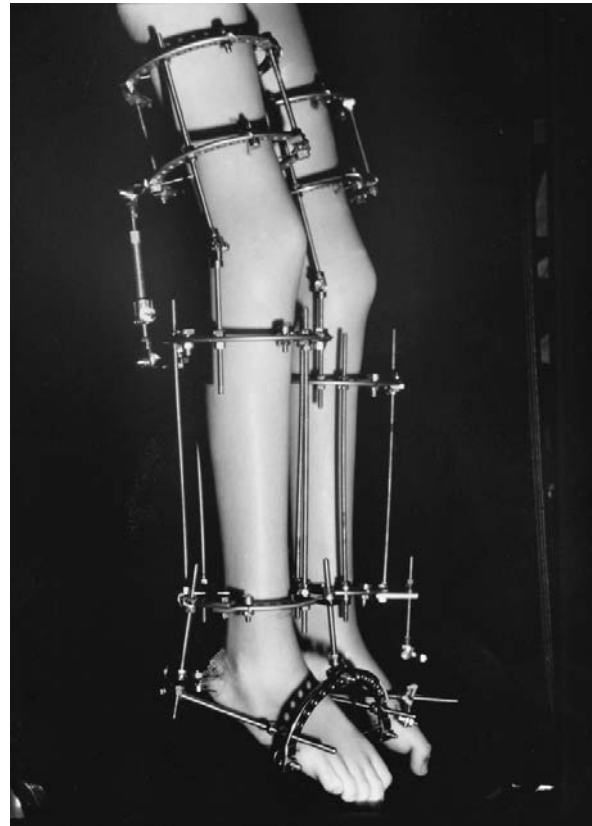
Since 1989 we have therefore used the *Ilizarov apparatus* to correct severe knee contractures [1]. At that time, this apparatus was already being used successfully for the correction of complex foot deformities [4, 6, 7]. The method involves the fitting of 2 circular rings to both the upper and lower leg, the linking of these ring systems with 2 lateral hinged joints and a dorsal distraction rod and a ventral compression rod (■ Fig. 3.347 and 3.348). We have treated 30 knees by this method to date. Fifty percent of the patients were suffering from arthrogryposis (■ Fig. 3.349). The flexion contracture was improved, on average, from 40° preoperatively to 6° postoperatively, although a subsequent deterioration to 18° was noted at the follow-up control after 3 years [1].

Recurrences can be expected as the child grows, particularly in those with arthrogryposis, since this condition involves impaired muscle growth and the muscles are unable to keep pace with the lengthening of the skeleton. Specific problems associated with the treatment of contractures in spastic cerebral palsies and flaccid paralyses are discussed in ► chapter 3.3.7.

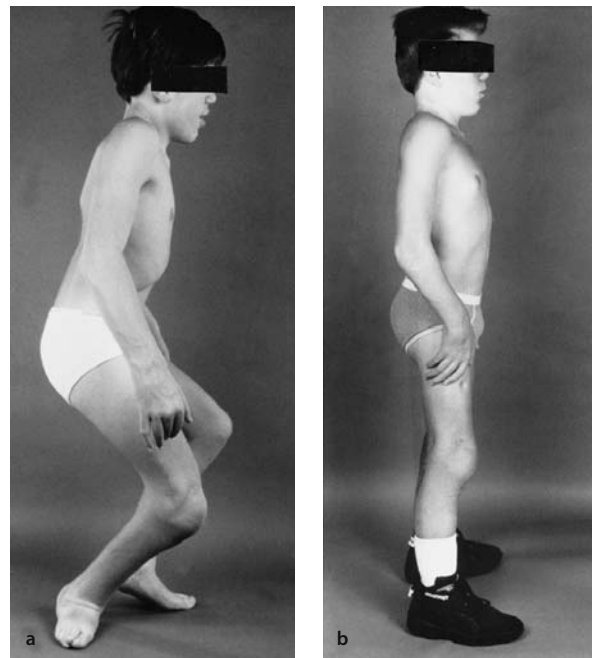
More recently we have started using the Tailor Spatial Frame for the correction of severe flexion contractures of the knee. This apparatus allows a more precise definition of the axis of rotation.



■ Fig. 3.347. Principle of correction of knee contracture with the Ilizarov apparatus



■ Fig. 3.348. Legs of a 16-year old girl with arthrogryposis and fitted Ilizarov apparatus on both sides for the correction of knee contractures



■ Fig. 3.349. 14-year old patient with arthrogryposis and severe knee contractures. a Preoperatively. b Postoperatively after extension with the Ilizarov apparatus

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### 3.3.14 Differential diagnosis of knee pain

Table 3.62 shows the differential diagnosis of knee pain.

Table 3.62. Differential diagnosis of knee pain				
History	Clinical features	Affected structured	Additional investigations	Differential diagnosis
<b>Joint effusion present</b>				
<b>Trauma present</b>	Swelling, instability  Giving way Locking Inability to walk	Capsular ligamentous apparatus  Menisci Bone	Depending on the individual situation: aspiration, radiography	Ligament lesion  Meniscal lesion
<b>No trauma</b>	Effusion With/without fever	Synovial membrane Bone/cartilage	CRP, ESR, blood count Serology, bacteriology Joint aspiration Radiography	Rheumatoid arthritis Infectious arthritis Osteomyelitis near the joint
<b>No joint effusion</b>				
<b>After exercise</b>	Possibly circumscribed swelling	Prepatellar or anserine bursa	–	Bursitis
<b>After exercise</b>	Pain on external rotation	Femoral condyles	Radiography (tunnel view)	Osteochondrosis dissecans
<b>After exercise</b>	Tenderness of tip of patella	Tip of patella	Knee x-rays: AP and lateral	Sinding-Larsen, jumper's knee
<b>After exercise</b>	Tenderness Tibial tuberosity	Tibial tuberosity	Possibly lateral x-ray	Osgood-Schlatter disease
<b>After exercise (particularly downhill)</b>	Tenderness patella	Patella	Possibly radiography	Patellofemoral syndrome
<b>After exercise</b>	Tenderness of medial femoral condyle	Synovial membrane	–	Mediopatellar plica (medial shelf)
<b>After exercise in popliteal fossa</b>	Bulging in popliteal fossa	Connective tissue	–	Popliteal cyst
<b>Giving way during exercise, pseudolocking</b>	Hypermobility of the patella	Patella	Knee x-rays: AP and lateral, axial view of patella, poss. CT	Habitual or recurrent dislocations of the patella
<b>Giving way during exercise (poss. no recollection of trauma)</b>	Instability (Lachman positive, lateral opening)	Ligamentous apparatus	Possibly x-ray with knee held in position	Ligament lesion
<b>Snapping of knee during exercise</b>	Possibly snapping elicited	Menisci	Possibly arthroscopy	Lateral discoid meniscus
<b>At night, on one side</b>	Possibly palpable bulge	Bone	Radiography, bone scan	Tumor
<b>At night, alternating sides</b>	No abnormal findings	Periosteum?	–	»Growing pains«



### 3.3.15 Indications for imaging procedures for the knee

The indications for imaging procedures for the knee are shown in **Table 3.63**.

<b>Table 3.63. Indications for imaging procedures for the knee</b>		
<b>Tentative clinical diagnosis</b>	<b>Circumstances/Indication</b>	<b>Imaging procedures</b>
Fracture	Trauma	Knee: AP and lateral (poss. axial view of patella)
Collateral ligament lesion	Trauma (bony avulsion? Stieda-Pellegrini shadow?)	Knee: AP and lateral
Meniscal lesion	Trauma (only if concurrent fracture suspected)	Knee: AP and lateral, possibly MRI
Osteochondritis dissecans	Locking	Knee: AP and lateral, tunnel view according to Frick
Dislocation of the patella	Typical incident	Knee: AP and lateral, axial view of both patellas, poss. CT in extension with and without tensing of the quadriceps
Tumor	Pain, swelling	Knee: AP and lateral, possibly bone scan, possibly MRI
Inflammation	Pain, fever, positive laboratory result	Knee: AP and lateral, possibly bone scan
Growing pains	If atypical (e.g. unilateral)	Knee: AP and lateral, possibly lower leg/thigh
Avascular bone necrosis (Osgood-Schlatter disease, Sinding-Larsen disease)	Exclusion of other lesions	Knee: AP and lateral
Axial deviations	Asymmetrical genua vara/valga, Blount disease?	Knee: AP and lateral

### 3.3.16 Indications for physical therapy in knee disorders

The indications for physical therapy in knee disorders are presented in **Table 3.64**.

<b>Table 3.64. Indications for physiotherapy in knee disorders</b>				
<b>Disorder</b>	<b>Indication</b>	<b>Goal/type of treatment</b>	<b>Duration</b>	<b>Additional measures</b>
Osgood-Schlatter disease	Pain	Alleviate pain  Strengthen the muscles  (Electrostimulation, quadriceps training)	12 sessions	Swimming, knee protection, warmth  Warmth  Knee support, poss. cylinder cast
Anterior knee pain	Pain	Strengthen the muscles  (Electrostimulation, quadriceps training, esp. vastus medialis)	12 sessions	Taping, knee support  (e.g. Genutrain), possibly temporary cylinder cast
Ligament instability	Instability, giving way	Strengthen the muscles  (Electrostimulation, training of the hamstring muscles)	As long as progress is still possible	Possibly exemption from gym classes; cycling and swimming
Postoperatively	After any operation	Strengthen the muscles, improve range of motion, improve gait pattern	As long as mobility is restricted, muscle atrophy is present and progress is still possible	Exemption from gym classes; cycling and swimming at a later stage

### 3.4 Foot and ankle

#### 3.4.1 Examination of the foot and ankle

##### 3.4.1.1 Infants

###### History

- Family history:  
Are there any known foot abnormalities in the family (clubfoot occurs with increased frequency in certain families, while polydactyly, syndactyly and split foot can be inherited, although most cases occur sporadically)?
- Birth history:  
Clubfoot is commonly associated with a breech presentation.

###### Inspection

*Abnormalities* of the foot that can be diagnosed at birth are usually also apparent on visual inspection. Thus, *polydactyly*, *syndactyly* and *split foot* are readily visible externally, as are *abnormalities of the great toes* (▶ Chapter 3.4.5). *Clubfoot* also shows a very characteristic picture, with adduction of the forefoot, marked varus of the hindfoot, an elevated calcaneus and an equinus foot position (▶ Chapter 3.4.3). Diagnosis by visual inspection is not always so easy for *congenital flatfoot (vertical talus)*. Here too the calcaneus is elevated, but the forefoot is usually abducted and pronated (▶ Chapter 3.4.4).

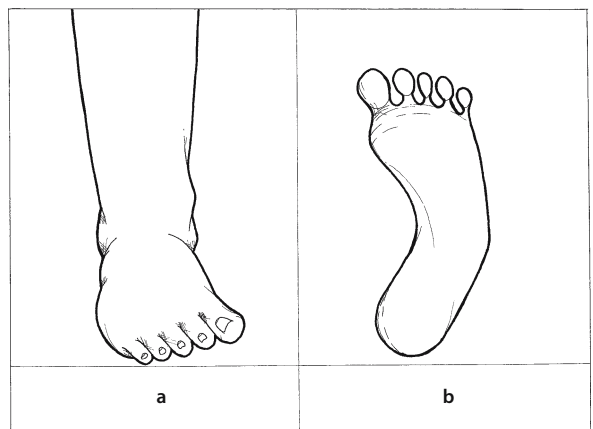
In addition to the actual abnormalities, *postural disorders* are also usually observed in the infant feet. *Pes calcaneus* is very frequently present at birth. In this condition, the foot is extended dorsally to its maximum extent, causing the back of the foot to touch the lower leg (■ Fig. 3.350). Although *metatarsus adductus*, or *pes adductus*, occurs as a congenital condition, it usually appears only during the first few months of life. It is characterized by adduction of the forefoot in relation to the rearfoot. This is best viewed from below (■ Fig. 3.351). The axis of the whole foot, or the rearfoot, in relation to the upper leg should always be evaluated at the same time (■ Fig. 3.352).

###### Palpation, examination of the range of motion

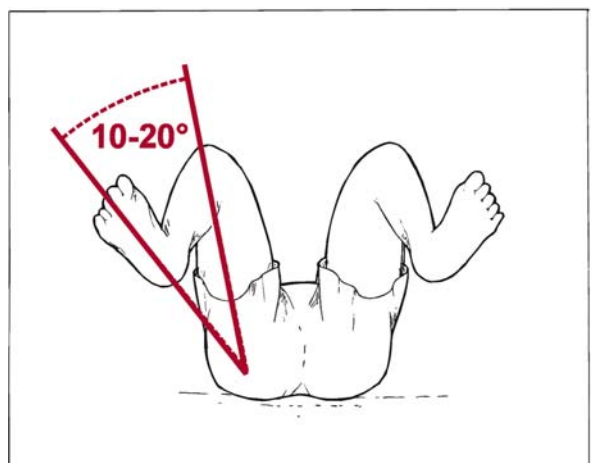
*Palpation* of the *calcaneus* and *talus* is important for diagnosing an elevated calcaneus or vertical talus. In the latter condition, the talus is very prominent on the plantar aspect of the foot. The *mobility* of the ankle and the subtalar joint is investigated according to the same procedure employed for children and adolescents (see below). The examination of the infant foot with abnormalities or postural disorders includes an *evaluation* of the correctability. If clubfoot or metatarsus adductus with adduction of the forefoot is present, the examiner grasps the heel with one hand while the other hand applies pressure to the forefoot in a medial to lateral direction (■ Fig. 3.353). If the normal position can be achieved with moderate pressure, the foot is correctable, otherwise not. A more detailed description of the



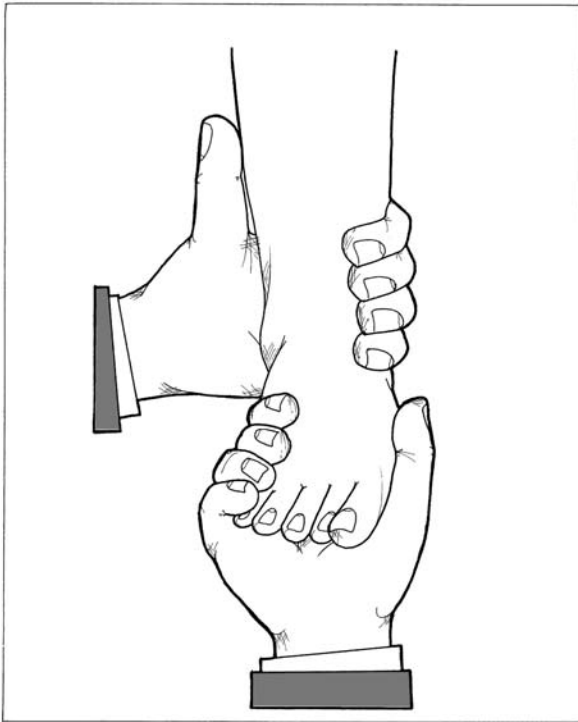
■ Abb. 3.350. *Pes calcaneus*: The back of the foot can strike the anterior edge of the tibia



■ Fig. 3.351a, b. *Metatarsus adductus*: The forefoot is adducted in relation to the rearfoot



■ Fig. 3.352. Determination of the *foot axis* in relation to the femoral axis in the supine infant



■ Fig. 3.353. Evaluating the *correctability* of the forefoot adduction. The examiner grasps the heel with one hand while the other hand corrects the forefoot in a medial to lateral direction

correctability of a clubfoot and its classification is provided in ► chapter 3.4.3. In a case of pes calcaneus, the examiner tests whether the foot can be pressed into plantar flexion or whether just the neutral position is achievable.

❗ **The examiner should not test whether an equinus foot position is correctable, since excessive pressure applied to the forefoot in a dorsal direction can damage the talus. The Achilles tendon is stronger than the small bones of the rearfoot, which are still in a cartilaginous state at this stage.**

*Stimulation* forms another part of the examination. Particularly in patients with metatarsus adductus, tickling of the lateral edge of the foot will activate the peroneal muscles, causing the child spontaneously to correct, at least partially, the metatarsus adductus position.

#### Other investigations

Many foot abnormalities are associated with *other deformities*. Since *hip dysplasia* and *clubfoot* often occur together, an ultrasound scan of the hips is always indicated in such cases. Clubfoot is also observed in connection with *arthrogryposis* and *diastrophic dwarfism*. Macrodactyly can be associated with the *Klippel-Trenaunay* and *Proteus syndromes*. Characteristic malformations of the great toes are observed in *fibrodysplasia ossificans progressiva*, *diastrophic dwarfism* and in *Rubinstein-Taybi syndrome*.

#### 3.4.1.2 Children and adolescents

##### Examination protocol for the upper ankle and foot in children and adolescents

The examination protocol is shown in ■ Table 3.65.

##### History

- Has trauma occurred? If so,
  - When did the trauma happen?
  - During what type of activity did the trauma happen (sport, play, daily routine)?
  - Is a direct or indirect injury involved?
  - What type of movement was involved (supination, eversion)?
- Pain history: Where is the pain located? When does it occur? Is the pain load-related, movement-related, or does it also occur at rest or even at night? If so, does the pain only occur when the patient changes position or does the patient awake at night because of the pain? For movement-related pain: What specific movements elicit the pain (dorsal extension, plantar flexion, inversion, eversion)?

##### Inspection

##### Examination of the walking patient

- Is there a *limp* (protective limp or stiff limp)? (see also ► chapter 4.2.3)?
- Does the foot roll normally from heel to toe, i.e. with a normal heel-ball gait, do the heel and forefoot strike the ground at the same time or is an equinus or steppage gait present? In a patient with an *equinus gait* neither active nor passive dorsal extension is possible, whereas only active dorsal extension is lacking in a patient with a *steppage gait*, which is associated with paralysis. Footdrop occurs in the latter case, but the feet can be extended passively in a dorsal direction. While the heel is not loaded at all in an equinus gait, heel loading also occurs secondarily in a steppage gait after the forefoot has struck the ground (ball-heel gait). The examiner must also observe whether the foot-strike is plantigrade or whether the foot supinates, placing most of the load on the lateral edge. The opposite picture, i.e. hyperpronation of the forefoot with hypervalgus of the rearfoot also occurs.
- The examination of the walking patient also includes observation of the *knees*. During the stance phase are the knees *extended* normally (i.e. approx. 5–10° flexion), *overextended* (complete extension or even hyperextension) or *insufficiently extended* (remain in flexion of more than 10°)?
- Examining the patient while *walking on tiptoes* and *heels* is also useful, as this is a quick and simple way of establishing whether coarse motor function is normal or not.
- See also ► chapter 2.1.3 for further information on gait analysis.

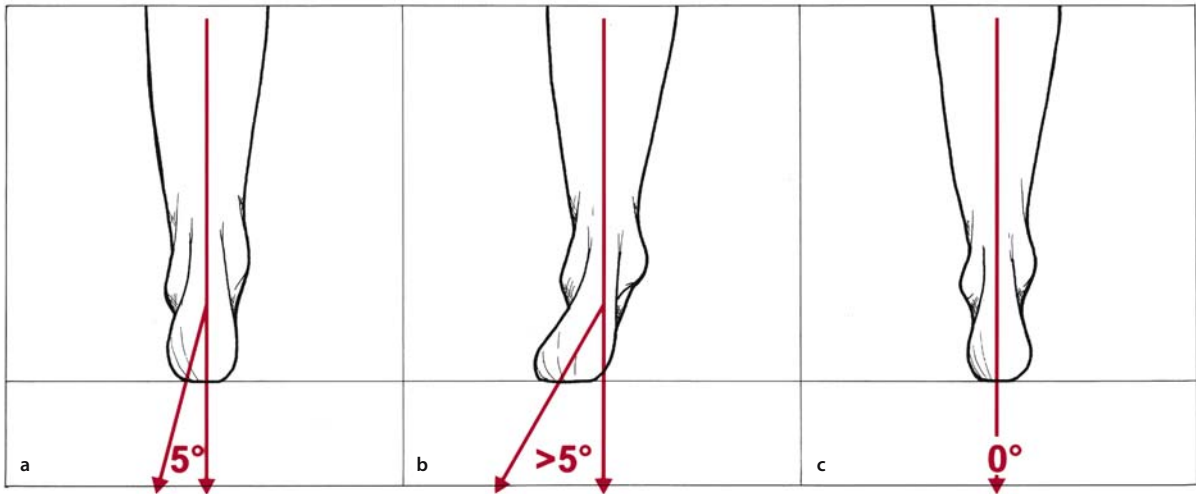
■ Table 3.65. Examination protocol for the upper ankle and foot

	Examination	Question
I. History	Ask about trauma	Type of trauma? Inversion or eversion?
	Pain history	Location? Duration? Load-related?
	Ask about stability	Feeling of instability? Frequent giving way (supination)?
II. Inspection	Gait pattern	Limping (protective, stiff?) Foot roll (heel-ball gait, equinus gait, steppage gait)? Extension of the knees? Walking on tiptoes and heels?
	Contours	Swelling, redness, bulging?
	Lower leg	Normal, atrophy?
	Rearfoot axis	Normal, valgus/varus position?
	Forefoot	Shape (Greek, Egyptian?) Hallux valgus, varus? Superduction?
	Medial longitudinal arch of the foot	Normal, lowered, medial weight-bearing, elevated, footprint?
III. Palpation	Tenderness	Calcaneus, malleoli, talus, navicular, forefoot
	Joint space in upper ankle	Effusion, capsule swelling?
	Para-articular soft tissues	Swelling? Tumor?
IV. Range of motion	Ankle joint	Dorsal extension/plantar flexion with extended (possibly also flexed) knee, active and passive
	Subtalar joint	Valgus and varus movement
	Forefoot	Pronation/supination
	Whole foot	Inversion/eversion
V. Stability	Lateral stability of ankle and subtalar joint	Forced inversion
	AP stability of upper ankle	Anterior drawer test

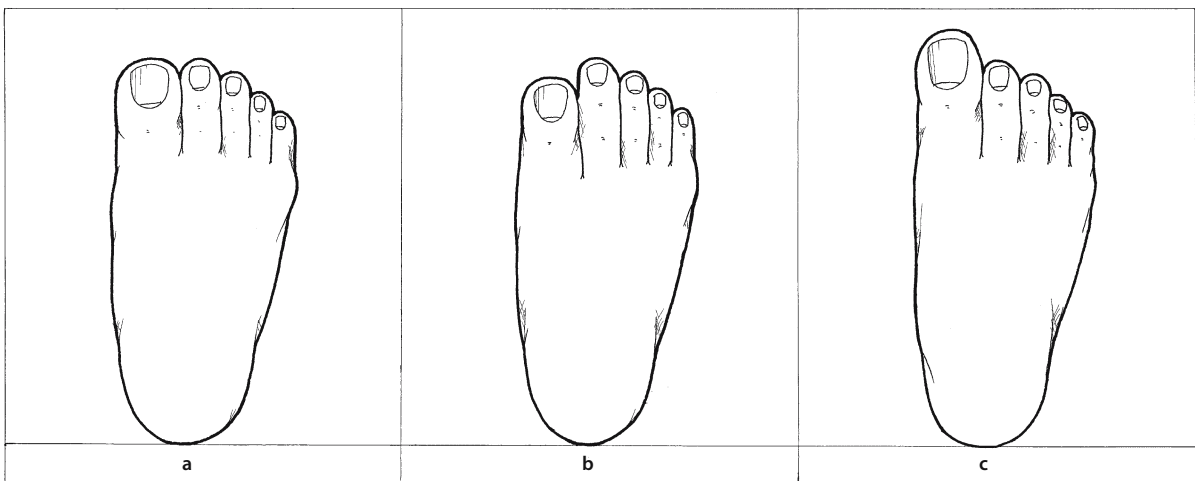
### Examination of the standing patient

- Is there any *swelling, redness* or *bulging*?
- Inspection of the *lower leg*: Do the calf muscles appear normal or atrophied?
- Observation of the *rearfoot axis*: Is this in a physiological valgus position of approx. 5°, or is a hypervalgus (>5°) or calcaneus varus (0° or less) present (■ Fig. 3.354)?
- *Shape of the forefoot*: Is it normal, thin or widened (splayfoot)? Variants of the forefoot are also observed in respect of *toe length* (■ Fig. 3.355). The *position of the great toe* must also be noted: neutral position, valgus deviation (in the metatarsophalangeal or interphalangeal joints?) or even a varus position. Any *superduction* or *subduction* of individual toes should also be noted.
- *Medial longitudinal arch of the foot*: Observe whether the medial arch of the foot is elevated as normal, touches the ground or is too high (■ Fig. 3.356).
- Evaluation of the *footprint*: The footprint under load can be visualized either on the podoscope (a glass plate mounted above a mirror; ► chapter 3.4.7) or with an ink-colored rubber plate placed on a piece of white paper. Alternatively, the whitened parts of the skin in the loaded zone can be inspected immediately after the foot is lifted off the ground. The callosity on the foot provides information about functional weight-bearing. The footprint (■ Fig. 3.357) is an important criterion for evaluating the formation of the longitudinal and transverse arches of the foot.

## 3.4.1 · Examination of the foot and ankle



■ Fig. 3.354a–c. *Rearfoot axis*: a in a physiological valgus position of approx. 5°, b in a hypervalgus position, c in a pathological varus position (neutral axis or genuine varus axis)

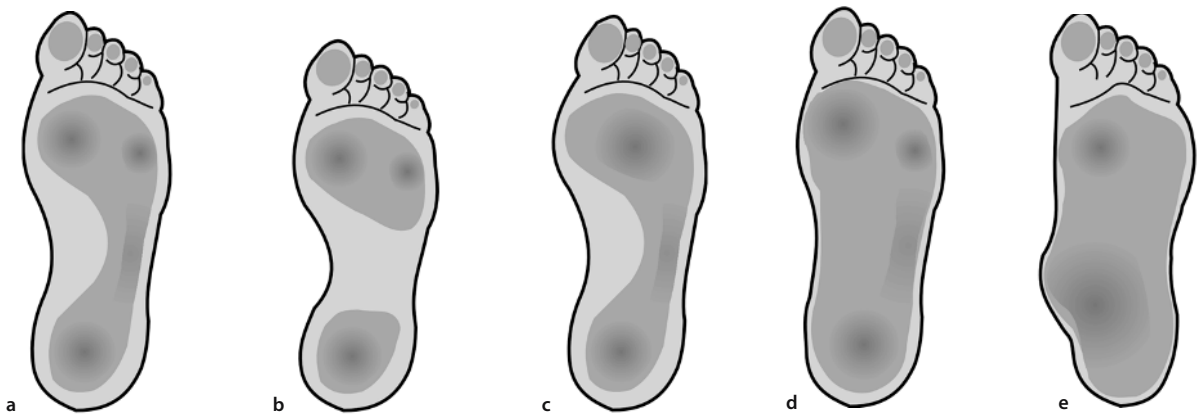


■ Fig. 3.355a–c. *Forefoot variants*: a intermediate foot (1st and 2nd toes roughly the same length), b Greek foot (2nd toe longer than the 1st), c Egyptian foot (1st toe longer than the 2nd)



■ Fig. 3.356a–c. *Medial arch of the foot from the medial side*: a normal foot (or »flat valgus foot«), b flexible flatfoot, c pes cavus





**Fig. 3.357a–e.** Footprints: a normal foot with callusing under the heel and the 1st and 5th metatarsal heads; b pes cavus with no weight-bearing in the metatarsal area; c splayfoot with widening of the forefoot and callus formation predominantly under the 2nd and

3rd metatarsal heads (rare in children and adolescents); d flexible flatfoot with a missing medial arch, but otherwise normal weight-bearing pattern; e heavy, rigid flatfoot with principal weight-bearing on the medial side in the midfoot area (under the talus)

### Palpation

#### Examination of the supine patient

- **Tenderness:** Typical painful sites in children and adolescents are the *heel* (in calcaneal apophysitis), the *lateral malleolus* and the *talar neck* (in injuries or instability of the lateral ligaments), the lateral aspect of the *talus* and *calcaneus* (in talocalcaneal coalition), the *navicular bone* (in cornuate navicular, accessory navicular or Kohler's disease), the *first metatarsal head* (in juvenile hallux valgus) and the 2nd, 3rd or 4th *metatarsal heads* (in Freiberg's disease or a stress fracture).
- **Palpation of an effusion:** Bulging of the joint capsule is readily observed and palpated in the ankle joint. Effusions occur after recent trauma, in juvenile rheumatoid arthritis or in hemophilia.
- Swelling of the *para-articular soft tissues* is observed after trauma and in association with inflammation and tumors.
- **Circumference measurement of the lower leg:** The circumference is measured with a tape measure at its maximum point.

#### Range of motion

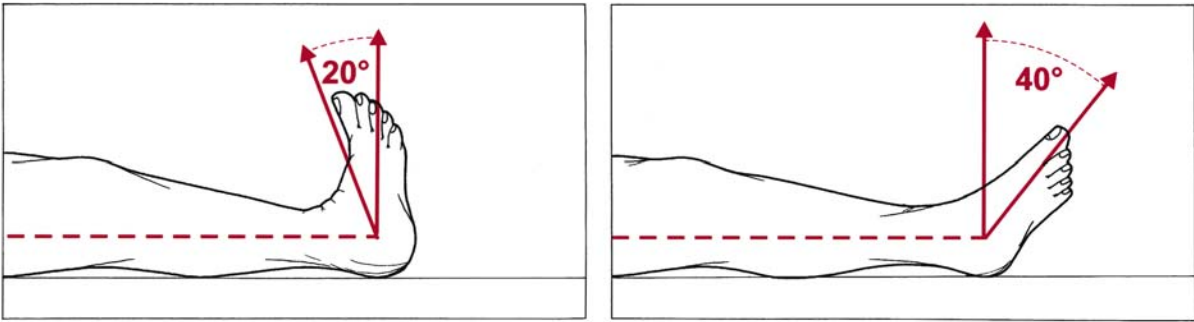
**!** Both sides should always be measured when examining mobility in the upper and lower ankle.

- **Neutral-0-position** = plantigrade foot.
- **Ankle joint: dorsal extension/plantar flexion:** The patient is examined in the supine position with the knee extended. *Passive:* The examiner grasps the forefoot and pushes it as far as it will go in the dorsal and plantar directions (Fig. 3.358). *Active:* The patient is asked to perform the same movement himself. In

functional respects, it is much more important to perform this examination with the knee extended rather than flexed, since the knee is extended during walking. Dorsal extension is restricted in the extended knee when the two-joint gastrocnemius is contracted. The normal range of motion is as follows: dorsal extension/plantar flexion = 20–0–40.

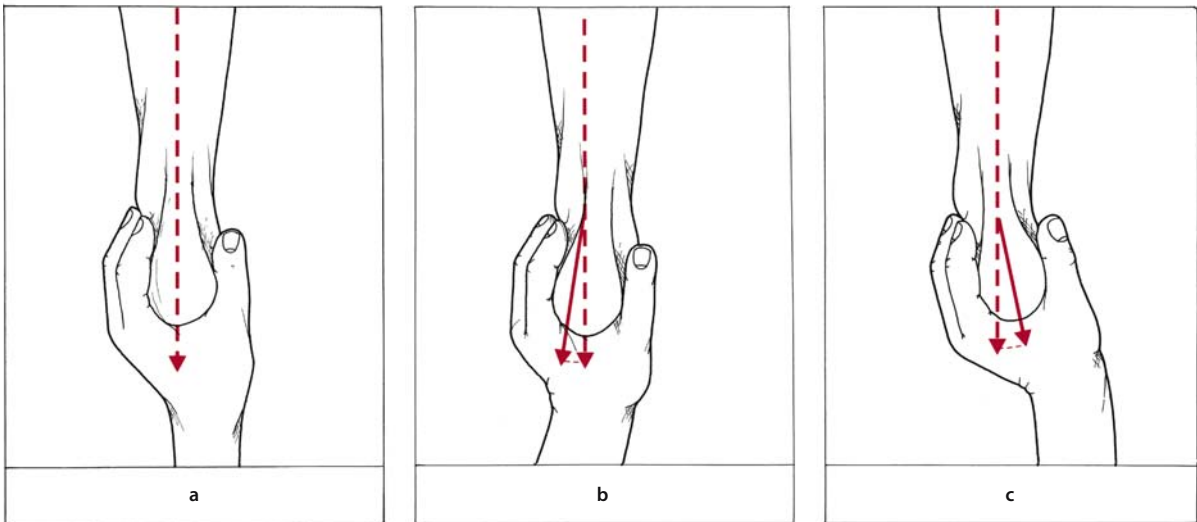
- **Subtalar joint:** The *varus* and *valgus* movement is tested. Grasping the lower leg with one hand, the examiner grasps the calcaneus with the other and turns it inwardly and outwardly (Fig. 3.359). The test is not very precise and angles specified in degrees are not very useful. We describe simply whether the movement is normal, slight or very restricted, or whether the joint is completely locked.
- **Forefoot:** *Pronation* and *supination* are measured. One hand firmly grasps the heel while the other turns the forefoot. Angles can be specified with this test since the plane of the forefoot in relation to the perpendicular to the lower leg can be determined very precisely. The normal range of motion is roughly as follows: pronation/supination = 20–0–30 (Fig. 3.360).
- The *combined rotational movement of the fore- and rearfoot* is termed *eversion* and *inversion*, and is tested by grasping the lower leg with one hand, the forefoot with the other and rotating both inwardly and outwardly. Since this test is likewise not very precise, we restrict ourselves to descriptions such as »normal«, »increased« (in instability), »slight«, »greatly restricted« or »locked«.
- **Toes:** *Dorsal extension* and *plantar flexion* in the metatarsophalangeal joint, and possibly the interphalangeal joints can be measured at this point.

## 3.4.1 · Examination of the foot and ankle



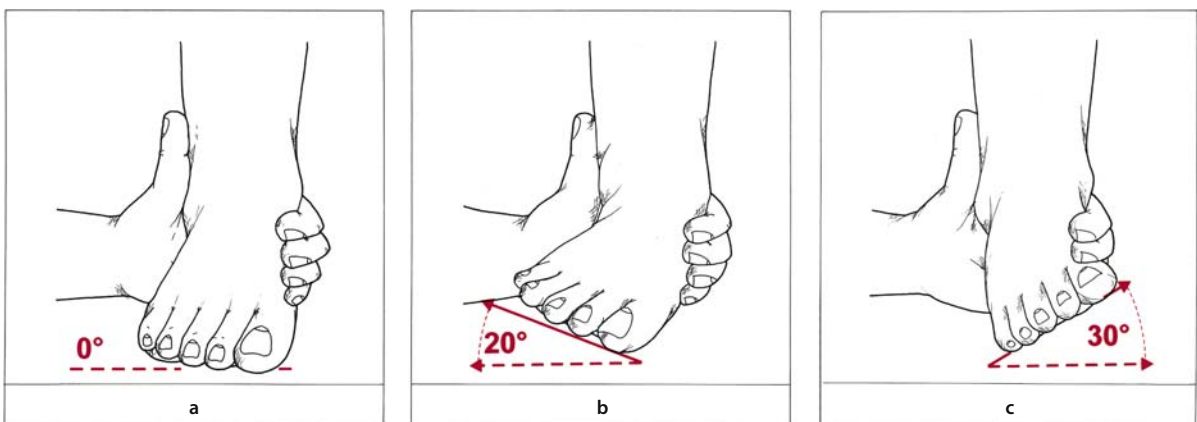
■ Fig. 3.358. Examination of *Range of motion in the ankle joint*. Dorsal extension and plantar flexion can be examined both with the knee flexed and extended. The extent of dorsal extension is always slightly greater with the knee flexed than extended because of the relaxed

gastrocnemius muscles. In functional respects, however, the examination with the knee extended is more important, since walking takes place in this position



■ Fig. 3.359a–c. Examination of *Range of motion in the subtalar joint*. a The heel is grasped with one hand and turned inwardly (b inversion) and outwardly (c eversion) in relation to the lower leg. Normally,

around twice as much inversion as eversion is possible. Stating the result in degrees is not very useful. The examiner should simply state whether the movement is normal, restricted or completely absent.



■ Fig. 3.360a–c. Examination of *mobility in the forefoot*. One hand stabilizes the heel (a), while the other rotates the forefoot inwardly (b pronation, 30–40°) and outwardly (c supination, 10–20°).

**Stability testing**

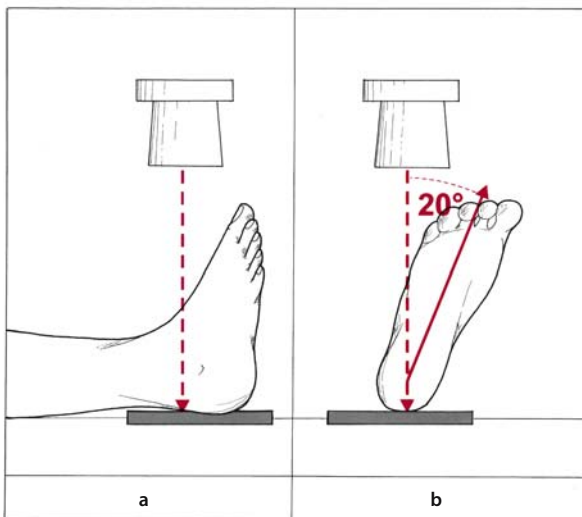
- *Test for lateral opening in the ankle:* The examiner grasps the lower leg with one hand and the foot with the other and attempts maximum inversion of the foot. If inversion is greater than normal, then instability is present, although it is not possible to differentiate between instability of the ankle and subtalar joint, for which a separate test for valgus and varus movement in the subtalar joint is required.
- *Anterior drawer test in the ankle joint:* The examiner grasps the lower leg with one hand and the rearfoot with the other and presses the latter forward and backward in relation to the lower leg. The movement is perceived in the hand and takes place in the ankle joint. This is always pathological and a sign of instability.

**Reference**

1. Debrunner HU (1982) Orthopädisches Diagnostikum. Thieme, Stuttgart, New York, pp. 102–105

**3.4.2 Radiographic techniques for the foot and ankle****Ankle joint: AP and lateral**

The patient lies in the supine position with the heel resting on a cassette. The foot forms a right angle in relation to the lower leg and is rotated inwardly by 20°, because this compensates for the physiological external rotation of the tibia and positions the malleoli at right angles to the x-ray beam (■ Fig. 3.361). The central beam is directed at the center of the ankle joint, i.e. 1 cm above the tip of



■ Fig. 3.361a, b. Recording the AP x-ray of the ankle. The foot must be rotated inwardly by 20° so that the ankle mortise is at right angles to the x-ray beam

the medial malleolus. For the lateral view, the patient is placed on the side to be viewed and the beam is aimed in a mediolateral direction. The central beam is directed on the medial malleolus.

**Ankle joint inclined at an angle of 45° internal and external rotation**

These views facilitate better evaluation of tears in the syndesmosis and of obliquely running fracture lines in joint fractures. The ankle joint is positioned and centered as for the AP view with a foam wedge angled at 45° on each side.

**Foot: DP (AP)**

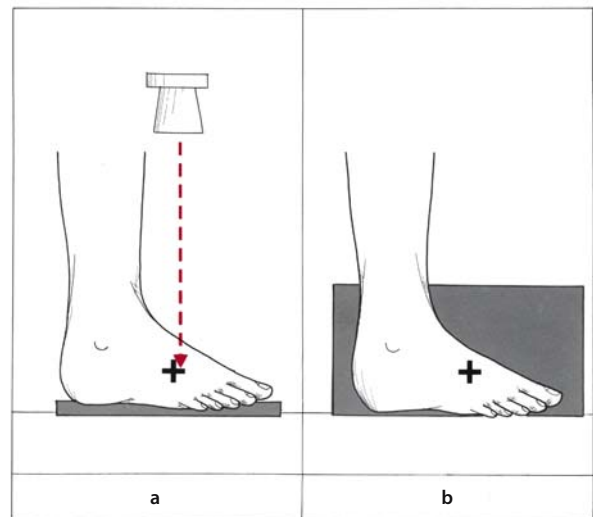
For the dorsoplantar view the patient sits on the x-ray table, with the hip and knee flexed and the sole of the foot on the same side resting on the cassette. The central beam is directed at the proximal end of the 3rd metatarsal (■ Fig. 3.362). An aluminum wedge filter over the forefoot prevents overexposure of the toes.

**Foot: lateral with the patient standing and weight-bearing**

The patient stands on a small wooden platform and the cassette is placed between both feet in a small slot in the platform. The central beam is aimed at the proximal end of the 4th metatarsal and travels in a lateromedial direction.

**Forefoot: AP oblique**

The lateral edge of the foot is raised by approx. 45° to simulate pronation. The central beam is directed at the proximal end of the 3rd metatarsal (■ Fig. 3.363). The



■ Fig. 3.362a, b. AP and lateral x-rays of the foot while seated (and standing)

view supplements the AP x-ray, providing a second plane and a clearer view of the structures of the individual rays compared to the lateral view, on which the metatarsals and phalanges are projected on top of each other.

#### Rearfoot: AP oblique

The lateral edge of the foot is raised by approx. 45° to produce eversion (■ Fig. 3.364). This x-ray provides a perfect view of, for example, coalition between the calcaneus and navicular or the talus and calcaneus, thus dispensing with the need for a CT scan.

#### Heel: lateral and axial in the supine position

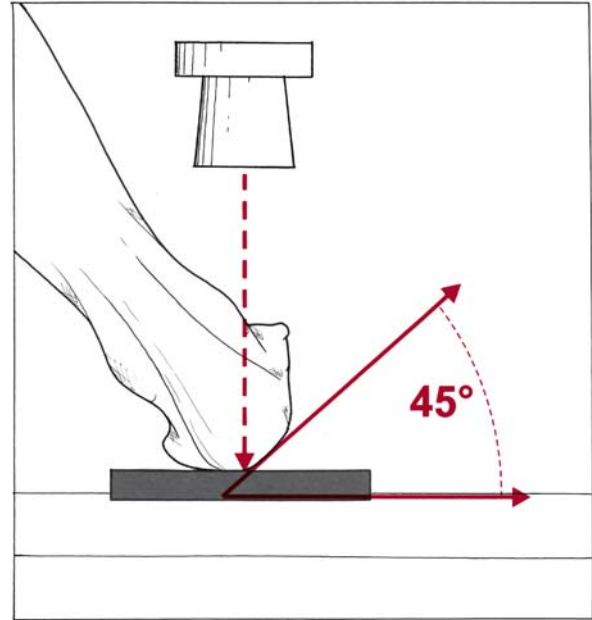
For the lateral view the lateral edge of the foot is placed on the cassette. The central beam is aimed at the center of the calcaneus. For the axial view, the patient lies on his back with the heel resting on the cassette and the foot at 90° to the lower leg. The central beam is aimed at the heel from below at an angle of 45°. Alternatively, the foot can be placed in a position of maximum dorsal extension, causing the central beam to strike the cassette from the cranial direction at an angle of 20° (■ Fig. 3.365).

#### Foot x-rays: AP and lateral in infants with the foot deformities of clubfoot, flatfoot, etc.)

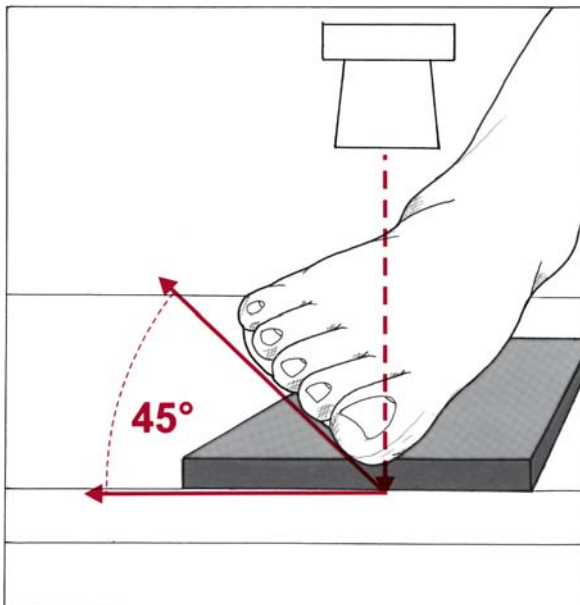
An important requirement here is weight-bearing of the foot in the AP and lateral projections, if possible in a position of corrected or overcorrected dorsiflexion and abduction.

#### AP projection (without correction)

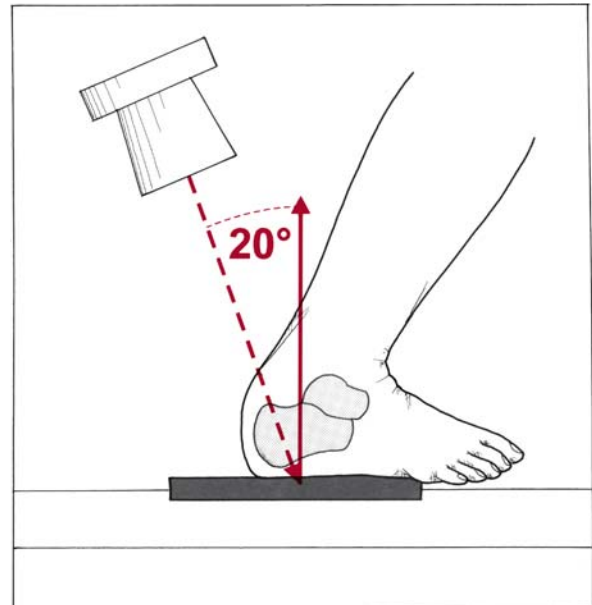
The foot of the lying infant is placed on the cassette and pressure applied via the tibia. The central beam is aimed at the tarsus at an angle of 30° from the caudocranial plane.



■ Fig. 3.364. Oblique x-ray of the rearfoot to visualize the joints between the calcaneus and navicular bone or between the talus and calcaneus



■ Fig. 3.363. Oblique x-ray of the forefoot to visualize the individual metatarsals and phalanges



■ Fig. 3.365. Axial view of the calcaneus

### 3.4.3 Congenital clubfoot

» I owe my life to my feet. They have led me astray into flights of fancy, caused me pain, forced me to read and use my imagination, to overestimate myself, to enthuse about the equality of creatures, to respect the imperfect, and they have rendered me unfit for military service (Hans Dieter Hüsch, German author and satirist, who was born in 1925 with bilateral clubfeet [from: »Du kommst auch drin vor«, Thoughts of a traveling poet, Kindler 1990]). «

#### ► Definition

Congenital abnormality of the foot with equinus and varus position of the rearfoot and adduction and inversion (supination) of the forefoot.

— Synonyms: Pes equinovarus, talipes equinovarus, strephopodia



#### Historical background

The corrective treatment of clubfoot was described very precisely long ago by Hippocrates (370 BC) [36]. He also described the fitting of bandages and corrective shoes.

#### Subsequent dates in the history of clubfoot treatment [36]

- 1574: Francisco Arceo: Description and pictorial presentation of a metal splint for clubfoot treatment.
- 1652: Wilhelm Fabricius Hildanus: Adjustable corrective splints.
- 1780: Jean-André Venel: Foundation of the first orthopaedic institute in Orbe (Switzerland), treatment of clubfoot with his »sabot de Venel«, the archetype of all current orthoses.
- 1784: First open division of the Achilles tendon by a surgeon, described by Moritz Gerhard Thilenius.
- 1972: Ponseti describes his modern concept of conservative clubfoot treatment [31].
- 1982: Crawford introduces the »Cincinnati incision« [7].
- 1987: Grill and Franke describe the use of the Ilizarov apparatus for severe cases of neglected clubfoot [15].

#### Classification

- Congenital clubfoot,
- Clubfoot posture,
- Congenital pes adductus,
- Neuromuscular clubfoot,
- Clubfoot in systemic disorders (e.g. arthrogyposis).

Two classifications for the severity of congenital clubfoot are now in widespread use. One was proposed by Dimeglio et al. [10] in 1995 (■ Fig. 3.366). This covers four grades:

- Grade I: benign, so-called »soft« clubfoot, readily reducible without significant resistance, similar to clubfoot posture),
- Grade II: moderate clubfoot (reducible with a certain degree of resistance),
- Grade III: severe clubfoot (only reducible against strong resistance),
- Grade IV: very severe clubfoot (not reducible, as in arthrogyposis).

The other common classification is that proposed by Pirani (■ Fig. 3.367). This is particularly suitable for monitoring the progress of clubfoot and can provide an indication as to the time of Achilles tendon lengthening. Although a comparative investigation of 4 classification systems found the Dimeglio system to be the most reliable [37], the Pirani classification is more commonly used in association with the Ponseti treatment.

#### Occurrence

The incidence of clubfoot among the white population is between 1.2 [4, 38] and 2.4 [2] per 1,000 births. A male:female ratio of 2:1 has been determined. The incidence varies considerably between races. Clubfoot is particularly rare, for example, among Chinese and Japanese (approx. 0.5/1,000), but common in black people (3.5/1,000 in South Africa), Australian Aborigines (3.5/1,000 [4]) and Polynesians (6.8/1,000 [5]). An interesting finding has been reported in a Danish study. At the start of the study (1979), a frequency of 1.2 per 1,000 births was calculated, but this had increased to 2.4 per 1,000 births by 1994 [2].

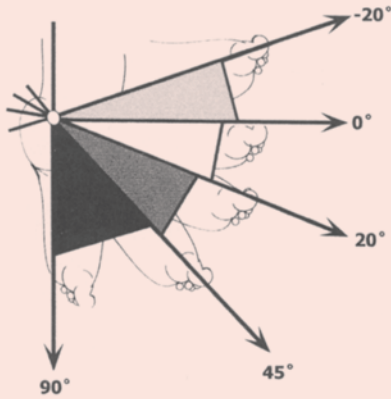
#### Etiology

Both genetic factors [38] and environmental influences during pregnancy play a role in the development of clubfoot. The *genetic component* is polygenic, i.e. the trait is carried not by a single gene but by several genes. More recent studies, however, have postulated a single dominant gene with a penetration of 33% [5, 9, 16]. Family studies have shown that the genetic component is very strong. Thus, the incidence declines from 2.9% if the clubfoot is present among first-degree relatives (siblings) to 0.6% among uncles and aunts, and to 0.2% among cousins [38]. The risk is particularly high if both parents are affected. The familial forms are generally more severe than the sporadic cases.

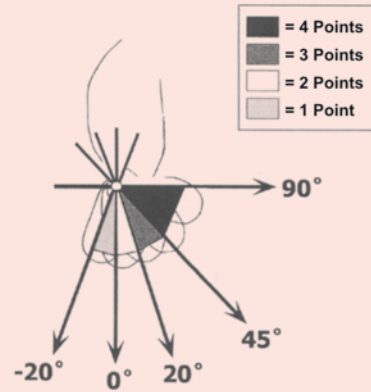


### Classification of clubfeet according to Dimeglio

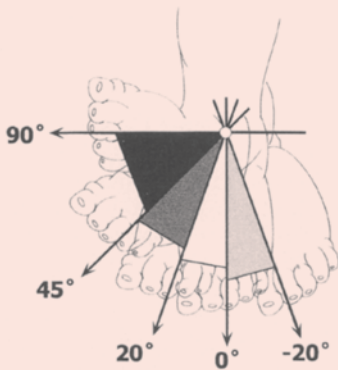
Grade	Type	Occurrence	Points
I	mild	20%	<5
II	medium	33%	5 to 9
III	severe	35%	10 to 14
IV	very severe	12%	15 to 20



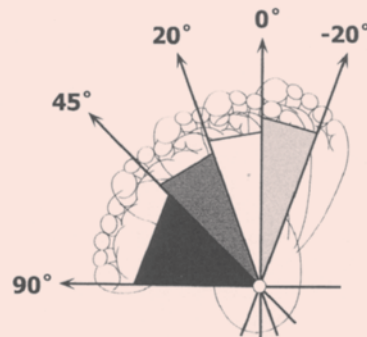
Evaluation of equinus in sagittal plane



Evaluation of varus in frontal plane



Evaluation of rotation of entire foot in horizontal plane



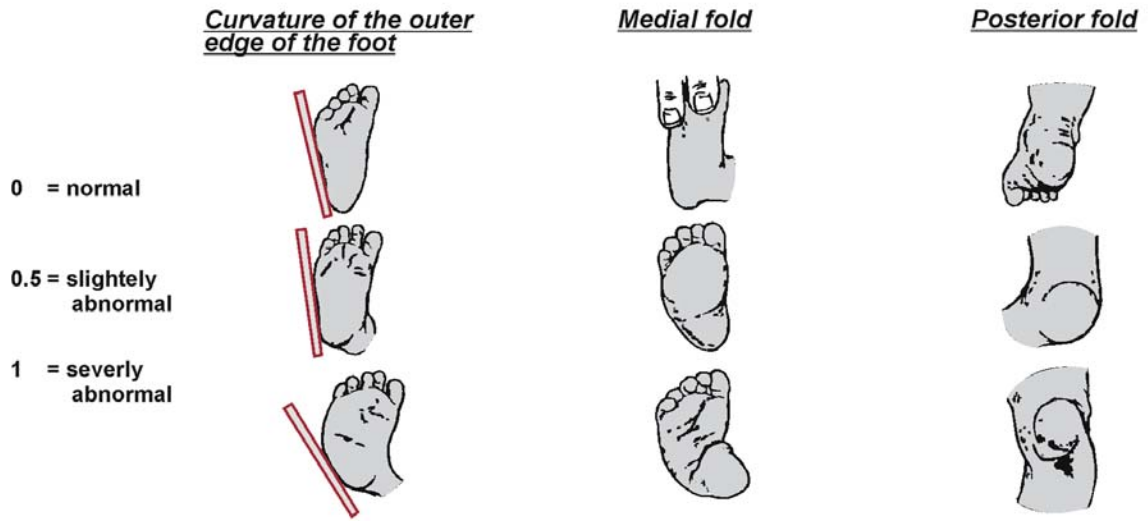
Evaluation of adduction of forefoot in frontal plane

#### Allocation of points

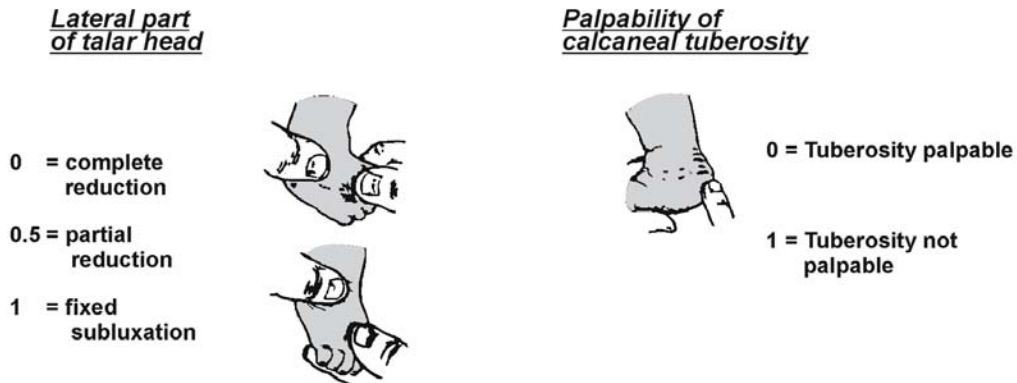
Redressability	Points	Other parameters	Points
90°-45°	4	Posterior groove	1
24°-20°	3	Medial groove	1
20°-0°	2	Cavus	1
<0° to -20°	1	Pronounced muscular atrophy	1

Fig. 3.366. Classification of clubfoot according to Dimeglio: A score of 1-4 is allocated for each parameter, and the total number of points indicates the severity

## SEE



## FEEL



## MOVE

### Rigidity of equinus deformity



Fig. 3.367. Classification of clubfoot according to Pirani. The classification is comprised of a *midfoot score* (curvature of the outer edge of the foot, medial fold and lateral part of the talar head) and a *rearfoot score* (dorsal fold, palpability of the calcaneal tuberosity and rigidity of

the equinus deformity). Each parameter is rated as follows: 0= normal; 0.5= slightly abnormal and 1= abnormal. The classification is particularly suitable for monitoring the progress of clubfoot. Achilles tendon lengthening is indicated if the midfoot score is less than 1

### 3.4.3 · Congenital clubfoot

Studies with fetal cadavers have shown that, *during pregnancy*, the foot remains in a physiological clubfoot position up until the 11th week, with medial rotation, varus of the rearfoot and adduction of the forefoot. Only after this point does the foot achieve its normal position [23]. The fibula grows faster than the tibia up until the 11th week, and tibial growth only starts beyond this point. Clubfoot has been experimentally induced in animals by administering toxins at a time corresponding to the 9th week of pregnancy in humans. A study has also shown that the risk of clubfoot is increased in smoking pregnant women [18]. Many authors have also assumed that milder forms can be induced by an incorrect intrauterine position. But this is countered by the fact that clubfoot does not occur more frequently than usual when the intrauterine space is restricted (twins, high birth weight, oligohydramnios) [38]. Since the introduction of routine ultrasound screening during pregnancy we also now know just how much the embryos move their legs, indicating that an otherwise healthy baby cannot adopt a constant »abnormal posture«.

#### Pathogenesis

Clubfoot is a complex deformity of the whole foot and lower leg involving a wide variety of anatomical structures. The identity of the *primarily affected tissue* has been the subject of much research and speculation. The following hypotheses have been proposed:

- **Neuromuscular defect:**  
The assumption that intrauterine pressure on the peroneal nerve induces clubfoot is now considered to be outdated. On the other hand, abnormal somatosensory and/or motor potentials have been observed in almost half of all clubfoot patients [26].
- **Anomaly of the arteries:**  
Arteriographic investigations involving children with clubfoot have shown that the anterior tibial artery is hypoplastic or that the dorsalis pedis [14] or posterior tibial arteries are missing in the majority of cases [24], although it is not clear whether this is a primary or secondary problem.
- **Shortening of the lateral ligaments** between the fibula and talus or calcaneus:  
These ligaments prevent the forward migration of the fibula in relation to the talus during dorsal extension, resulting in an increasing equinus position.
- **Medial deviation of the talar neck:**  
The medial deviation of the talar neck is the primary deformity, all others are secondary [34]. This theory is now generally accepted.
- **Shortening of the medial ligaments:**  
An investigation of 16 fetuses found fibromatosis in the area of the medial ligaments of the ankle joint (deltoid ligament) as the first change in an incipient clubfoot [13].

#### Pathological anatomy

The following anatomical changes are observed in clubfoot:

- **Talus:**  
The primary deformity in clubfoot consists of a deviation of the anterior section of the talus (talar neck) in a medial and plantar direction (■ Fig. 3.368). The angle between the trochlea of the talus and the talar neck is greater in neonates than in adults, and greater still in clubfoot (■ Fig. 3.369). The talar neck is also shortened and the typical shoulder is lacking. The anterior articular surface is rotated inwardly. The whole talus is smaller than normal and ossification is delayed.
- **Calcaneus:**  
The deformation of the calcaneus is much less pronounced than that of the talus. The calcaneus shows a slight medial deviation and the sustentaculum tali is slightly hypoplastic.
- **Metatarsal and forefoot bones:**  
These are slightly hypoplastic, i.e. shortened but normal in width.



■ **Fig. 3.368.** 3-D reconstruction of a CT scan of a clubfoot (9-year old boy). Note the deviation of the talus and the subluxation of the navicular bone

— **Tibia:**

As we know from ultrasound studies [25], the tibia shows slight internal rotation of approx. 10° on average. However, this internal rotation of the tibia is masked by the posterior displacement of the fibula, giving the impression that the lower leg is externally rotated. Clubfoot patients do not show an increased incidence of rotational deviations of the tibia compared with their normal-footed counterparts [8].

— **Ankle joint:**

The deviation of the talus and the raised position of the calcaneus cause the talus to be pushed forward out of the ankle mortise. A third of the talar joint surface does not articulate in a case of clubfoot. Whether medial rotation also takes place at the same time remains a contentious issue.

— **Subtalar joint:**

The calcaneus is rotated medially and tilted ventro-caudally in relation to the talus, i.e. the normal upward slope in a dorsal to ventral direction is absent.

— **Talonavicular joint:**

The navicular bone is displaced in a medial and plantar direction in relation to the talus. In a pronounced case of clubfoot, the lateral section of the anterior talar surface does not articulate with the navicular (■ Fig. 3.370).

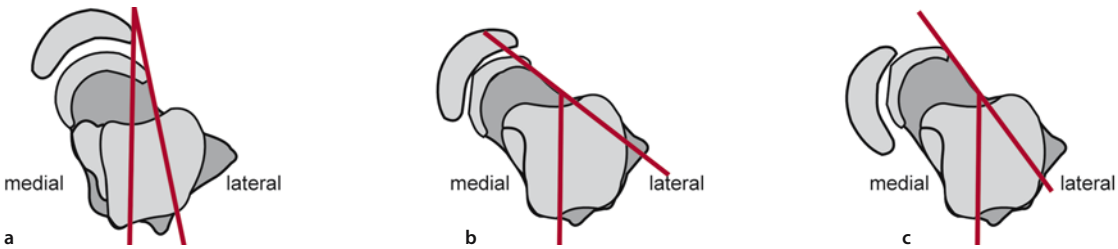
— **Soft tissue changes:**

The soft tissues on the anteromedial and posterolateral sides of the talus are shortened. All tissue types (skin, ligaments, tendons, muscles, blood vessels, nerves) are contracted to the same extent. Histological studies of these tissues have revealed certain changes in clubfoot patients [22], but not in fetuses [19], which suggests that the alterations in the ultrastructure are secondary. The structures most affected by shortening are the posterior fibulocalcaneal and talocalcaneal ligaments, the talonavicular joint capsule, the talocalcaneonavicular ligament, the tendon of the posterior tibial muscle and the fibrous ligament at the intersection of the tendons of the flexor hallucis longus and flexor digitorum longus muscles. Another striking finding is atrophy of the calf muscles.

**Associated anomalies**

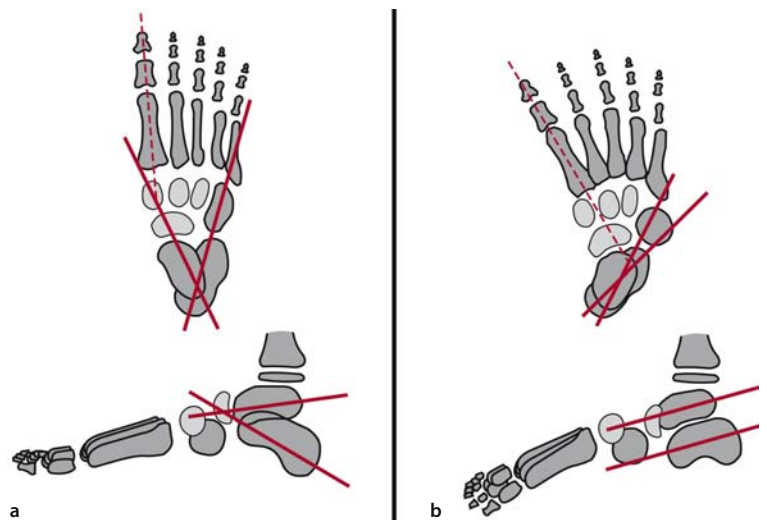
**Isolated clubfoot deformity, associated with other »independent« anomalies**

Associated anomalies are found in approx. 15% of patients with clubfoot. Since congenital hip dysplasia is one such, especially common, associated anomaly, an ultrasound scan of the hips is always indicated in patients with a clubfoot if this investigation has not already been performed in connection with a general screening program.



■ Fig. 3.369a–c. Configuration of the talus in clubfoot. a Configuration in the normal foot. b Medial deviation of the talar neck in clubfoot. c Medial deviation of the talar neck and additional subluxation of the navicular in a medial direction in clubfoot

■ Fig. 3.370a, b. Position of the bones of the foot: a in the normal foot, b in clubfoot (in each case top DP view, bottom lateral view). Lighter area Tarsal bones that are still purely cartilaginous at birth: navicular, cuneiform bones. In the normal foot, the angle formed by the talus and calcaneus on the DP and lateral views ranges from 30–50°, while these two bones are more or less parallel in both planes in clubfoot. Instead of sloping upwardly in a dorsal to ventral direction, the calcaneus is aligned horizontally or even shows a downward slope. The forefoot is adducted, the navicular dislocated medially to a lesser or greater extent (see also ■ Fig. 3.368)



### Clubfoot in connection with hereditary disorders

Clubfoot often occurs in connection with an *arthrogryposis multiplex congenita* (▶ Chapter 4.6.7.1). Since other parts of the body are almost always affected by this condition, with restricted mobility in other joints, the whole locomotor apparatus of the neonate will need to be investigated. Clubfoot is also frequently observed in connection with *Larsen syndrome* (▶ Chapter 4.6.2.21). This condition involves multiple congenital dislocations of various joints (knee, hips) and often also deformities of the spine. Clubfoot is also frequently present in patients with amniotic ring constriction (congenital band) syndrome (▶ Chapter 3.4.5.12 and 3.5.3.8), diastrophic dwarfism (▶ Chapter 4.6.2.6), Freeman-Sheldon syndrome and Möbius syndrome (▶ Chapter 4.6.5.4).

### Secondary clubfoot

Clubfoot can occur as a secondary condition, primarily in neuromuscular disorders (for example in Charcot-Marie-Tooth disease, poliomyelitis or infantile cerebral palsy), and occasionally also in muscular disorders [32].

### Diagnosis

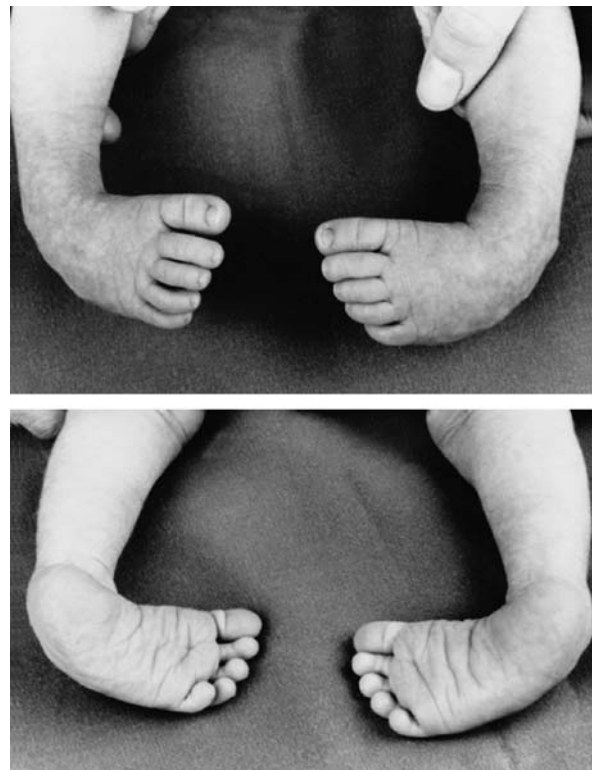
The clinical diagnosis is easily made at birth. The rearfoot is in an equinus and varus position, the forefoot in adduction and eversion (pronation) in relation to the rearfoot (■ Fig. 3.371). The deviation of the forefoot is often incorrectly described as supination, but this only applies in respect of the lower leg and not in relation to the rearfoot. The prominent end of the anterior parts of the talus are palpated on the lateral side. The skin is generally very thin at this point and the skin fold that is normally present is missing, although very fine creasing of this thin section of skin can be produced by everting the foot. The shortened Achilles tendon can be palpated as a tough cord. The heel is small and raised. The lateral malleolus is displaced posteriorly. Calf atrophy is even noticeable at birth in a case of unilateral clubfoot, and its extent is also the best prognostic criterion [6]. The more atrophied the calf muscles are, the greater the expected resistance to treatment. It is very important to classify the foot at birth (■ Fig. 3.366, 3.367), otherwise it will not be possible to assess the subsequent result.

**! The forefoot in clubfoot is pronated, and not supinated, in relation to the rearfoot.**

On the *x-ray*, the ossification centers of the talus and calcaneus – in contrast with a normal foot in which the axes of the two bones form an angle of approx. 40° (20°–50°) on both the AP and lateral views – are largely aligned in parallel with each other (■ Fig. 3.369, 3.372). At birth the ossification centers of the talus, calcaneus, cuboid and metatarsals can be seen on the x-ray, but not the navicular bone, which hampers the evaluation of the extent of

subluxation in the talonavicular joint. The navicular bone only starts to ossify around the 3rd year of life.

It is essential to employ a standardized radiographic technique (▶ Chapter 3.4.2). On an appropriate x-ray it is possible to derive the position of the navicular from the angle between the axis of the talus and that of the first metatarsal (■ Fig. 3.370). We usually record the x-ray not at birth but during the corrective treatment at the age of around 4 months. This is then used to establish the indication and planning for surgery.



■ Fig. 3.371. Clubfeet in the neonate: Top from the ventral side, bottom from the dorsal side. Note especially the pronounced adduction and supination of the feet



■ Fig. 3.372. DP and lateral x-rays of a clubfoot in a 5-month old boy. Talus and calcaneus are largely parallel, the calcaneus slopes downward in a dorsal to ventral direction, the forefoot is also in a downward sloping oblique position



### Differential diagnosis

A distinction must be made between genuine clubfoot and a clubfoot *posture*. In the latter condition the foot is likewise in an adducted varus position, but it is much more flexible and fully reducible, the lateral malleolus is not posteriorly displaced, the skin folds are normal and the fine creasing over the lateral aspect of the talus is absent, the heel is normal in size, there is no, or only a minimal, equinus component, and the calf atrophy is hardly present at all. A distinction must also be made between clubfoot and *congenital pes adductus*. In this (rare) foot disorder, the changes in the rearfoot are absent, but the adduction of the forefoot is already present at birth in contrast with the common condition of acquired pes adductus. The adduction may be accompanied by subluxation in the talonavicular joint.

### Treatment

❗ **In hardly any other area of pediatric orthopaedics has the treatment changed so fundamentally in recent years as in clubfoot, even though Ponseti's basic therapeutic method now in widespread use is over 50 years old. Even just a few years ago, the extensive surgical peritalar reduction by means of a »Cincinnati incision« was the benchmark treatment. It is thanks to the Internet and patient pressure that the conservative Ponseti method with minimal surgical intervention has caught on in recent years. It is much more suitable for preserving a mobile foot than procedures involving comprehensive operations.**

The treatment of a severe clubfoot consists of 4 phases, whereas phase 1 alone applies in a case of clubfoot posture, while phases 1–3 are sufficient for a normal, unproblematic and effectively treated clubfoot.

1. Corrective treatment,
2. Operation,
3. Retention treatment,
4. Correction of recurrences and late deformities.

### Corrective treatment

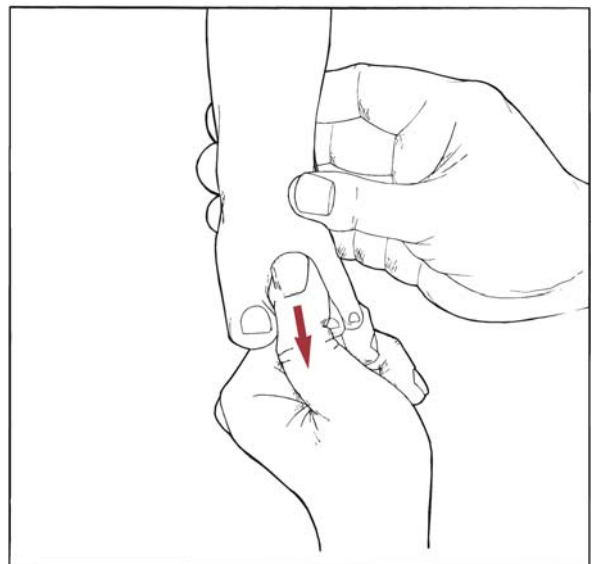
The corrective treatment according to Ponseti involves manipulation of the forefoot deformity. The thumb of one hand stabilizes the talus from the lateral side, while the other hand pulls and supinates the forefoot. This manipulation reduces the navicular from its position of medial subluxation (■ Fig. 3.373). The formerly employed pronating correction of the forefoot should no longer be practiced as the foot will then be corrected in the wrong direction.

❗ **Under no circumstances should an attempt be made to correct the forefoot in the direction of dorsal extension. Since the Achilles tendon is stronger than the**

**metatarsal ligaments and the cartilage of the talus, this can result in the formation of a »rocker-bottom flatfoot« with a high-standing heel, low midfoot and dorsally extended forefoot (■ Fig. 3.374). In addition, the risk of talar necrosis is increased. In clubfoot the soft tissues are hard and the bones soft!**

Massaging of the heel in a caudal direction, on the other hand, is a suitable treatment for the equinus deformity. The corrective treatment should start as soon as possible after birth and is administered by a physical therapist on an outpatient basis. The mother should also be involved in performing the corrective measures under the direction of the physical therapist, who must have received meticulous training in the Ponseti method. After the correction, the position must be held in place by a retaining measure. In our case, we do not use a cast in the very first days of life as this interferes with the close physical contact between the mother and child that is so important at this time. Additionally, not only does the neonate have very thin and fragile skin, but it also grows very quickly at this stage and would thus require daily changing of the cast. We therefore secure the position obtained after correction with casts after 1–2 weeks. We use long-leg casts for this purpose (■ Fig. 3.375), changing them weekly during the first 2 months of life. The foot is growing rapidly at this stage. During the first year, it increases from an average of 7.5 cm to 12 cm, i.e. by 60%.

Below-knee casts have not proved effective as they can easily slip down – particularly if there is a strong equinus component – leading to pressure points. The traditional



■ **Fig. 3.373.** Principle of clubfoot correction according to Ponseti: The thumb of one hand stabilizes the talus from the lateral side and valgizes the calcaneum, while the other hand pulls and supinates the forefoot and thus reduces the navicular bone

### 3.4.3 · Congenital clubfoot

plaster of Paris is by far the best material for modeling purposes, giving it a clear advantage over modern plastics (e.g. Scotchcast). The weight saving offered by the latter compared to normal plaster is of secondary importance in these small infants. Nor have we found Softcast to be particularly effective. Although it is softer than plaster, can be removed by the mother and does not require a cast saw, it is not very good at retaining the foot position and wrinkling of the cast can lead to pressure points.

As an alternative to the combination of physical therapy and cast, physical therapy alone is employed in some centers, allegedly with good results. In France, a brace that exerts traction on the calcaneus has been developed, while a »continuous passive motion« device is even used elsewhere. Our experience to date with these methods has not proved particularly positive.

We re-assess the situation at the age of around 2 months: We clinically evaluate the position of the forefoot, the subluxation of the navicular and the equinus component and repeat the Pirani classification. We also record an x-ray at this time. If the corrective treatment has not managed to restore a completely normal situation (which is usually only possible with the »clubfoot posture«), we consider surgery to be indicated.

The use of botulinum toxin has been advocated as an alternative to surgical Achilles tendon lengthening [1]. The initial reports are encouraging, but it is too early to assess the value of this treatment.

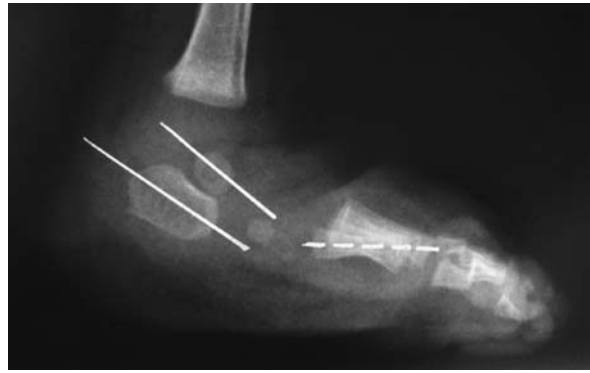
#### Operation

Achilles tendon lengthening is performed at the age of 2–4 months. Achilles tendon lengthening is indicated if the midfoot score is less than 1 in the Pirani classification. In experienced hands, this should be the case in 90% of the feet after 5 or less casts [28]. Ponseti proposed the percutaneous tenotomy [30]. For our part, we prefer percutaneous (or open) cutting of the tendon medially and laterally at two different levels, because of concerns about overcorrection or a pes calcaneus position. At any rate, this is a procedure with minimal morbidity that can be performed on an outpatient basis or with 1-2 days hospitalization. Postoperatively, a cast is fitted for 3 weeks.

Careful evaluation of the foot should include a radiographic assessment. In particular, the correct position of the navicular must be checked.

A peritalar reduction is occasionally required for the rare extremely severe forms of clubfoot (Dimeglio grade IV, Pirani midfoot and hindfoot scores both >2).

This operation for these severe cases involves lengthening of the Achilles tendon, an extensive posterior release (division of the posterior joint capsules of the upper and lower ankle and of the lateral and medial talocalcaneal ligaments). If navicular subluxation is present a medial release is also required, with division of the ligaments between the talus, navicular and medial cuneiform bones,



■ Fig. 3.374. Lateral x-ray of the foot of a 6-month old boy with an incorrectly treated clubfoot that has developed into a *rocker-bottom flatfoot*. As a result of correction of the forefoot in a cranial direction, the Achilles tendon was not lengthened, but the forefoot was pushed upwards. The calcaneus is elevated and slopes downward in a dorsal to ventral direction, whereas the forefoot rises up and the center of the foot is the lowest point, producing a crescent-shaped foot that resembles a »rocker bottom« sole



■ Fig. 3.375. *Clubfoot cast*. For modeling purposes, traditional plaster of Paris is superior to modern plastics. For clubfoot we prefer to use long-leg casts rather than below-knee casts as, firstly, they produce a better outward correction of the foot and, secondly, the below-knee cast (particularly if an equinus deformity is present) readily slips down and produces pressure points. The knee must be flexed by at least 60° in order to ensure that the foot as a whole can be corrected outwardly in relation to the thigh. In addition the forefoot is abducted and pronated in relation to the rearfoot

reduction of the navicular and possibly lengthening of the tendon of the posterior tibial muscle. This operation can be performed either with a dorsolateral incision next to the Achilles tendon and, if necessary, an additional incision on the medial edge of the foot or the so-called Cincinnati incision. [7], a horizontal incision at heel level proceeding, on the lateral side, from the cuboid around the heel medially to the navicular. This incision permits concurrent correction of all contracted components of the clubfoot. At the end of the operation, we use a Kirschner wire to transfix the navicular from the direction of the talus, and the talus and tibia from the direction of the heel and apply a long-leg cast. The cast is changed after 3 weeks and the wire is removed at the same time. The treatment is concluded after 6 weeks.

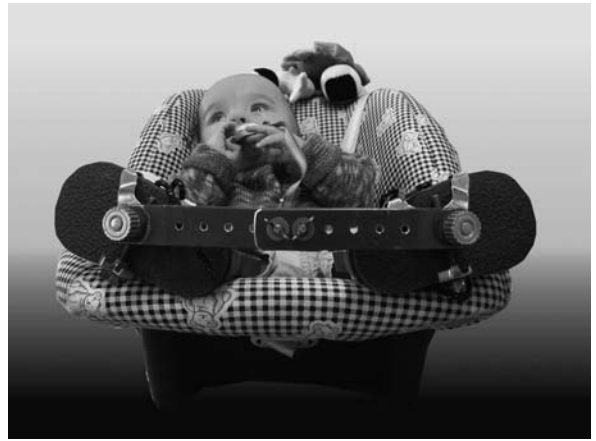
### Retention treatment

The pathological changes that have provoked the clubfoot seem to persist during the first 4 years of life and can cause recurrence. Ponseti therefore advocates prophylactic use of a splint until the age of 4 years. The following conservative measures may be considered:

- Ponseti brace (■ Fig. 3.376),
- long-leg splint (■ Fig. 3.377),
- anti-varus shoe,
- fitting shoes the wrong way round,
- shoes with a plantar joint,
- insert with varus pad (■ Fig. 3.378),
- inner shoe.

The *Ponseti brace* is our standard brace after clubfoot treatment. It allows the child to kick and move his legs while still producing a corrective effect. The brace is worn day and night until the child starts walking, and thereafter only at night. It is important to fit the brace particularly after a conservative (or a minimal surgical) Ponseti treatment. A recently published study showed that poor compliance by parents who disliked the brace resulted in a recurrence in a high proportion of cases [11]. The Ponseti brace must be worn at night until the age of 4. The *long-leg splint* (■ Fig. 3.377) is particularly suitable postoperatively after peritalar reduction. The knee must be flexed in the splint so that the foot can be held in abduction.

Of the corrective options for the walking child, the *anti-varus shoe* and an *insert with varus pad* (■ Fig. 3.378) produce roughly the same (slight) effect. Since these measures do not involve any counterforce (the forefoot is not corrected against another fixed structure, e.g. the heel or thigh), the muscles are still able to exert traction. In principle, the same result can be achieved more cost effectively by *fitting the shoes the wrong way round* (i.e. the left shoe on the right foot and the right shoe on the left foot). Since the lateral edge of the shoe is curved more strongly toward the center of the foot compared to the medial edge, this



■ Fig. 3.376. *Ponseti brace*. In this simple and cost-effective brace, the shoes are fixed to a cross-bar in an outwardly rotated position. The brace allows a certain freedom of movement while still exerting a corrective effect

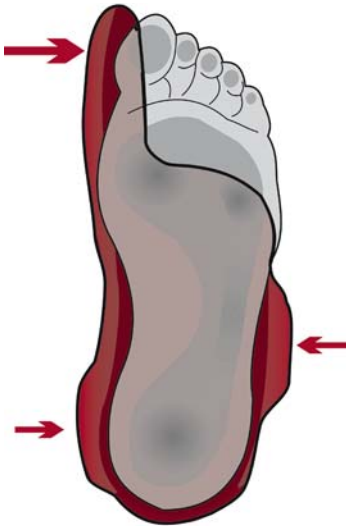


■ Fig. 3.377. *Long-leg splint*: While the splint cannot correct the foot, it does maintain the achieved position by rotating the foot outwardly in relation to the thigh. The splint must be flexed by at least 60° at the knee. The forefoot can also be held in an abducted position in relation to the rearfoot

produces a certain (slight) corrective effect. The *shoe with plantar joint* likewise only produces a moderate effect, and the heel-to-toe roll is hampered by the rigid sole required for the metal joint.

An adequate corrective effect can only be achieved in the walking child with a so-called *inner shoe*. This is a lower leg orthosis in which the heel is firmly grasped so that the forefoot can be corrected in relation to the heel.

## 3.4.3 · Congenital clubfoot



■ Fig. 3.378. Insert with varus pad or »three-tab insert«. This functions according to the three-point principle (arrows). The forefoot (particularly the great toe) is abducted in relation to the rearfoot

This type of orthosis can be worn inside a normal shoe. In our experience this is the most effective measure for children who adduct the forefoot abnormally when they start walking.

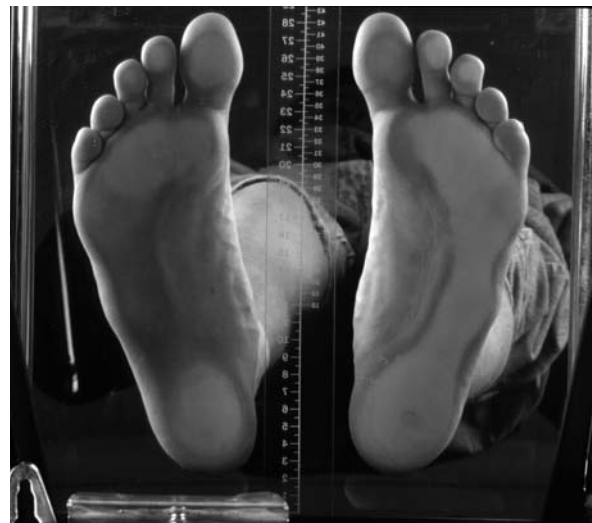
#### Correction of recurrences and late-occurring abnormal positions

A distinction must be made between »recurrences« and the consequences of inadequate treatment, this is only possible if the foot was classified at birth (e.g. according to Dimeglio or Pirani, see above). Reports of short-term or medium-term therapeutic results without any classification of the feet are of no value these days. In addition to recurrences of the equinus deformity and the adduction of the forefoot, secondary hallux varus in particular should be mentioned at this point. This varus position of the great toe is the compensatory response to supination of the forefoot and the lack of weight-bearing under the first metatarsal head (■ Fig. 3.379).

The following measures have been proposed for recurrences or late-occurring abnormal positions:

- Soft tissue corrections,
- Tendon lengthening procedures,
- Tendon transfer procedures,
- Osteotomies,
- Correction with the external fixator (ring fixator, e.g. Ilizarov-type apparatus).

Soft tissue corrections (e.g. repeated medial release) are not very successful since the scars that are constantly



■ Fig. 3.379. 15-year old boy with secondary hallux varus with supination of the forefoot and absence of weight-bearing under the head of the 1st metatarsal after congenital clubfoot

being reformed can contract and lead to a further recurrence. We no longer perform these operations and instead correct the foot with osteotomies or a ring fixator (see below). Nor has the repeated lengthening of the Achilles tendon proved effective, primarily because, in the event of a recurrence of the equinus deformity, the posterior joint capsules of the ankle and subtalar joints are also contracted. Here, too, the use of a ring fixator can produce much better results. Of the tendon transfer procedures, the transfer of the tendon of the anterior tibial muscle (or part of this tendon) laterally is recommended in particular for correcting the adduction and supination of the forefoot. We used to perform this operation more often in the past, but have since noted the not infrequent occurrence of flatfoot in our long-term follow-up after 20 years [29]. The operation is therefore useful only if there is a pronounced pes cavus component and must then be implemented very gradually (a part of the tendon must be left in place). For the above reasons, only osteotomies



and corrections with the ring fixator will be addressed in greater detail at this point.

A special and underestimated problem is the overlength of the Achilles tendon. This results in a decreased force for plantarflexion, and equinus gait is almost impossible. In such cases shortening of the Achilles tendon is sometimes indicated. In severe cases the foot extensors have to be lengthened.

### Osteotomies

Osteotomies are performed on the following bones:

- Tibia,
- Calcaneus,
- Talus,
- Cuboid and medial cuneiform bones,
- Metatarsals.

#### Tibia: lower leg derotation osteotomy

Since the forefoot is often internally rotated in relation to the upper leg, an externally rotating osteotomy of the tibia is frequently indicated. The fact that the abnormal position is in the foot rather than the lower leg is usually disregarded. Although the tibia is often slightly rotated inwardly in clubfoot [25], the fibula is posteriorly displaced. Derotation of the tibia will intensify this effect and the abnormal position will not be corrected at its actual location, i.e. the midfoot. We therefore consider that a tibial derotation osteotomy is rarely indicated in clubfoot. The surgical technique is described in ► chapter 4.2.1.

#### Calcaneal osteotomies

Dwyer proposed (originally for the treatment of pes cavus) the removal of a wedge from the calcaneus (■ Fig. 3.380a). Later he recommended an operation at the same site, but with the insertion of a wedge, for clubfoot [12]. This operation is implemented if the heel is in an abnormal varus position. It is only indicated in rare cases, however, since the soft tissues over the heel are very tight on the medial side, hampering the insertion of a bone wedge and risking problems with skin closure. On the other hand, the calcaneus is usually too short in clubfoot, and the removal of a wedge would also prove problematic since it would make the calcaneus even shorter. In patients with a very short calcaneus in a varus position we therefore no longer remove a wedge, but perform a lateral displacement of the dorsal section at the same site as the osteotomy, as suggested by Mitchell [27] (■ Fig. 3.380b). To date we have an accumulated experience of over 100 calcaneal osteotomies [17].

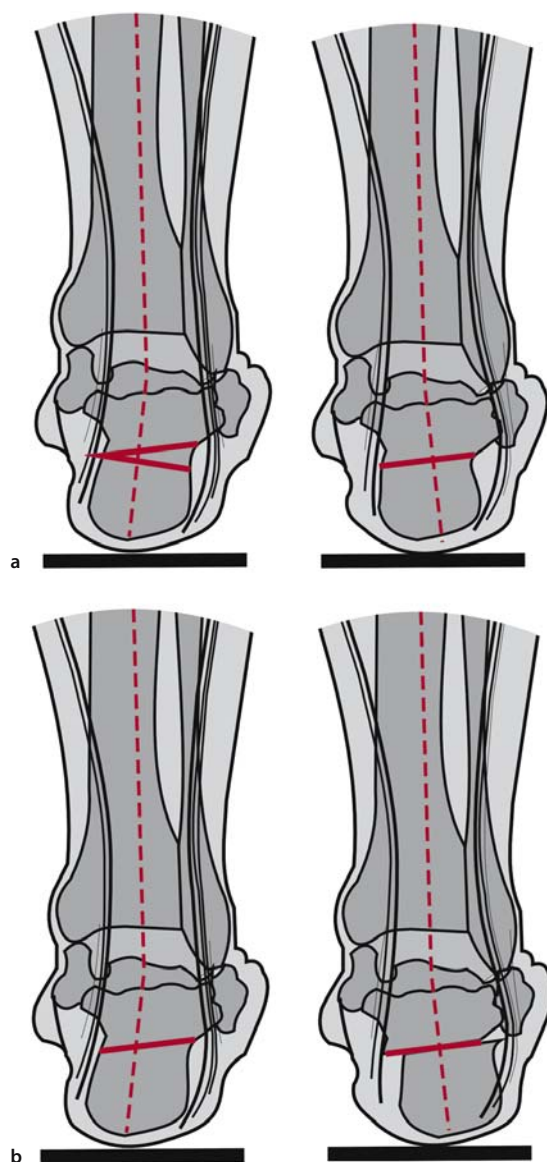
#### Talar osteotomy

Descriptions of osteotomies in the area of the talar neck are repeatedly being published, although we have no ex-

perience with such operations. The persuasive underlying argument is that the operation is performed at the site of the deformity. However, the very poor circulation in the talus raises perfectly legitimate fears about the complication of talar necrosis.

#### Metatarsus: Closing cuboid and opening wedge cuneiform osteotomy

The closing cuboid and opening wedge medial cuneiform osteotomy corrects the deformity at the base of the forefoot, i.e. very close to the actual site of the abnormal position. The bone wedge removed from the cuboid is



■ Fig. 3.380a, b. Principle of calcaneal osteotomy according to Dwyer. a Conventional osteotomy with removal of a lateral wedge and valgization of the calcaneus. b Modified technique (according to Mitchell) without wedge removal, with lateral displacement of the posterior fragment



### 3.4.3 · Congenital clubfoot

inserted into the osteotomy gap in the medial cuneiform (■ Fig. 3.381).

Each osteotomy is transfixed with a Kirschner wire, and a below-knee cast is fitted for 4 weeks. If the forefoot is so rigid that it prevents insertion of the laterally removed wedge on the medial side, we temporarily distract the medial osteotomy gap at operation with a small external fixator. This operation is very efficient and has a high success rate. We have performed the procedure in our hospital for the past 12 years. A follow-up study involving 30 patients found that permanent correction was achieved in 90% of cases [33]. The operation is suitable for patients with pronounced adduction of the forefoot at the age of 6 years or older. If a strong supination component is present, we join the medial and lateral osteotomies to produce a full-width osteotomy and pronate the whole forefoot. This is indicated particularly in cases of pronounced secondary hallux varus.

#### Metatarsals

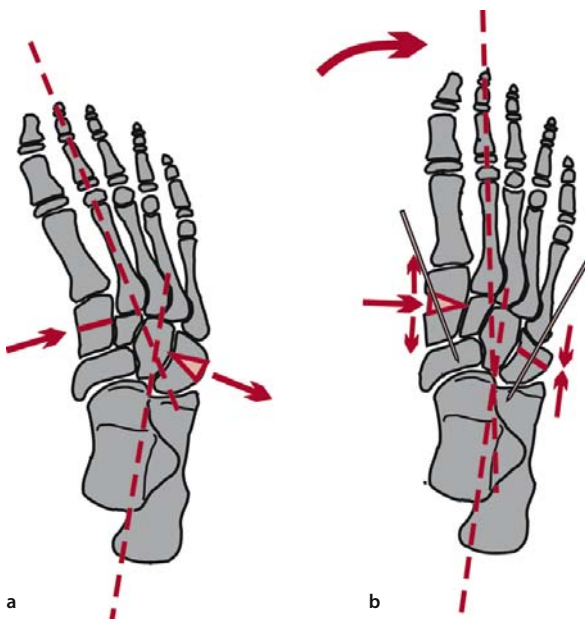
The correction of forefoot adduction at the base of the metatarsals is practiced in many centers. This procedure corrects the deformity, but less efficiently than the

cuboid and cuneiform osteotomy since it is more distal and further away from the pivot point. Because of the epiphyseal plates it can only be performed after the child has stopped growing. In our view, it is only indicated for a varus position of the first metatarsal alone. This operation is described in ► chapter 3.4.8 (juvenile hallux valgus).

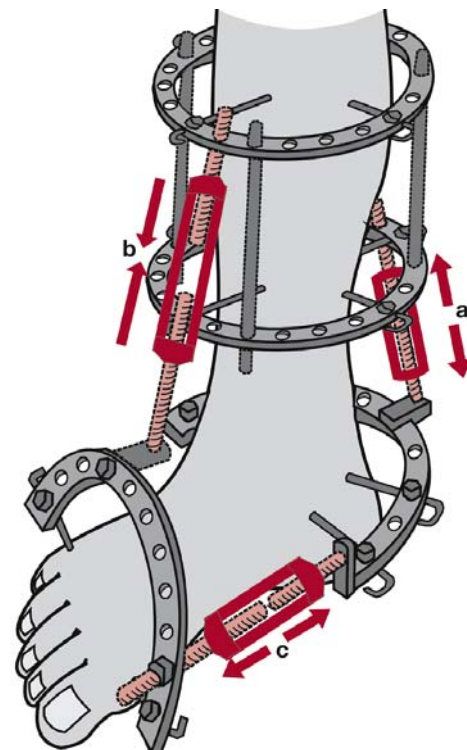
#### Correction with the external ring fixator

The ring fixator was developed in the 1950's by Ilizarov in Russia for the treatment of fractures and for limb lengthening [21]. However, the western world only became aware of this system towards the end of the 1970's. It soon became apparent that the ring fixator was suitable not only for stabilizing or lengthening bones, but also for lengthening soft tissues and thus correcting deformities. The system of half rings is very versatile and can be used for an almost limitless range of applications. We began wyeears ago and have treated around 50 cases to date [3]. Other authors have likewise reported on its use for this application [15]. The therapeutic principle is illustrated in ■ Fig. 3.382.

The simplest technical procedure is correction of the



■ Fig. 3.381a, b. Principle of the *closing cuboid and opening wedge cuneiform osteotomy*. (a) The bone wedge removed from the cuboid is (b) inserted into the medial cuneiform. If the foot is very rigid, the cuneiform bone may need to be opened up with the aid of 2 Kirschner wires or even an external fixator. This is the most effective operation for correcting adduction of the forefoot since the correction takes place very close to the site of the deformity. The operation can be performed from the age of 6 at the earliest (depending on the bone maturation in each case)



■ Fig. 3.382. *Correction of clubfoot deformity with the Ilizarov apparatus*: Two rings are attached to the lower leg. Half rings are fixed to the calcaneus and forefoot. A distraction element (a) corrects the elevated heel while, at the same time, a compression element (b) pulls the forefoot upwards. An additional distraction rod on the medial side of the foot can correct the adduction (c)

equinus component. The rearfoot is held in place by a half ring fixed with 2 Kirschner wires, another half ring is fitted to the dorsal forefoot and two rings are attached to the lower leg. The deformity is corrected by applying distraction dorsally and compression ventrally (■ Fig. 3.382). As with the standard bone lengthening procedure, the threaded rod is lengthened or shortened by 1 mm a day. The threaded rod anchored on the lateral side of the anterior half ring pulls the foot into pronation. A distraction rod can be inserted medially between the half rings on the fore- and rearfoot to correct the adduction of the forefoot. However, since this rod produces a translational movement, whereas a rotational movement is really required to correct the adduction, we use another method for severe deformities: Two rings are fitted to the distal lower leg, one inside the other to produce inner and outer rings. A threaded rod is used to rotate the outer ring in relation to the inner. The outer ring is linked to the half ring on the forefoot and the inner ring to the half ring on the rearfoot. Rotation of the outer ring corrects the forefoot adduction with a rotational movement (■ Fig. 3.383).

The ring fixator is very efficient at correcting foot deformities (■ Fig. 3.383), and even very contracted abnormal positions can be normalized, albeit with a lot of patience. The procedure is also often painful and associated with a high level of complications [15], particularly repeated infections at the entry points of the wires. Oc-

asionally the wires also pull out or contracture of the toe flexors occur. The treatment usually lasts 2–3 months and the patient is rewarded with a very efficient and usually permanent correction. We have observed recurrences particularly in foot deformities connected with arthrogryposis. In severe contracture deformities it can therefore prove worthwhile to combine the soft tissue correction with the ring fixator with an osteotomy at the most contracted point. We usually perform an osteotomy on the first metatarsal or the medial cuneiform bone. The contracture of the toe flexors can be corrected with the help of physical therapy. After removal of the ring fixator, the position must be consolidated for a prolonged period with a (plastic) walking cast.

We have recently started using the Taylor Spatial Frame to carry out these corrections. This apparatus allows an even more precise correction with better control of the pivots.

#### **Our therapeutic strategy for clubfoot**

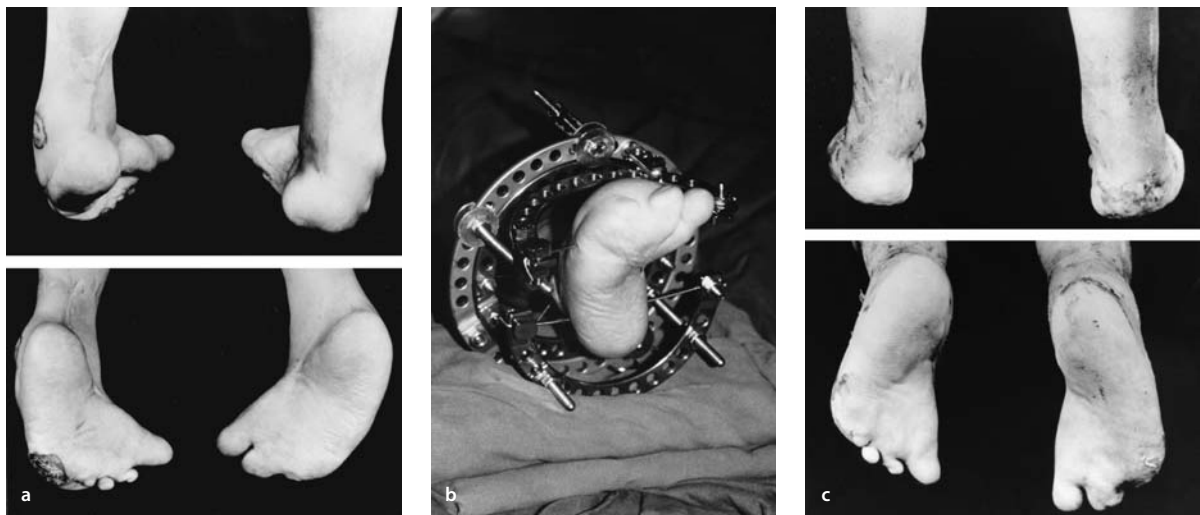
Our therapeutic strategy for clubfoot is shown in ■ Table 3.66.

» *The limping Hephaestus of Greek myth probably suffered from clubfoot, yet he became the husband of Aphrodite, the goddess of beauty and love.* «

■ Table 3.66. Therapeutic strategy for clubfoot

	Therapeutic measures
<b>Primary treatment</b>	
First 3 weeks of life	Manual correction according to Ponseti, corrective dressings
4th week to approx. 2nd (to 4th) month	Manual correction according to Ponseti, retention with long-leg casts (change weekly and, from the 2nd month, every 2 weeks)
2nd to 4th month	When Pirani midfoot score is less than 1, but calcaneus is too high, then Achilles tendon lengthening; 2 months postoperatively long-leg casts For Dimegl io grade IV or Pirani midfoot and hindfoot scores both >2 at birth, surgical peritalar reduction
up to 4 years of age	Ponseti brace at night Treatment usually concluded by this time
<b>Secondary treatment (for refractory clubfoot or recurrences)</b>	
Adduction (and supination) of the forefoot	Combined closing cuboid and opening wedge medial cuneiform osteotomy (from the age of 6/7)
Varus of the calcaneus (hindfoot)	Lateral displacement osteotomy of the calcaneus according to Mitchell
Equinus foot (and adduction and supination of the forefoot, possibly also leg shortening)	Correction with the Ilizarov apparatus
Calcaneus foot (because of overlength of the Achilles tendon)	Shortening of the Achilles tendon, possibly also lengthening of the foot extensors
Internal rotation of the lower leg (extremely rare!)	Supramalleolar tibial derotation osteotomy

## 3.4.3 · Congenital clubfoot



■ **Fig. 3.383a–c.** Correction of *adduction and supination of the foot with the Ilizarov apparatus*: An Ilizarov apparatus was fitted to an 8-year old girl with a severe clubfoot deformity (with a ring constriction syndrome (a)). Since the deformity was so pronounced, the translation

principle shown in Fig. 3.382 was not used. Rather, a double-ring that produces derotation of the forefoot was fitted (b). After 3 months, a plantigrade foot position is achieved with substantial correction of the deformity (c)

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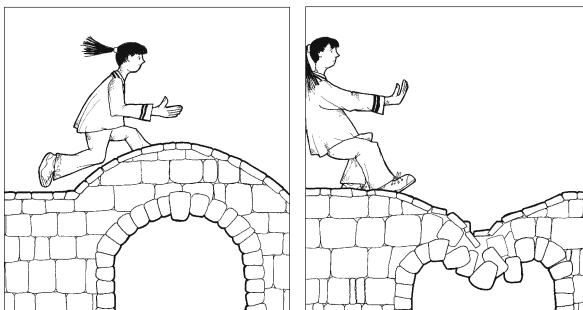
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### 3.4.4 Congenital flatfoot (vertical talus)

#### ► Definition

Rare congenital deformity of the foot with vertical orientation of the talus and dislocation of the talocalcaneonavicular joint dorsally and laterally (the acquired forms of flatfoot are discussed in ► chapter 3.4.7).

— **Synonyms:** Congenital vertical talus, congenital rigid flatfoot, congenital convex pes valgus, congenital rocker-bottom flatfoot, platypodia



#### Historical background

In contrast with clubfoot, which was known as a clinical diagnosis back in ancient times, the presence of congenital flatfoot was only discovered after the invention of the x-ray. The condition was first described by Henken 1914 [9].

#### Occurrence

While we are not aware of any epidemiological studies, flatfoot can be described as a fairly rare deformity. Most

authors that have reported on the treatment of flatfeet have involved populations of 5–35 patients [4, 5, 11]. At our hospital we treat around 1 case of vertical talus a year (compared to approx. 15–20 cases of clubfeet). Both sexes are affected with equal frequency, and other anomalies exist concurrently in roughly fifty percent of patients.

#### Associated anomalies

Congenital vertical talus occurs alone or in connection with neurological disorders (particularly myelomeningocele) or other systemic illnesses. Only approx. 50% of cases occur in isolation, while additional anomalies are found in the remainder. Congenital vertical talus is observed in approx. 10% of patients with myelomeningocele and is already present at birth. It should not be confused with secondary neuromuscular pes planovalgus, which is very common in myelomeningocele and a consequence of the missing muscle function (► Chapter 3.4.10 and ► chapter 4.7.4). Congenital flatfoot has also been observed in connection with arthrogryposis, neurofibromatosis, trisomy 18 [14], Prader-Willi syndrome, De Bary syndrome [12] and prune-belly syndrome [6].

#### Classification

Hamanishi proposed the following classification on the basis of observations made in 69 cases of congenital vertical talus [7]:

- Type 1: vertical talus associated with spinal anomalies,
- Type 2: vertical talus associated with neuromuscular disorders,
- Type 3: vertical talus associated with malformation syndromes,
- Type 4: vertical talus associated with chromosomal anomalies,
- Type 5: idiopathic vertical talus
  - 5a: resulting from an intrauterine disorder,
  - 5b: with digitotalar dysmorphism,
  - 5c: with vertical talus in a close relative,
  - 5d: not associated with any other skeletal anomaly or genetic component.

#### Etiology

The frequency of a very wide variety of associated anomalies underlines the fact that vertical talus is a very heterogeneous condition in etiological respects. Vertical talus in isolation appears to be the result of a problem during pregnancy. Up until the 7th week of pregnancy the foot is in pronounced dorsal extension and gradually plantarflexes over the course of the following weeks. The damage must occur during this phase, possibly as a result of the concurrent shortening of both the triceps surae muscle and the foot extensors. A hereditary component has been observed both for flatfoot in isolation and in association with other anomalies [13].



### 3.4.4 · Congenital flatfoot (vertical talus)

#### Pathological anatomy

The pathoanatomical changes have been investigated in several children with multiple deformities who died at an early age [3]. The principal element is the dislocation of the navicular bone in a cranial direction. It no longer articulates with the anterior joint surface of the talus, but is located dorsal to the talar neck (■ Fig. 3.384). The talus is tilted downward on the medial side of the calcaneus and stands vertically. At the same time, the calcaneus is rotated posterolaterally. The sustentaculum tali is hypoplastic, allowing the talus to slip past it. All ligaments and tendons on the medial aspect of the rearfoot are lengthened substantially and form a dislocation pouch in which the talus resides. The triceps surae muscle and the foot extensors are shortened and contracted.

#### Diagnosis

The diagnosis of congenital flatfoot can usually be confirmed at birth just on the basis of *clinical examination*. The sole of the infant's foot is convex and palpation reveals the prominent talus instead of the medial arch. The forefoot is abducted and dorsally extended. The heel stands high and the calf muscles are shortened. An equinus deformity is present, though this may be masked by dorsal extension of the forefoot. Occasionally, the cranially dislocated navicular bone can also be palpated. In a case of genuine vertical talus the foot is contracted and cannot be manipulated into the normal position. The lateral x-ray shows an almost vertically standing talus the head of which may also appear lower than the calcaneus. The latter is horizontal and lacks the normal upward slope from a dorsal-caudal to ventral-cranial direction. Sometimes the calcaneus even tilts in a dorsal-cranial to ventral-caudal direction.

The angle between the calcaneus and talus is increased, usually to around 90°. This angle may also be reduced, however, if the calcaneus tilts downward. An abnormally high talocalcaneal angle is also usually mea-

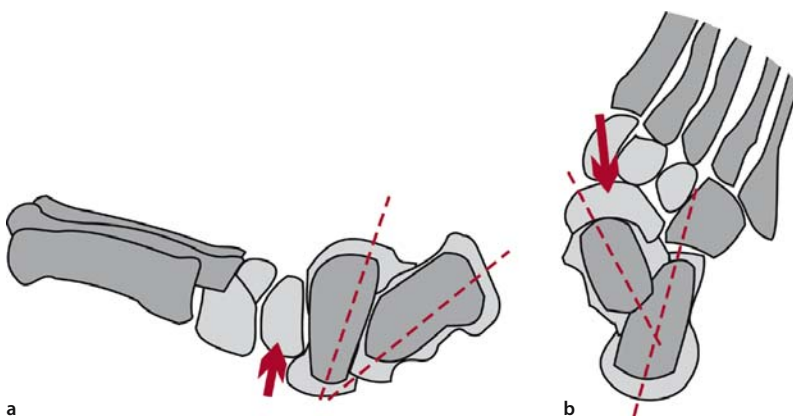
sured on the DP view (■ Fig. 3.385). A useful method for differentiating between a vertical talus and a flexible flatfoot or oblique talus is to record lateral x-rays of the foot firstly in a plantigrade position and then in maximum plantar flexion. In a patient with flexible flatfoot, plantar flexion reduces the abnormal configuration of the talus and navicular, causing the 1st metatarsal to form a continuation of the talar axis. This is not the case with vertical talus (■ Fig. 3.386). No other imaging procedures (MRI, CT) are required to confirm the diagnosis, although ultrasound may be useful for visualizing the dislocation of the navicular.

#### Differential diagnosis

Differentiating between vertical talus and *flexible flatfoot* (► Chapter 3.4.7) is not always easy as the latter can likewise be present even at birth. However, the foot is not nearly as contracted as in congenital flatfoot, the navicular is not dislocated and the triceps surae muscle is not yet shortened at the time of birth. Flexible flatfoot is usually conspicuous only after the child starts walking, if the medial arch has not formed by this time (usually as a result of extreme ligament laxity).

#### Treatment

Vertical talus is a serious abnormality of the foot. All authors of recent studies now agree that purely conservative treatment cannot produce a successful outcome [4, 5, 10, 11]. Disagreement exists, however, as to whether surgical correction should be performed as a single-stage procedure [5, 14] or in two steps [4]. In our hospital we try as far as possible to correct the whole deformity in a single step and at the earliest possible point [15]. The operation should preferably be performed during the first 3 years of life. The procedure involves a posterior capsulotomy of the upper and lower ankle, Achilles tendon lengthening (this part of the operation is similar to the procedure for clubfoot) and open reduction of the navicular, closure of



■ Fig. 3.384a, b. Bones of the foot in vertical talus. **a** In the lateral view the talus stands almost vertically and the calcaneus slopes downward, rather than upward, in a posterior to anterior direction. The still cartilaginous navicular in the neonate is subluxated (or dislocated) in a cranial direction. **b** Seen from above, the angle between the talus and calcaneus is increased.





■ Fig. 3.385a, b. Bilateral *vertical talus* in a boy. a X-rays at the age of 4 months. The vertical talus is apparent on the lateral views, but the axis of the dysplastic calcaneus cannot be assessed. Pronounced medial deviation of the talus is seen on the AP views. b Lateral views of the

foot 1 year after open reduction of the navicular and talus, Achilles tendon lengthening and closure of the dislocation pouch. The foot now shows a substantially normal configuration

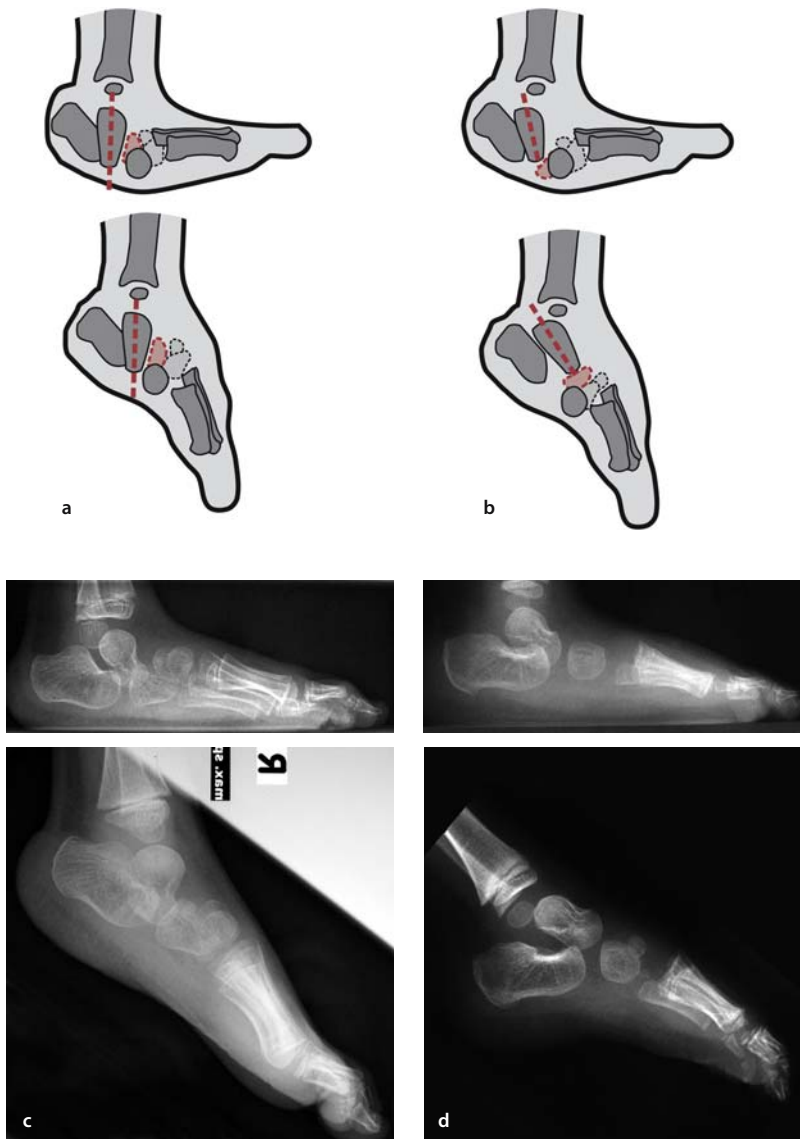
the medial dislocation pouch and transfixation of the talonavicular joint. Postoperatively, a below-knee cast with good modeling of the medial arch is fitted.

❗ **The follow-up management is particularly important after this operation. Since the talus has a strong tendency to slip back to its old position, countermeasures must be continued for several years until the situation has consolidated sufficiently to rule out the risk of recurrence.**

We therefore fit lower leg orthoses providing good medial support for at least 2 years after the operation. During this period we do not allow the child to take a single step without the orthosis in order to prevent renewed overstretching of the soft tissues. Compliance is best when below-knee casts are used, although these must be changed frequently so that the foot can be manipulated from time to time to prevent tendon adhesions. Since implementing this follow-up treatment consistently we

have only encountered a single recurrence (in this case the compliance of the parents was less than ideal). Some authors support the reduction with an extra-articular talocalcaneal arthrodesis according to Grice [11]. We do not consider this measure to be necessary initially. If necessary, it can be implemented at a later date if recurrence occurs (this operation is described in ► chapter 3.4.7). The opening wedge osteotomy of the calcaneus according to Evans [5, 8] in vertical talus is a suitable operation for improving the mechanics of the foot if the outcome of the initial treatment is unsatisfactory. This procedure is described in greater detail in ► chapter 3.4.7. Some authors recommend transfer of the anterior tibial tendon to the talar neck at the same time as the reduction, while others suggest transfer of the peroneus longus tendon dorsally to the talar neck [10]. We have not performed either of these operations ourselves. They are both associated with the basic problem of tendon transfers, i.e. that it is extremely difficult to restore the impaired muscle equilibrium to its correct state.

## 3.4.4 · Congenital flatfoot (vertical talus)



**Fig. 3.386a–d.** Vertical talus and flexible flatfoot or oblique talus. **a top:** Vertical talus. Schematic view of an x-ray in plantigrade position; **bottom:** same foot in maximum plantar flexion. The navicular is not reduced and the talus and 1st metatarsal are not aligned (radiological example in c). **b top:** Flexible flatfoot or oblique talus. Schematic view of an x-ray in plantigrade position; **bottom:** foot in maximum plantar flexion. The navicular is reduced and the talus and 1st metatarsal are parallel and aligned (radiological example in d)

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### 3.4.5 Other congenital anomalies of the foot

#### 3.4.5.1 Accessory ossification centers, talus partitus

##### ➤ Definition

Ossification anomalies of the foot skeleton with the occurrence of surplus (accessory) bones, usually at the tendon attachments. These are normal variants.

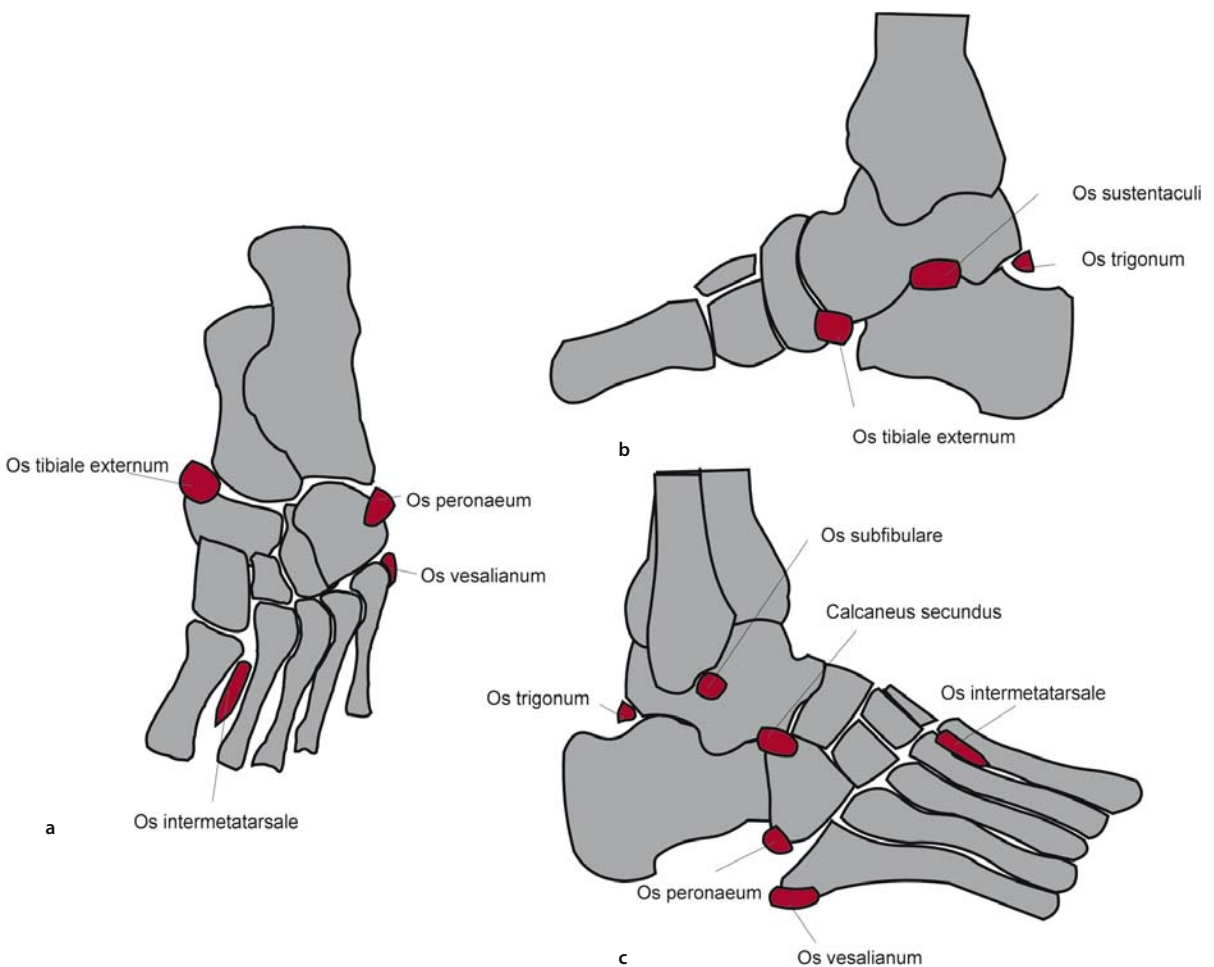
##### Nomenclature, occurrence

■ Fig. 3.387 shows the sites, names and frequencies of the various accessory tarsal bones [49]. Accessory ossification centers are common, with approx. 15% of the population possessing such variations. The commonest are the os trigonum, the os peronaeum and the os tibiale externum (accessory navicular). The only accessory

bones that are of clinical significance are the accessory navicular bone and the subfibular bone (os subfibulare). Rather than a spontaneously occurring accessory ossification center, the os subfibulare usually forms after a traumatic cartilaginous rupture of the anterior talofibular ligament and subsequent ossification of the tendon attachment.

##### Clinical features, diagnosis

Accessory ossification centers of the foot are usually unearthed as chance findings on conventional AP and lateral x-rays of the foot. Since they are not usually clinically significant, no further diagnosis is required, although it is important to be aware of them so that the innocuous nature of the finding is correctly assessed. Possible exceptions are the os accessory navicular bone (■ Fig. 3.388) and the os subfibulare. The os accessory navicular bone may, particularly in connection with a flexible flatfoot,



■ Fig. 3.387a–c. Schematic presentation of the commonest *accessory ossification centers* (mod. after [49]): a from top, b from medial side, c from lateral site. The commonest accessory bone is the accessory navicular bone (os tibiale externum). The os subfibulare corresponds

to a ruptured cartilaginous attachment of the anterior talofibular ligament. The os vesalianum [49] was described by A. Vesalius in 1533, but has not been mentioned by subsequent authors. It is usually confused with the separate apophyseal center that is often present at this site

### 3.4.5 · Other congenital anomalies of the foot

give rise to symptoms. As the bone often protrudes significantly on the medial side it can rub against hard shoes, leading to inflammation and swelling. The resulting pain will then depend on the respective footwear worn by the patient. Occasionally these symptoms also occur at this site even when no accessory bone is present. Instead, the navicular bone is very prominent on the medial side, in which case it is described as a »cornuate navicular bone«.

A projecting bone in the area of the navicular can be classified as one of 3 types [42]: In type I, an ossification center exists in the tendon of the posterior tibial muscle. In type II, the os tibiale externum forms a synchondrosis with the navicular while, in type III, no separate ossification center is present, but rather the aforementioned cornuate navicular bone. Type II may develop over time into a type III situation. The os subfibulare can also occasionally cause pain and is located at the distal end of the fibula, slightly in front of the lateral malleolus. As already mentioned, this is usually a traumatically avulsed ossification center. It can cause symptoms particularly in connection with loosened lateral ligaments and chronic instability. Usually, however, local tenderness is also present (■ Fig. 3.389). In very rare cases, the os trigonum can cause symptoms, generally after trauma to what is actually a very common accessory ossification center.

Besides accessory ossification centers, *congenital cleft formations* can also occur, although these are extremely rare. Isolated cases involving the talus and the calcaneus



■ Fig. 3.388. X-ray of the left foot in a 12-year old girl with an accessory navicular bone (*os tibiale externum*)




■ Fig. 3.389. AP and lateral x-rays of the left upper ankle in a 15-year old boy. The *os subfibulare* is not an accessory ossification center, but a traumatic rupture of the cartilaginous attachment of the anterior talofibular ligament that subsequently ossifies



■ Fig. 3.390a, b. *Talus partitus* in a 14-year old girl. a Lateral x-ray, b MRI scan with frontal and sagittal sections



have been observed.  Fig. 3.390 shows a talus partitus that was giving rise to symptoms. Such symptoms may be attributable to a loosening of the connection between the two parts of the bone.

### Treatment

Since accessory ossification centers rarely produce symptoms, treatment is required in only a small proportion of cases, most commonly in relation to an accessory navicular bone. If this is removed, the surgeon must be careful to preserve, or restore, the stability of the ligamentous apparatus. If pain is experienced medially over the navicular, treatment with an insert does not usually bring any major improvement. In such cases, relief is provided only by surgical removal of the os tibiale externum or chiseling off of the cornuate navicular. The patient himself must decide whether surgery is indicated. When the accessory bone is removed, the attachment of the tendon of the posterior tibial muscle is preserved, thereby producing complete freedom of movement in almost every case [4, 29]. A transfer of the posterior tibial tendon, as recommended by some authors [29], is not necessary, nor does it produce any further improvement in the outcome. Occasionally, an os trigonum will also need to be removed because of symptoms.

### 3.4.5.2 Tarsal coalition

**» The boy's heel elicits a painful perception, indicating the presence of a bony connection. «**

#### ➤ Definition

Bony or connective tissue bridge between two bones of the rearfoot and/or metatarsus.

#### Historical background

Cruveilhier [13] was the first to describe a calcaneonavicular coalition back in 1829.

#### Occurrence


In one epidemiological study with 2,000 recruits, tarsal coalitions were observed in 21 cases [48], which corresponds to an incidence of approx. 1%. So it is a relatively common anomaly, and one that is usually associated with clinical consequences as well.

#### Classification

Tarsal coalition can be subdivided as follows:

1. Isolated coalition:
  - Calcaneonavicular coalition (53% [45])
  - Talocalcaneal coalition (37% [45])
  - Talonavicular coalition
  - Calcaneocuboid coalition
  - Naviculocuneiform coalition

2. Multiple forms, part of a complex syndrome:
  - Apert syndrome (▶ Chapter 3.4.5.12, 4.6.3.1)
  - Carpal coalition (▶ Chapter 3.5.3)
  - Symphalangism (▶ Chapter 3.4.5.4)
  - Longitudinal deformities (▶ Chapter 3.2.7.2, 3.3.6.1, 3.4.5.11)
  - Congenital ball-and-socket ankle joint (▶ Chapter 3.4.5.6)
  - Fibular deficiency (▶ Chapter 3.3.6.1)
  - Proximal focal femoral deficiency (▶ Chapter 3.2.7.2)


Tarsal coalition can also be classified according to the *type of connection* [26] ( Table 3.67). This classification is purely descriptive and based on observations made during operations. However, it is probably a dynamic process in which types II and III can develop into a type I.

#### Etiology

While the etiology is not fully understood, tarsal coalition appears to involve a disorder of differentiation and segmentation of the primitive mesenchyme resulting in the failure to form a proper joint. A coalition between tarsal bones has even been seen in fetuses [25]. In an investigation of 142 fetal cadavers a talocalcaneal bridge was found in a total of 16 cases (9%). The defective differentiation appears to occur in the 9th–10th week of pregnancy. An increased incidence of tarsal coalition has also been observed in connection with club feet [44], fibular deficiency or proximal femoral deficiency [17]. In 26 children with a fibular deficiency requiring a foot amputation, a talocalcaneal coalition was present in 14 cases (54%) [17], although this was radiographically visible only in 4 children. Individual authors also report on a familial occurrence of tarsal coalition [28].

#### Clinical features, diagnosis

Not all patients with tarsal coalitions are symptomatic. In early childhood, in particular, pain is usually absent. But even adults with deficient mobility in an ankle can remain symptom-free, in which case the coalition may only be discovered as a chance diagnosis [28]. The first occurrence of symptoms is characteristic of the site of the coalition and connected with the time of ossification. Talonavicular coalitions can become symptomatic in children as young as 2 years, and the onset of symptoms

 **Table 3.67. Classification of tarsal coalition according to the type of connection**

Type	Type of connection
I	Bony
II	Cartilaginous
III	Fibrous



### 3.4.5 · Other congenital anomalies of the foot

in calcaneonavicular coalitions is generally between the ages of 8 and 12, while talocalcaneal coalitions do not cause symptoms until adolescence [12].

The nature of the symptoms also depends on the site of the coalition. Talonavicular coalition usually involves a prominence on the medial side of the foot without the presence of major symptoms. In particular, the longitudinal arch of the foot is usually preserved. With calcaneonavicular and talocalcaneal coalition, on the other hand, mobility of the lower ankle is impaired. The foot deviates increasingly into a rigid abnormal valgus position result-

ing, in turn, in spasticity of the peroneal muscles. The extent of the valgus deformity of the calcaneus can vary considerably: It can be severe enough to completely flatten the medial arch, resulting in a rigid flatfoot.

**! The possibility of tarsal coalition should always be considered in a case of rigid pes planovalgus or unclear pain in the mid/rearfoot. Such bridge formations are one of the most commonly overlooked diagnoses in pediatric orthopaedics.**

Such rigid flatfoots are often painful, particularly in the midfoot area. The pain is load-related and can also occur in the area of contracted peroneal muscles. The opposite deformity, namely a varus position of the rearfoot with a pes cavus component, has been observed in isolated cases of tarsal coalition [46]. A congenital ball-and-socket ankle joint can also develop as a consequence of tarsal coalition and the lack of mobility in the lower ankle (► Chapter 3.4.5.6).

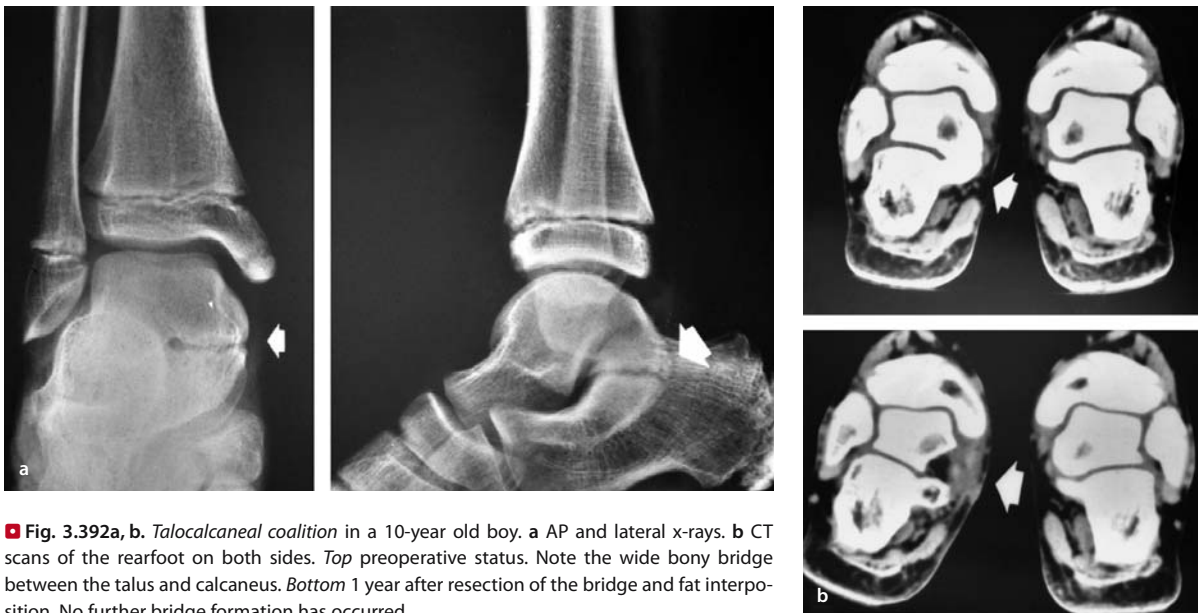
The coalition is not always readily visible on conventional AP and lateral *x-rays*. Since the subtalar joint runs at an oblique angle to the horizontal, the joint is not clearly visualized on the lateral view. In particular, the coalition cannot be seen on a conventional *x-ray* when the connection is fibrous or purely cartilaginous. A much clearer picture is obtained with an oblique *x-ray* of the rearfoot, in which the beam is inclined at 45° to the horizontal. Views in various oblique positions are sometimes needed to visualize a bone connection clearly since the overlaying of bone structures can be mistaken for a bridge formation (■ Fig. 3.391).

For this reason, *computed tomograms* are also helpful and indicated if a coalition is seriously suspected.

■ Fig. 3.392 shows an *x-ray* of a patient with a talocal-



■ Fig. 3.391a, b. *Calcaneonavicular coalition* in a 13-year old boy. a The coalition is much more difficult to see on the lateral *x-ray* than on the oblique view (b)



■ Fig. 3.392a, b. *Talocalcaneal coalition* in a 10-year old boy. a AP and lateral *x-rays*. b CT scans of the rearfoot on both sides. *Top* preoperative status. Note the wide bony bridge between the talus and calcaneus. *Bottom* 1 year after resection of the bridge and fat interposition. No further bridge formation has occurred

canal coalition. An *MRI scan* shows a characteristic, hyperintense subchondral zone, even if the coalition is only fibrous [43].

### Treatment

The treatment of symptomatic tarsal coalition is surgical. The operation always involves resection of the bony, fibrous or cartilaginous bridges. The procedures vary only in the way in which the resulting gap is filled. Normally, fatty tissue is interposed, although some authors have also used tendons for this purpose [16, 39]. The advantage of fatty tissue is that it does not show any tendency to ossify. Langenskiöld introduced this procedure to prevent the renewed formation of bone bridges in the area of the growth plates. Experimental studies have shown that the free fatty tissue transplant can be replaced by viable fat cells [27]. The results of resection of the coalition are usually good if the joint is actively moved post-operatively. Recurrences occur in less than 10% of cases [36]. We usually leave the epidural catheter in place for several days while administering intense physiotherapy. After the patient is discharged, movement of the rearfoot must continue to be actively assisted on a daily basis for several months.

#### 3.4.5.3 Polydactyly

##### ► Definition

The presence of more than 5 toes on one foot. The supernumerary toe(s) can be in differing stages of development.

##### Occurrence, etiology

Polydactyly of the feet (and of the hands) is a relatively common malformation. The incidence among the white population is roughly 30:100,000 [51], and it occurs more frequently in girls than in boys. Substantial differences exist between races. The incidence calculated for the black population is 30:100,000 [51], compared to 45:100,000 for Indians [34]. Most cases of polydactyly occur as an isolated deformity, unilaterally or bilaterally on the feet alone or on both hands and feet. The condition is inherited as an autosomal recessive trait. A large-scale epidemiological study involving 5927 Brazilian children with polydactyly [10] found that associated anomalies were present in 14.6% of cases, most rarely with postaxial polydactyly and most commonly with central polydactyly (over 50% in the latter instance). Duplication of the 5th toe is the commonest abnormality. 75% of the polydactylies associated with syndromes involved either trisomy 21, trisomy 13 or Meckel syndrome. The latter is a rare autosomal recessive disorder involving, in addition to the polydactyly, abnormalities of the kidneys, liver and CNS. Isolated cases

of polydactyly are also observed in Ellis-van-Creveld syndrome.

### Classification

The *traditional classification* is as follows:

- *Preaxial*: Duplication on the side of the great toe
- *Central or axial*: Duplication in the area of toes 2 – 4
- *Postaxial*: Duplication on the side of toe 5

The commonest forms of polydactyly are postaxial, less common are preaxial duplications, while the axial type is extremely rare [9].

### Classification according to Blauth [6]

*Syndrome classification* in two directions: *longitudinal and transverse*:

- The transverse axis refers to the affected toe (1, 2, 3, 4, 5).
- The longitudinal axis refers to the site of the duplication: distal phalanx, middle phalanx, proximal phalanx, metatarsus, tarsus.

### Clinical features, diagnosis

The diagnosis is always obvious even at birth. Supernumerary toes are usually most striking if they are not aligned in parallel with the other toes (■ Fig. 3.393). Since such protruding toes will cause problems when footwear is worn and are very unsightly, the decision to remove them will usually be made before the child starts walking. If the bifurcation occurs at the level of the metatarsals, the toes will often show parallel alignment, but the foot will be much wider than normal. This can also pose problems when it comes to the provision of footwear (■ Fig. 3.394). On the great toe side, the supernumerary toe usually shows pronounced varus deviation (■ Fig. 3.395). This deviation often occurs at the metatarsophalangeal joint, causing both sections of the duplicated great toe to show varus alignment (congenital hallux varus, see ► chapter 3.4.5.7). Polydactylies are not infrequently combined with *syndactylies*. An association seems to exist between duplication of the hallux and anterior bowing of the tibia. The latter resembles the deformity encountered with congenital pseudarthrosis of the tibia, but has a much better prognosis [31].

### Treatment

The ideal age to remove supernumerary toes is between 9 and 12 months. Simple resection of the surplus toes is generally sufficient (particularly for the postaxial forms). The surgeon should be careful to ensure that the scar does not occur on the lateral edge of the foot, where it could rub uncomfortably against footwear. If both part-

## 3.4.5 · Other congenital anomalies of the foot



■ Fig. 3.393. Polydactyly in a 1-year old child. This is a *postaxial* type with 7 toes

ner toes in a case of postaxial polydactyly are roughly equivalent, it is worthwhile removing the medial ray. Although this is technically more difficult, it does result in a more stable and sturdier lateral foot margin [35]. On the medial side, simple resection of the supernumerary toes is not usually sufficient, since a varus deformity is additionally present and the 1st metatarsal is often shortened. The deviation of the metatarsophalangeal joint occasionally needs to be corrected by means of an osteotomy (■ Fig. 3.395). If the 1st metatarsal is severely shortened, a lengthening osteotomy with an external fixator may be necessary.



■ Fig. 3.394. Polydactyly with duplication of the 1st metatarsal in a 16-year old girl. Left preoperatively, right after removal by chiseling of the medially projecting part in the area of the 1st metatarsal head



■ Fig. 3.396. After separation of syndactyly with scar-related adhesion on the great toe



■ Fig. 3.395a–c. Female patient with polydactyly of the *preaxial* type (DP views). a Duplicated great toe at the age of 6 months before resection of the medial ray. b At the age of 16 years a pronounced hallux val-

gus is present due to the asymmetrically formed metatarsophalangeal joint. c After subcapital corrective varization osteotomy

### 3.4.5.4 Syndactyly

#### ► Definition

Missing or incompletely formed web space between two toes.

#### Occurrence

Syndactyly of the feet, though not as common as syndactyly of the hands, is not a rare deformity, and also occurs particularly in connection with polydactyly.

#### Clinical features, diagnosis, treatment

Although syndactyly is not associated with cosmetic or functional disadvantages, the parents of the affected child often ask for the anomaly to be removed. This is where the idea mentioned in ► chapter 1.1 applies, i.e. that a congenital abnormality is a punishment from God, a visible sign of original sin. Moreover, such parents are rarely persuaded by the argument that other people are hardly ever aware of the syndactyly or that it is even possible to be married to a spouse for 20 years without the latter realizing that their partner has syndactyly on a foot. *Surgical treatment* is therefore strongly *discouraged*, because the risk of complications in the foot are much greater than those in the hands. Postoperatively, it is not possible to keep the resulting web space as dry as one between the fingers, ultimately leading to potential wound adhesions and scar formation, which can then (in contrast with the original syndactyly) cause functional problems (► Fig. 3.396).

### 3.4.5.5 Split foot (Lobster claw foot)

#### ► Definition

Hypoplasia or the absence of one or (usually) several central ray(s) on the foot.

#### Occurrence

An epidemiological study in Hungary calculated an incidence of 1.3 in 100,000 neonates [14]. Boys are more frequently affected than girls, and the right side is more affected than the left [14]. An autosomal dominant component has been detected in some cases [14]. Split feet have also been observed in connection with tibial aplasia. The autosomal dominant form with incomplete penetrance is always bilateral, while the unilateral form is not associated with any detectable inherited component. The hereditary form is frequently associated with cleft hand, possibly also with cleft lip and palate or with syndactyly and polydactyly, and possibly with deafness [38].

#### Classification

Blauth proposed the *classification* shown in ► Table 3.68 [8]. The development of split foot starts on the 2nd or 3rd ray and progresses in a distal to proximal direction [8].

► Table 3.68. Classification of split foot. (According to Blauth [9])

Type	Features
I	2nd–4th toes missing, normal metatarsals
II	2nd–4th toes missing, all metatarsals present, but partially hypoplastic
III	Only 4 metatarsals present
IV	Only 3 metatarsals present
V	Only 2 metatarsals present
VI	Monodactylous split foot



► Fig. 3.397a, b. Split feet, left type II, right type IV in a 16-year old boy. a Clinical picture, b X-ray



### 3.4.5 · Other congenital anomalies of the foot

Only the 5th ray is present in a monodactylous split foot. The defect is always greater at the distal end compared to the proximal end. Occasionally, synostoses are found at the edges of the cleft.

#### **Clinical features, diagnosis**

The diagnosis of split foot is easy and always apparent just from the outward appearance (■ Fig. 3.397). The type of split foot can be classified with the aid of an x-ray. No further diagnostic investigation is required. In functional respects, split feet are usually very efficient since the rays that bear the main weight, 1 and 5, are invariably present (except in the monodactylous type).

#### **Treatment**

Surgery is relatively rarely indicated for split foot [7]. In some cases the split feet may be so wide that shoes cannot be fitted. In such cases, an osteotomy to narrow the foot is indicated. Occasionally, other complex corrections must be performed or interfering elements removed. In general, however, such indications are rare and most split feet can be left in their natural state since the patients are not particularly affected in functional terms. Of course, the cosmetic appearance is always unsatisfactory, but this cannot really be improved without substantial effort. Naturally the patients will avoid being seen barefoot in public.

### 3.4.5.6 Congenital ball-and-socket ankle joint

#### ► **Definition**

Congenital deformity of the ankle joint with dome-shaped deformation of the talus and a rounded distal end of the tibia.

#### **Occurrence**

This is a very rare deformity, and the author is not aware of an epidemiological study. Only isolated cases are reported in the literature [5, 22, 47]. The anomaly appears to be slightly more common in Japan and is usually associated with tarsal coalition [5, 22, 47], although it may also be accompanied by other foot deformities.

#### **Etiology**

If tarsal coalition is present, the development of the upper ankle as a ball-and-socket joint is a secondary phenomenon [5, 22, 47]. The loss of mobility in the lower ankle leads to a secondary change in the upper ankle: instead of just being able to move in one axis of movement, it acquires the ability, over time, to move in 2 axes, as has been confirmed by the findings on x-rays recorded from birth to early childhood [5, 22, 47]. Tarsal synostoses are particularly common in connection with longitudinal deficiency of the fibula (► Chapter 3.3.6.1). If no tarsal

coalition is present, the underlying mechanism is unclear. An increased frequency of valgus deformity of the distal femur has also been observed [37].

#### **Clinical features, diagnosis**

While a congenital ball-and-socket ankle joint does not usually produce any symptoms, it can lead to lateral instability and thus an increased incidence of supination-related trauma. Together with the loss of mobility in the subtalar joint, this can lead to premature osteoarthritis in the ball-shaped ankle joint. The latter has a characteristic appearance on the x-ray (■ Fig. 3.398): On the AP view, the talus is convex and lacking its flattened, slightly concave shape. The distal ends of the tibia and fibula have adapted themselves to this shape.

#### **Treatment**

No causal treatment is known. Symptomatic treatment is based on the tarsal coalition (► Chapter 3.4.5.2). If substantial symptoms are present and if osteoarthritis develops, an arthrodesis may be necessary in adulthood.



■ Fig. 3.398. AP x-ray of the upper ankle in congenital ball-and-socket ankle joint in an 11-year old boy



### 3.4.5.7 Congenital hallux varus

#### ➤ Definition

Congenital medial deviation of the 1st metatarsophalangeal joint and the great toe.

#### Occurrence

This is a fairly rare deformity that is occasionally combined with a shortened 1st metatarsal. We are not aware of any epidemiological data. Congenital hallux varus is very typically seen in polydactyly with duplication of the great toe. More common than the congenital form is secondary hallux varus, which occurs secondarily after overcorrection in soft tissue procedures for hallux valgus. This form also frequently occurs in middle age in people who walk barefoot [24]. Secondary hallux varus is also very typically seen in a residual clubfoot with rigid supination of the forefoot. Since the ball of the great toe cannot provide support, the great toe is drawn inward until it is able to provide support (▶ Chapter 3.4.3).

#### Clinical features, diagnosis

The diagnosis is confirmed by clinical examination: The great toe deviates medially to a greater or lesser extent (■ Fig. 3.399) and may show supination. The hallux varus causes symptoms while footwear is worn. Just putting on the shoe can cause problems, and pressure points can also occur over the tip of the toe or the interphalangeal joint. Always note any supination of the forefoot. If no callus has formed under the ball of the great toe, then the hallux varus is secondary and not congenital.

#### Treatment

The treatment of congenital hallux varus is always surgical. The aim is to restore the normal axis of the great

toe, which can usually be achieved with a subcapital osteotomy of the 1st metatarsal (■ Fig. 3.399). An opening wedge osteotomy with insertion of a medial wedge is the best procedure since the 1st metatarsal is often too short. If the bone is of normal length, a closing wedge osteotomy with the removal of a lateral wedge is also appropriate. If the deformity is very pronounced, the osteotomy should be performed in the area of the medial cuneiform bone, again supplemented by the insertion of a wedge. Postoperatively we transfix the great toe with a Kirschner wire. Sometimes the soft tissues must also be lengthened medially. The correction of congenital hallux varus is not easy and there is also a certain risk of recurrence. The correction of secondary hallux varus due to supination of the forefoot is addressed in ▶ chapter 3.4.3.

### 3.4.5.8 Macroductyly

#### ➤ Definition

Disproportionate growth of one or more toes.

#### Occurrence

Macroductyly is a very rare deformity. If several toes or the whole foot are affected, there may be an underlying disease such as neurofibromatosis, Proteus syndrome or congenital hemihyperplasia [3, 18, 33].

#### Clinical features, diagnosis

Macroductyly (■ Fig. 3.400) is a very troublesome deformity as shoes are difficult to fit because of the protruding toe. The condition is diagnosed by clinical examination. The bone structures are unchanged provided no additional malformations are present.



■ Fig. 3.399. Congenital hallux varus in an 8-year old boy with dysplasia of the 1st metatarsal and synostosis between the 1st and 2nd metatarsals. Left preoperatively; right after division of the synostosis and corrective osteotomy



■ Fig. 3.400. Macroductyly of the right 2nd toe in an 8-year old girl. Syndactyly of the left 3rd and 4th toes is also present

### 3.4.5 · Other congenital anomalies of the foot

#### Treatment

The main problem concerns footwear provision. Differing shoe sizes often have to be worn on the left and right foot. The orthotist must fill the gap in front of the normally growing toes with padding, otherwise the shoe will not fit properly. A greatly enlarged toe can be made smaller by a multi-step surgical procedure: First, the proximal phalanx is removed and the growth plate of the middle phalanx is closed. The fatty tissue on one side is removed, and the toe is fused with the adjacent toe. In the second step, the fatty tissue on the other side is removed. If the metatarsal is also enlarged, the epiphyseal plate can be closed here too. The timing of the operations is difficult. Amputation should be avoided as a rule, otherwise axial deviation of the adjacent toes can occur, which can also lead to symptoms.

#### 3.4.5.9 Brachymetatarsia

##### ➤ Definition

Congenital shortening of a single metatarsal bone.

##### Occurrence

The shortening of a single metatarsal in isolation is not all that rare. Generally the 1st metatarsal is affected. No epidemiological data are available however. It should be borne in mind that the 1st metatarsal is not always longer than the 2nd metatarsal, but can also be slightly shorter, even in the normal foot. In 40% of feet, the 1st metatarsal is longer than the 2nd, while the reverse applies in another 40%, and both metatarsals are the same length in the remaining 20%.

##### Clinical features, diagnosis

Slight differences in the length of individual metatarsals are usually of no clinical relevance. A more pronounced shortening, however, can impair the statics of the foot. Significant shortening of the 1st metatarsal, for example, interferes with the normal heel-to-toe roll, since the ball of the great toe bears most of the weight during this maneuver. If the metatarsal is too short, the weight cannot roll correctly over the ball of the great toe. Shortening of the middle rays can also impair the transverse arch and occasionally cause symptoms.

##### Treatment

If the heel-to-toe roll mechanism is impaired and symptoms are present, lengthening of the shortened metatarsal may occasionally be indicated, ideally by means of a small external fixator and callotaxis. However, the extent of the lengthening should not exceed 40% of the length of the metatarsal, otherwise there will be a high risk of stiffening of the metatarsophalangeal joint [32].

#### 3.4.5.10 Accessory muscles

##### ➤ Definition

Additional muscles in the retromalleolar region as accessory muscle bellies of the flexor digitorum longus, flexor hallucis accessorius longus or soleus accessorius muscles. These are normal congenital variants.

##### Occurrence

Two studies with cadavers have shown an incidence of 5% for accessory muscles in the retromalleolar area [50]. This is therefore a common anomaly, but one that is rarely diagnosed.

##### Clinical features, diagnosis

Clinical examination reveals an asymptomatic thickening in the hollow alongside the Achilles tendon. Although no symptoms are involved, an awareness of these accessory muscles is important as achillodynia is very common in active sporting adolescents and the possibility of accessory muscles must be borne in mind in the differential diagnosis of swelling in the retromalleolar area [21]. The diagnosis can be confirmed by a CT or MRI scan.

##### Treatment

Since this is a normal variant without any pathological significance treatment is not required.

#### 3.4.5.11 Foot abnormalities with longitudinal malformations (fibular or tibial deficiency)

##### ➤ Definition

The combination of femoral hypoplasia or a proximal focal femoral deficiency (► Chapter 3.2.7.2) with fibular hypoplasia or aplasia (► Chapter 3.3.6.1) and the absence of lateral foot rays is known as a *longitudinal deficiency of the fibula (fibular deficiency)*. If the tibia is hypoplastic or aplastic (► Chapter 3.3.6.2) and the medial foot rays are missing, the condition is known as a *longitudinal deficiency of the tibia (tibial deficiency)*. In the much more common fibular deficiency, the 5th ray alone may be missing or several rays from the lateral to the medial side may be deficient. Because of the malformed fibula, instability and valgus deviation of the upper ankle is usually present, very often combined with talo-calcaneal coalition. In the much rarer condition of tibial deficiency, the 1st ray, or several rays from the medial to lateral side may be missing.

##### Occurrence

The incidence of fibular deficiency was calculated to be 2:100,000 neonates in one epidemiological study [40], although far from all patients show foot abnormalities in addition to the fibular defect.

**Clinical features, diagnosis**

The foot abnormality is always combined with a hypoplasia or absence of the fibula and a shortening of the lower leg. At birth the foot is usually in an equinovalgus position and one or more lateral rays of the foot are missing. The bone structures of the rearfoot may be incompletely formed and tarsal coalition is present. In the much rarer condition of tibial hemimelia, the 1st ray or the 1st and 2nd rays may be defective. In this anomaly the foot is in a varus-equinus position. The x-ray shows the defectively formed bone structures and the bone connection if tarsal coalition is present (■ Fig. 3.401). The latter is very commonly seen in longitudinal deficiency of the fibula, as is the formation of a congenital ball-and-socket ankle joint [5, 47].

**Treatment**

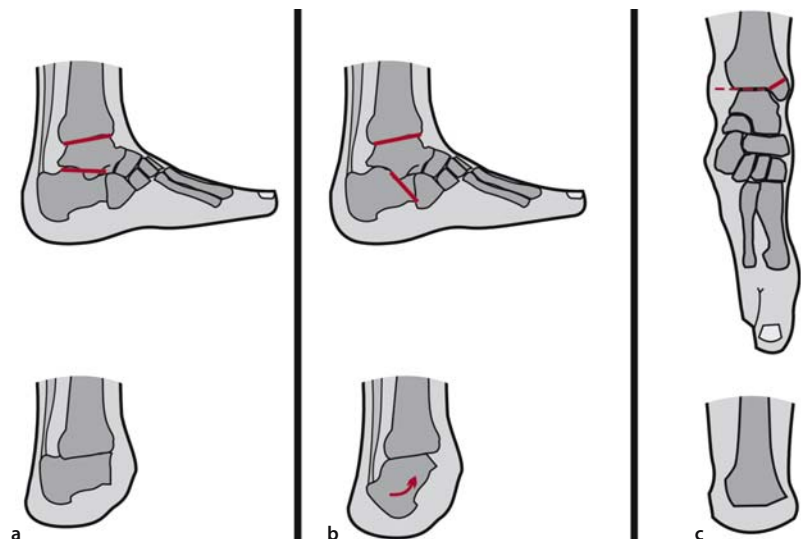
The main problem associated with fibular deficiency is stabilization of the ankle. If a type I B or type II fibular

hemimelia is present (▶ Chapter 3.3.6.1), the ankle is unstable and the rearfoot shows severe valgus deviation. If the fibula is completely absent and if no more than 3 lateral foot rays exist then preservation of the foot is not usually a sensible option. In such cases the patients should be gently encouraged to agree to a Syme amputation (■ Fig. 3.402), although this is not always acceptable to the parents or child. The orthotic management of such a foot is usually extremely difficult, since muscle tension always tends to cause the talus to subluxate laterally. If good heel control can be achieved it is generally possible to guide the foot to a certain extent and place it in the orthosis in an equinus position. If an attempt is made to put the foot in a plantigrade position, however, it will prove even more difficult to prevent the valgus deviation. If the foot can be preserved until the completion of growth, a stable plantigrade foot position can then be achieved with an arthrodesis of the ankle joint (or possibly a triple arthrod-

■ Fig. 3.401. X-rays of the left foot in a 9-year old boy with fibular deficiency. Fibular aplasia is present and two rays of the foot are missing on the fibular side. The main problem is the absence of stability in the upper ankle



■ Fig. 3.402a–c. Amputations in the area of the mid- and rearfoot: a According to Boyd: The calcaneus is fused with the tibia in a horizontal position, the mid- and forefoot are amputated. b According to Pirogoff: The calcaneus is aligned vertically and fused with the tibia in this position, the mid- and forefoot are amputated. c According to Syme: The foot is amputated at the upper ankle with resection of the medial malleolus



### 3.4.5 · Other congenital anomalies of the foot

esis). If the ball of the great toe is normal, then a largely physiological heel-to-toe roll is possible and walking will not be significantly impaired. Of course, the patient is unlikely to be able to run or jump. The equinus foot deformity can sometimes be corrected with the Ilizarov apparatus, although this procedure is only appropriate if the ankle joint is stable. Lengthening of individual rays is almost never indicated. An important requirement for the orthosis is a good foot bed with filling of the gaps caused by the missing rays.

#### 3.4.5.12 Foot abnormalities in systemic disorders

##### **Apert syndrome (Acrocephalosyndactyly)**

###### 🔴 **Definition**

Simultaneous occurrence of synostoses on both hands and feet, and also on the skull and spine. This is an autosomal dominant hereditary disorder (for a detailed description see ► chapter 4.6.3.1).

###### **Clinical features, diagnosis**

The foot deformity in Apert syndrome is very characteristic, and its development can be predicted. The synostosis is progressive. The great toe becomes increasingly shorter and deviates in a medial direction. The phalanx becomes delta-shaped [30]. The mobility of the metatarsophalangeal joint gradually deteriorates during the course of growth. The metatarsus and rearfoot also increasingly show bony connections between the individual bones. The 5th metatarsal is prominent with callus formation beneath its head. Other metatarsal bones may also pro-

trude in a plantar direction. The defective mobility very rapidly leads to the formation of painful pressure sites (🔴 Fig. 3.403).

###### **Treatment**

The treatment of Apert syndrome is a multidisciplinary problem requiring collaboration between orthopaedists, microsurgions, neurosurgeons, plastic surgeons, pediatricians, orthotists and psychologists. The treatment of the foot is determined by the severity of the patient's symptoms. It should be borne in mind that the extremely rigid feet in a patient with Apert syndrome almost rule out completely any possibility of compensation. Accordingly, the aim is to facilitate the heel-to-toe roll as much as possible. If the great toe shows pronounced medial deviation or the metatarsal head shows plantar protrusion, it may be necessary to prevent any additional handicap by means of an osteotomy (🔴 Fig. 3.403b). Occasionally a ray has to be shortened. In view of the lack of compensation options, surgery is indicated relatively often [1]. Secure bedding of the feet in the shoe, possibly with orthotic support, is an important requirement.

###### **Foot abnormalities in various syndromes**

Foot abnormalities are observed in various syndromes. Thus macrodactyly, syndactyly and polydactyly all occur in Klippel-Trenaunay syndrome (► Chapter 4.6.6.4) and Proteus syndrome (► Chapter 4.6.6.3) [3, 18, 33]. A metatarsus varus and clinodactyly are also frequently observed. The ring constriction syndrome (amniotic band syndrome) [15] can be associated not only with clubfoot deformities, but also vascular malformations and trophic disorders of the foot (🔴 Fig. 3.404).



🔴 **Fig. 3.403a, b.** Type III *Apert syndrome* in a 16-year old girl. **a** Clinical picture. Note the lack of segmentation between the toes, the medial deviation of the great toes and the plantar projection of the 2nd metatarsal on the right. **b** Oblique views of the right foot, *top* exostosis

on the 1st metatarsal and plantar deviation of the 2nd and 3rd metatarsals, *bottom* after removal by chiseling of the exostosis on the 1st metatarsal and osteotomy of the 2nd and 3rd metatarsals



**Fig. 3.404.** AP and lateral x-rays of the right foot in a 10-month old girl with *amniotic band syndrome*. Note the constriction at metatarsal level and the rudimentarily formed and incompletely segmented toes

The Prader-Willi syndrome (▶ Chapter 4.6.5.8) is occasionally associated with congenital pes planus. In fibrodysplasia ossificans progressiva (▶ Chapter 4.6.2.31), anomalies of the great toes are invariably present, although in widely varying forms. Toenail changes are generally observed in pterygium syndrome (▶ Chapter 4.6.5.2). Foot abnormalities also commonly occur in diastrophic dwarfism (▶ Chapter 4.6.2.6): In 43% of cases a metatarsus adductus is present, in 37% an equinovarus adductus and, in 8%, an equinus deformity [41]. There is increasing evidence to suggest that amniocentesis can cause foot abnormalities, particularly if it is performed before the 13th week of pregnancy [52].

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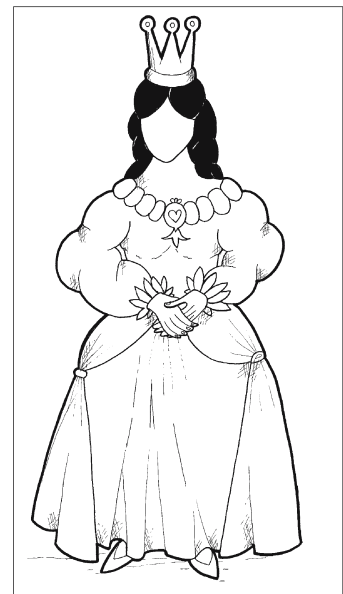


### 3.4.6 · Do skewfeet stop Cinderella from turning into a princess?

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### 3.4.6 Do skewfeet stop Cinderella from turning into a princess? or: Should one treat metatarsus adductus?

» *It happened, however, that the king appointed a festival which was to last three days, and to which all the beautiful young girls in the country were invited, in order that his son might choose himself a bride... After the festival the king's son looked for the beautiful girl with whom he had danced the whole evening. He picked up the small, dainty golden slipper that the girl had worn and then lost. The slipper didn't fit any woman's foot throughout the land. Except Cinderella, on whose foot the slipper fit like a glove. Then the prince knew whom he wanted as his wife... « (Brothers Grimm)*



Metatarsus adductus is the commonest foot deformity in infants. It almost always develops only *after* birth as a result of the unequal muscle tension on the medial and lateral sides of the foot. Parents are understandably worried and often think that the problem will persist into adulthood if nothing is done.

#### ► Definition

Metatarsus adductus = adduction of the forefoot in relation to the rearfoot in the infant.

— **Synonyms:** Pes adductus, postural metatarsus adductus, skewfoot

#### Occurrence

Metatarsus adductus is a common deformity of the foot that is not usually present at birth but only develops during the first few weeks of life. Interestingly it is only

observed in children who are born at term. Metatarsus adductus does not occur in premature neonates [3]. Precise epidemiological data are not available.

### Etiology

Various factors are responsible for the development of metatarsus adductus. The fact that it does not occur in premature neonates [3] suggests that restricted space in the uterus may play a role. Metatarsus adductus was observed more frequently in the 1980's and 1990's, when children were regularly placed in the prone position. Resting the feet on a blanket also appears to play a role in the development of metatarsus adductus. Since babies have been consistently placed on their back in recent years, because of the risk of sudden infant death, we are now observing metatarsus adductus less frequently. The constitutional adduction of the metatarsals is another etiological factor. The importance of the muscles as a triggering component should not be underestimated. Since small children lack muscle balance, the adductor hallucis, tibialis anterior and tibialis posterior muscles are stronger in relation to the peroneal muscles and therefore promote the adduction tendency of the forefoot.

### Clinical features, diagnosis

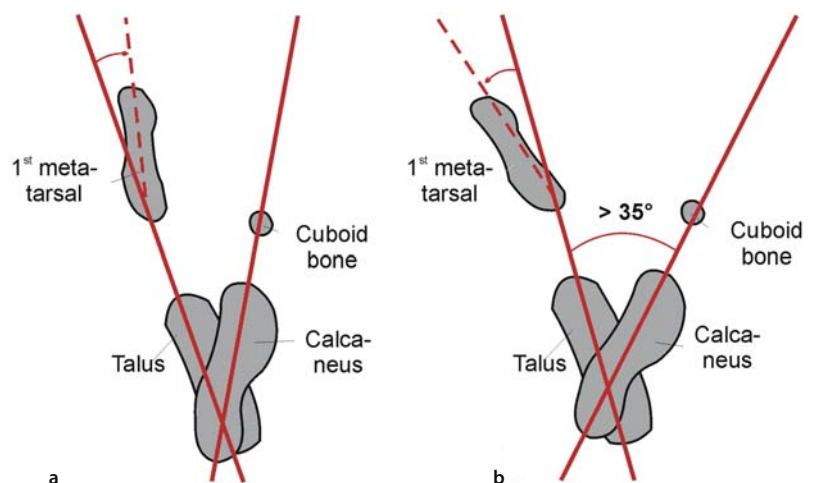
The most obvious clinical finding is the adduction of the forefoot in relation to the rearfoot. The examiner should note whether the rearfoot is in an abnormal valgus position or not to establish whether skewfoot or a standard metatarsus adductus is present. However, the inward turning of the foot can be caused by other factors: The internal rotation of the tibia can likewise produce a foot axis that is rotated inwards in relation to the thigh axis (► Chapter 3.4.1.1). The orthopaedist must carefully establish whether the inward rotation originates in the foot

itself or the lower leg, or whether a combination of both factors is involved. The distinction is important because it has consequences for treatment.

Medial torsion of the tibia is promoted by hyperactivity of the medial muscles (adductor hallucis, tibialis anterior and tibialis posterior muscles). An x-ray is not necessary for a case of straightforward metatarsus adductus. Radiographic clarification is recommended only if the presence of skewfoot is clinically suspected. MRI studies of skewfoot have shown that lateral subluxation of the navicular bone and medial subluxation of the 1st metatarsal is present even before the child starts walking [2]. The radiographic assessment of the foot is explained in ► Fig. 3.405. A clinical example of a skewfoot is shown in ► Fig. 3.406. A simple method of documenting the metatarsus adductus is to stand the child on the glass plate of a photocopier (provided the child is not too heavy) [7].

### Prognosis, treatment

Whether treatment is required for a simple case of metatarsus adductus is a matter of dispute. Naturally the need for treatment is closely connected with the prognosis. Although very few studies are available on the natural history of metatarsus adductus, one of the few serious investigations has shown that, out of 130 untreated feet with metatarsus adductus, 14% were still deformed after 7 years [6]. In a more recent study, 8% of 85 feet treated with below-knee casts still showed a residual deformity after an average of 4 years [4]. Thus, most cases of metatarsus adductus will return to normal spontaneously. Of the persistent cases, some will go on to develop a juvenile hallux valgus, particularly if a 1st metatarsal is in the varus position [1]. Another long-term study over 32.5 years and involving 31 patients showed that a slanting angle of the joint between the 1st metatarsal and the cuneiform



► Fig. 3.405a, b. Radiological assessment of the infant foot according to Berg. a Normal configuration; calcaneus and talus form an angle of less than 35° and the axis of the 1st metatarsal is either parallel to the talar axis or turned inwards. b In *pes adductus* the axis of the 1st metatarsal is turned outwards compared to the talar axis. In *skewfoot* the talocalcaneal angle is also greater than 35°

## 3.4.6 · Do skewfeet stop Cinderella from turning into a princess?

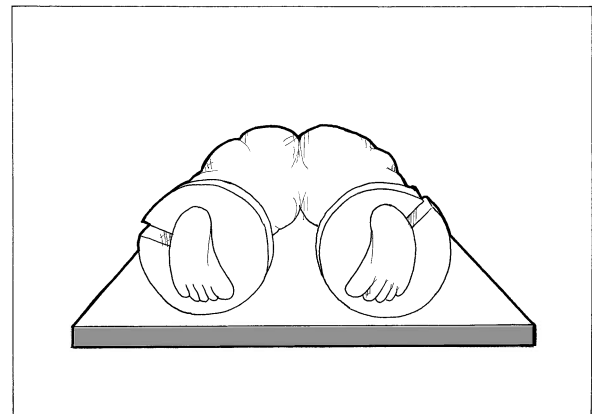


■ **Fig. 3.406.** 9-year old boy with *skewfeet*: In addition to hypervalgus of the rearfoot, there is adduction of the forefoot with hallux varus, but normal weight-bearing under the ball of the great toe (in contrast with Fig. 3.379)

bone persisted in 68% of cases, but that juvenile hallux valgus was relatively rare [1]. There is no doubt that metatarsus adductus is a benign deformity, even though it can lead to subsequent problems in individual cases during adolescence and adulthood. Since the treatment is simple and not especially irksome we tend to opt for cast treatment of pronounced cases of metatarsus adductus in order to prevent a later persistent and troublesome problem.

Overall, however, treatment is only required in very few children with metatarsus adductus. A simple and cost-effective preventive measure is the use of foam rings (■ Fig. 3.407). Such rings prevent the foot from resting against the bed, and thus accentuating the adduction of the forefoot, when the child is in the prone position. But this is only a preventive measure. Actual treatment is indicated if the abnormal position cannot be eliminated by stimulation of the lateral edge of the foot or the application of slight medial pressure. Treatment involves cast correction and is unproblematic in infants who are not yet able to walk. Only after the child starts to walk do the casts prevent motor development to a greater extent. We therefore try to complete the treatment before the onset of walking.

While a below-knee cast would seem to be sufficient in a simple case of metatarsus adductus [4], the risk of the cast slipping down and causing pressure points in very



■ **Fig. 3.407.** Use of *foam rings* in infants with metatarsus adductus in the prone position. The rings prevent the feet from resting on the mattress and consequent exacerbation of the metatarsus adductus

small children is considerable. We therefore generally prepare long-leg casts with a flexed knee joint for small children (see ■ Fig. 3.375). Such long-leg casts can also positively influence any medial torsion of the tibia, which would not be the case with below-knee casts.

We generally use Softcast for preparing a cast for metatarsus adductus. The cast does not need to be as accurately shaped as for a case of clubfoot, and the Softcast can be removed by the mother with little effort (and without the noise of the cast saw). Below-knee casts are sufficient however for larger children. The heel must be carefully shaped and correct the position of the forefoot in relation to the rearfoot. For this corrective treatment we use either the traditional plaster cast or Scotchcast, since Softcast is not so easy to shape.

The treatment of *skewfoot* is more problematic [2, 5]. Fortunately this deformity is relatively rare. Surgical treatment is occasionally indicated if the midfoot is in pronounced supination [5]. We consider that a soft tissue operation (medial release) is ineffective in cases of metatarsus adductus. If substantial adduction actually persists into later childhood, the combined closing wedge cuboid osteotomy and opening cuneiform osteotomy is a more effective way of correcting the deformity (► Chapter 3.4.3).

### References

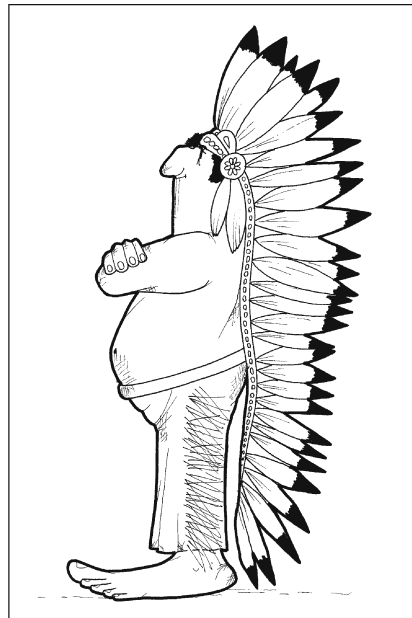
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#### 3.4.7 Flatfoot Indians – which ones must be treated so that they can later become chiefs? – or: How do we distinguish between flat valgus foot and flexible flatfoot?

Concern about »flatfeet« is one of the commonest reasons prompting a mother or father to take their children to a pediatrician or orthopaedist.

Like the back and the knee, the foot is also often used in everyday linguistic usage in a symbolic sense. Although certain figures of speech are emotionally colored, the actual shape of the foot is not used to represent a characteristic trait of the person in question. When we are anxious about the outcome of a development we get cold feet. When a situation has turned out favorably we have fallen on our feet. Someone who thinks on his feet is capable of making good decisions quickly. Someone who drags his feet is unnecessarily delaying a decision. To put one's foot down is to exert one's authority. In a new situation we have to find our feet, while in a busy situation we are rushed off our feet.

The aesthetic qualities of the foot are also worth bearing in mind. Footwear is strongly influenced by fashion trends. The importance that we attach to the beauty of



the naked foot is subject to cultural differences. In southern countries (for obvious reasons), parents' concern about a normal foot shape is much greater than in North America or northern Europe, where the foot tends to go uncovered only at night. Accordingly, inserts or even custom-made shoes are produced in substantial quantities in Italy and Spain, and surgical procedures involving the foot are also much more common in these countries than north of the Alps.

Naturally it is concern about problems with walking in later life that primarily prompts the visit to the doctor. We must distinguish between various conditions. A common feature is the reduction in size, or complete absence, of the arch over the weight-bearing area in the center of the medial edge of the foot. The differing forms of flatfoot and valgus foot are listed in ■ Table 3.69.

This chapter deals with physiological flat valgus foot, its differentiation from flexible flatfoot and their differential diagnosis. The other conditions involving a flattened medial arch are addressed in other chapters (see notes in ■ Table 3.69).

#### 3.4.7.1 Physiological flat valgus foot

##### ► Definition

Increased valgus position of the heel and flattening of the longitudinal arch in children, compared to adults, as a result of increased anteversion of the femoral neck.

##### Etiology

Children show more pronounced anteversion of the femoral neck compared to adults. This forces them to

■ Table 3.69. Differential diagnosis: flattened longitudinal arch of the foot

	Etiology	Clinical findings	Investigation	Treatment
<b>Congenital flatfoot</b> Congenital flatfoot, congenital convex pes valgus, congenital vertical talus, congenital rocker-bottom flatfoot (chap. 3.4.4)	Congenital	Present at birth Valgus position of the heel Abduction and pronation of the forefoot Missing longitudinal arch Sole poss. with convex curve (»rocker-bottom flatfoot«) Shortened Achilles tendon Poss. palpable talonavicular subluxation	Clinical X-ray: vertical talus	Operation during the 1st year of life (talar reduction) Cast and orthotic treatment for 2–3 years
<b>Flexible flatfoot</b> Severe pes planovalgus, flexible flatfoot, flexible pes planovalgus	Ligament laxity Constitutional disposition Overweight Muscle weakness	No medial arch over weight-bearing surface Restricted dorsal extension Shortened Achilles tendon Increased valgus of the rearfoot Hyperpronation of the forefoot Foot often normal at birth	Clinical X-ray: Increased talocalcaneal angle	Initially conservative (casts) Possibly inserts, high medial supports with heel control and supination wedge Possibly operation (calcaneal lengthening osteotomy, at the earliest after the start of walking) Postoperative cast and orthotic treatment for 2–3 years
<b>Contracted flatfoot</b> Rigid flatfoot, talocalcaneal coalition; (chap. 3.4.5.2)	Tarsal coalition (usually talocalcaneal coalition)	Manifestation at 8–10 years, heel in hypervalgus, no mobility in the lower ankle	Clinical X-ray (possibly oblique views) CT	Surgical: Resection of the bone fractures and fat interposition
<b>Physiological flat valgus foot</b>	Increased antever-sion of the femoral neck	Increased valgus of the rearfoot Hyperpronation of the forefoot	Clinical	Reassure the parents No treatment required
<b>Neuromuscular pes planovalgus</b> Neuromuscular flatfoot; (chap. 3.4.10)	Neuromuscular functional disorder	Shortening of the Achilles tendon Heel in very pronounced valgus Hyperpronation of the forefoot Hallux valgus	Clinical Neurological investigation X-ray: Secondary damage?	Orthoses with heel support Possibly surgical lengthening of the triceps surae Possibly double arthrodesis

adopt an intoeing gait. To avoid tripping over their own feet, the child unconsciously tries to correct its intoeing gait by turning the feet outwards. This external rotation of the weight-bearing foot results in valgization of the heel and flattening of the foot arch. Anyone can try it for themselves: Turning your upper body, and thus the lower leg as well, internally over the weight-bearing foot will produce valgus rotation of the heel and, automatically, hyperpronation of the forefoot with flattening of the longitudinal arch.

### Diagnosis, measures

Because of the mechanism for correcting the intoeing gait, children's feet appear unlike adult's feet, a fact that often worries parents. Not infrequently, the mothers and fathers

had to wear shoe inserts during their own childhood and therefore consult the doctor in order to ensure that their own child doesn't miss out on any necessary treatment. During the examination we check whether a medial arch is present or not. Note that this arch is lacking in children under 3 years of age because of the fat pad and that the foot arch may not be properly delineated until the age of 6. With the child standing on tiptoe, we can also observe how the foot arch forms with the varization of the heel. Naturally the height of the longitudinal arch is subject to considerable variation. Some patients will show a relatively deep arch even in adulthood, but this is not associated with any morbidity, i.e. the risk of subsequent foot problems is not increased at all. The early provision of medi-ally-supporting inserts will not influence the height of the



foot arch. In a study conducted in our hospital, the results for the group of patients fitted with inserts were worse than those for those without inserts [7]. This is explained by the fact that the inserts make the muscles »lazy« and the active development of the arch is less likely to occur than when inserts are not fitted. Even the wearing of shoes has an adverse effect on the development of the foot arch [13]. The problem of flat valgus foot is accentuated by overweight, by an abnormal valgus axis of the lower leg and by general ligament laxity [9]. The transitions to flexible flatfoot are blurred. *Genua valga* (► Chapter 4.2.1) can be influenced to a slight extent by fitting a wedge to the medial side of the sole of the shoe. *Genua valga* will usually persist, however, if obesity is present. All children and adolescents with weight problems should be given dietary counseling, and a successful outcome depends greatly on the extent to which the parents also cooperate. There is no treatment for general ligament laxity, which must be accepted as a given, although the problem will tend to decline steadily over time.

❗ **A general recommendation in a case of childhood flat valgus foot is to walk barefoot a lot, and to rely on the fact that the foot arch will develop spontaneously with the physiological derotation at the femoral neck and that any slightly lowered arch will not have any negative effect on the performance of the foot in adulthood.**

*Childhood flat valgus foot is a physiological condition and should not be a reason for extensive and useless diagnostic and therapeutic measures.*

### 3.4.7.2 Flexible flatfoot

#### ➤ Definition

Foot condition with a *missing medial arch* over the weight-bearing surface, *restricted dorsal extension*, a *shortened Achilles tendon*, an *increased valgus position* of the rearfoot and *hyperpronation* of the forefoot. However, the foot remains flexible, and the medial arch can be restored by *varization* of the heel. An *increased talocalcaneal angle* is measured on the lateral x-ray.

— *Synonym: Talipes planovalgus*

#### Occurrence

Since a substantial grey area exists between a clear case of flatfoot with a missing longitudinal arch and predominant weight-bearing on the medial side of the sole of the foot and a case of physiological childhood flat valgus foot, and since the classification of individual cases is usually arbitrary, calculating the frequency of flexible flatfoot poses considerable problems. Add to this the fact that the fat pad on the sole of the foot fills out the medial arch during the first few years of life, which makes any differentiation

difficult from the outset. Only when the child reaches the age of around 6 years does the weight-bearing surface correspond to that of an adult [14]. It can be stated confidently, however, that flexible flatfoot is a rare condition (in this part of the world it probably affect fewer than 1 in a thousand children) if one accepts as a criterion the fact that the medial arch is completely filled in on a footprint recorded in a school-age child. One interesting study in India showed that the widening of the weight-bearing surface occurred 3 times more frequently in children who regularly wore shoes than in those who always walked barefoot [13].

#### Etiology

The most important etiological factor is general ligament laxity which, during weight-bearing, causes the talus to tilt over the calcaneus in a medial and caudal direction. The calcaneus then pronates into an extreme valgus position. The problem is accentuated if the child is overweight. Any general muscle hypotonia will also contribute to the accentuation of the deformity. One reported rare cause of flexible flatfoot is injury to the tendon of the tibialis posterior muscle [11].

#### Associated anomalies

Any condition involving general ligament laxity can also be accompanied by flexible flatfeet. This particularly applies to the Ehlers-Danlos syndrome (► Chapter 4.6.6.7), although children with other syndromes, e.g. Down syndrome (► Chapter 4.6.4.1), Rubinstein-Taybi syndrome (► Chapter 4.6.3.3) or Marfan syndrome (► Chapter 4.6.6.5), also show an increased incidence of flexible flatfoot. The flexible flatfoot is often accompanied by a shortening, or even dislocation, of the peroneal tendon.

#### Diagnosis

Children with a flexible flatfoot do not experience any pain. It is the outward appearance of the foot that prompts the parents to visit the doctor. Symptoms may occur during adolescence (particularly if the patient is overweight). Examination reveals the absence of the medial arch over the weight-bearing surface. Callusing of the sole of the foot is often observed at this site. A worthwhile exercise is to view the foot on a podoscope through a glass plate or prepare a *footprint* by placing a sheet of paper with a carbon paper backing (■ Fig. 3.408). Two forms can be differentiated: In the milder form, the medial arch is missing, but the foot shape is normal and weight-bearing is regular (■ Fig. 3.408b), in the more severe form, the talus protrudes medially and weight-bearing is greater on the medial than on the lateral side (■ Fig. 3.408c). In the most severe form, lateral weight-bearing is completely absent (■ Fig. 3.408d).

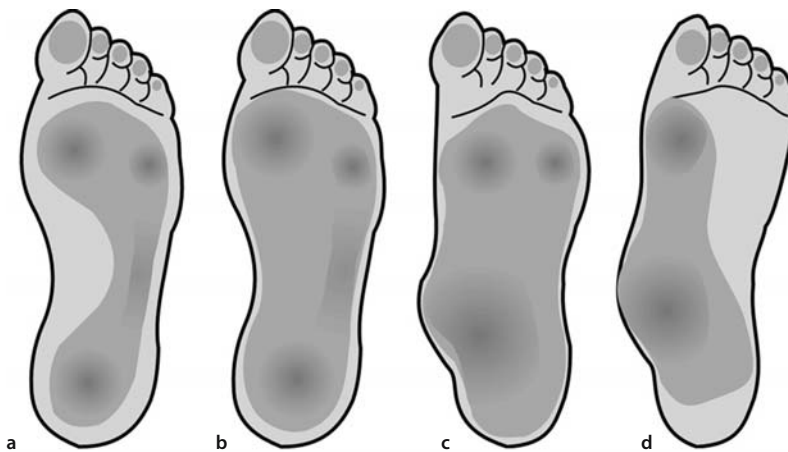
It should be noted, however, that the absence of the medial arch is physiological after the child starts to walk,

## 3.4.7 · Flatfoot Indians – which ones must be treated so that they can later become chiefs?

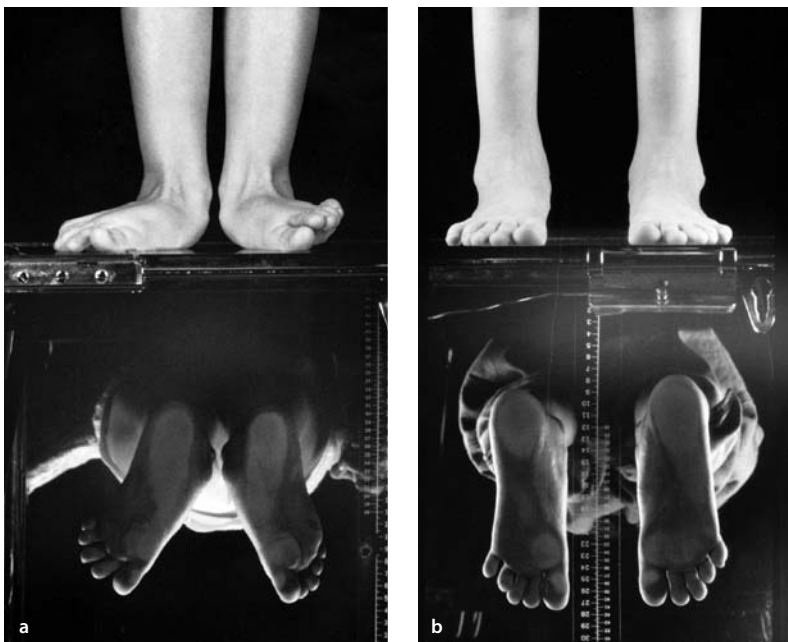
since the longitudinal arch is filled out with fatty tissue. This tissue declines during the first few years of life. By the age of around 3 years, the medial arch starts to appear on the footprint, although it is only fully formed by the age of 5 or 6 [15]. Once the child starts walking, a case of flexible flatfoot can be diagnosed only if weight-bearing is greater on the medial than on the lateral side or if the lateral x-ray of the standing patient shows a talocalcaneal angle greater than  $60^\circ$  (■ Fig. 3.409).

On *clinical examination* the foot arch becomes visible when the child stands on tiptoe or, while the child is standing with normal weight-bearing, when the great toe is pushed up (■ Fig. 3.410). Another examination finding

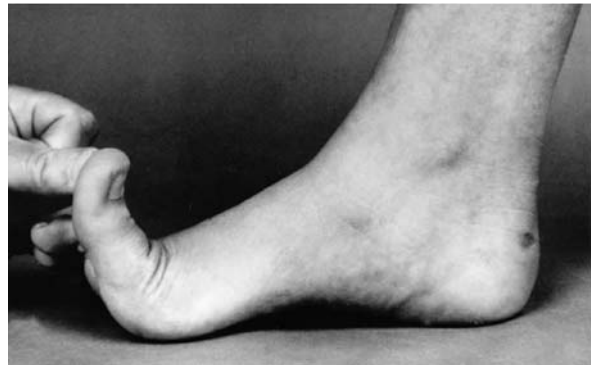
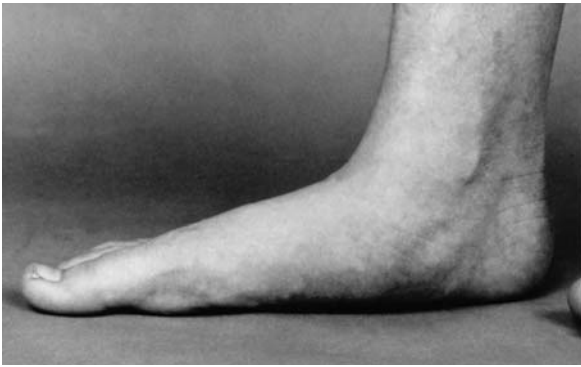
is *general ligament laxity*. This can be established on the basis of the thumb-to-forearm distance, hyperextensibility of the long fingers of over  $90^\circ$  at the metacarpophalangeal joint and hyperextensibility of the elbow and knee joints. The severe forms of flatfoot are also regularly accompanied by *shortening of the calf muscle*. This can be masked by the fact that the child is able to extend the foot dorsally at the ankle, but this is only possible with pronounced valgization of the heel, which shortens the travel of the Achilles tendon. If the heel is held in the neutral position, however, dorsal extension via the plantigrade position is no longer possible. The shortening of the calf muscles is always secondary. Placing the heel in the valgus position



■ Fig. 3.408a–d. Footprints in normal foot and various degrees of flatfoot. a Normal foot with no weight-bearing under the medial arch. b Flexible flatfoot with flattened medial arch, but otherwise normal weight-bearing pattern with callusing mainly under the heel and the 1st and 5th metatarsal heads. c Severe flatfoot with weight-bearing medially under the talus. d Very severe flatfoot with eversion and pronation of the whole foot and abduction of the forefoot and complete absence of weight-bearing on the lateral side



■ Fig. 3.409a, b. Feet on the podoscope in a case of severe flexible flatfoot (same patient as in Fig. 3.411). a At the age of 2 years, pre-operatively. Weight-bearing occurs almost exclusively on the medial side, the forefoot is abducted. b At the age of 4 years, 2 years after open reduction of the talus, lengthening of the triceps surae and orthotic provision. The feet now show a normal weight-bearing pattern



**Fig. 3.410.** Flexible and rigid flatfoot can be distinguished by *pushing up the great toe*. In a flexible flatfoot the medial longitudinal arch is

produced with dorsal extension of the great toe, which is not the case with rigid flatfoot



**Fig. 3.411a, b.** Lateral x-rays of both feet of a patient with severe flexible flatfoot. **a** At the age of 2 years. Note the enlarged talocalcaneal angle on both sides. **b** 2 years after open reduction of the talus, lengthening of the triceps surae and orthotic provision. Normal configuration has been restored

causes the travel of the Achilles tendon to be shorter than normal even in the plantigrade position. Dorsal extension causes the heel to pronate even more into a valgus position, thereby canceling the physiological stretching of the calf muscles.

On the *x-ray*, the talocalcaneal angle is increased on both the AP and lateral views (**Fig. 3.411**); on the lateral view this should be greater than 60° for a diagnosis of flexible flatfoot. The talus is usually relatively steep on the lateral x-ray. The calcaneus is often largely horizontal and lacks the normal upward slope from a dorsal-caudal to ventral-cranial direction, which is an indication of the shortening of the Achilles tendon. In order to produce meaningful results, the x-rays of the foot should be recorded with the child standing and weight-bearing. Additional imaging investigations are not necessary in a case of flexible flatfoot. Only if a rigid rearfoot is present would there be a need for further investigation to exclude a bone coalition, which is often not visible on the plain x-ray. CT scans are usually indicated in such cases, possibly incorporating oblique views as well (**Chapter 3.4.7**).

**Treatment**

**!** Before deciding on a treatment, the orthopaedist must carefully consider whether any treatment is even necessary.

Mild forms of flexible flatfoot do not usually lead to any significant functional problems even in adulthood, nor are they painful. These patients are also usually able to participate in sports without any restrictions [17]. In children, flatfoot is more of a cosmetic than a functional problem, and one that worries the parents much more than the children themselves. Only severe forms of flatfeet, in which weight-bearing is greater on the medial than on the lateral side, adversely affect function over time. Particularly if obesity is also present, troublesome foot symptoms can become a long-term problem.

### 3.4.7 · Flatfoot Indians – which ones must be treated so that they can later become chiefs?

The following therapeutic options are available:

- **Conservative:**
  - cast treatment,
  - foot exercises, walking barefoot, stretching of the calf muscles,
  - loose inserts,
  - shoe modifications,
  - lower leg orthoses.
- **Surgical:**
  - navicular suspension with or without naviculocuneiform arthrodesis,
  - lengthening of the triceps surae muscle and/or Achilles tendon,
  - extra-articular subtalar arthrodesis according to Grice,
  - insertion of a dowel implant in the tarsal sinus,
  - calcaneal varus osteotomy,
  - calcaneal lengthening osteotomy according to Evans
  - double arthrodesis.

A summary of the measures for the various conditions is shown in ■ Table 3.69.

#### Conservative treatment

##### Infancy

If the flexible flatfoot is associated with an abduction of the forefoot, it is occasionally manifest even at birth. In such cases it is worth straightening the foot with a cast during the first few months of life. We use long-leg casts as below-knee casts can easily slip down and lead to pressure sores. Moreover, the correction of the foot is better with a long-leg cast. We generally use Softcast since the parents can then remove the cast without any difficulty. In this form of correction the rearfoot is pushed in a varus direction and the forefoot is supinated and adducted. At the same time the medial longitudinal arch is shaped by the cast. As a rule, we start corrective treatment only after the 2nd month of life and continue the treatment until the foot shape has returned to normal, generally after 2–3 months, by which time the foot has a normal shape in the non-weight-bearing state. Whether a flexible flatfoot will continue to persist after the start of walking cannot be predicted with certainty since this depends to a great extent on the quality of the ligaments – and this is difficult to assess in the infant.

##### Walking age

If a flexible flatfoot persists after the start of walking, the possibility of inserts can be considered. The foot is particularly difficult to assess at this age since the medial foot arch is filled with fatty tissue, which means that no arch is visible on the footprint even in the normal foot. We make a diagnosis of flexible flatfoot at this age only if weight-

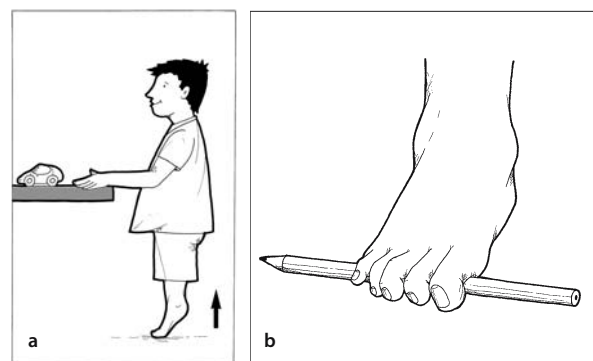
bearing is greater on the medial than on the lateral side or if the x-ray shows the corresponding signs.

A measure that is always useful is to recommend *walking barefoot a lot* as this strengthens the foot muscles. Another appropriate measure is to provide instruction in *foot exercises* that the mother can playfully encourage the child to practice. However, this is not the time to prescribe physical therapy. Only rarely is the physical therapist successful in getting the still very small child to follow her instructions, and exercises are only useful if they are practiced several times a day. It is pointless therefore to place this unnecessary financial burden on the public healthcare system (or the insurance funds).

Useful foot exercises include walking on tiptoe and heel-walking. Walking on tiptoe is an ideal way of training the foot muscles (■ Fig. 3.412), while heel-walking stretches the calf muscles. While other types of exercise (e.g. lifting objects with the toes) may liven up the daily training program, they are not particularly important for foot function.

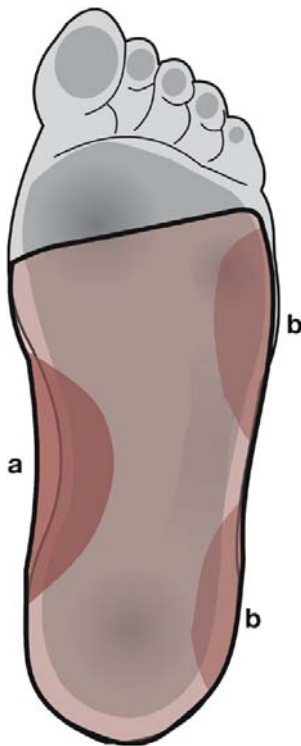
If shortening of the triceps surae muscle is already present, special *stretching exercises for the calf muscles* should be provided. In this case, physical therapy is appropriate since the stretching in flexible flatfoot is effective only if the heel is simultaneously placed in a varus position, which the child is unable to achieve on its own. Moreover, the mother may be unable to manage this exercise, or at least will require careful instruction.

We consider that the *provision of inserts* is appropriate in pronounced forms of flexible flatfoot. Although the efficacy of insert treatment has not been completely proven scientifically, we nevertheless manage feet with fallen medial arches with inserts or shoe modifications. Various studies have shown that the outcome is no different for treated and untreated feet [16, 17]. A study conducted in our own hospital with two groups of approx. 20 children with fallen arches with and without insert



■ Fig. 3.412a, b. *Foot exercises.* a The simplest and most useful exercise is to stand on tiptoe. b Exercises with the child can be made more fun by competing with the child to grasp long objects with the toes





■ Fig. 3.413. Loose *derotation insert* with medial support (a). Lateral counter supports are inserted to prevent the foot from sliding laterally (b). The »diagonal cut«, i.e. the fact that the insert is longer at the front on the lateral side slightly corrects the child's intoeing gait

treatment showed that the end result was not influenced by the supports [7]. However, all these studies involved mild forms of flexible flatfoot or even physiological flat valgus foot. Other studies, by contrast, have shown that the supporting of the medial arch and varization of the heel with an insert or shoe modification certainly does produce an effect in more pronounced forms of flexible flatfoot [12]. The underlying principle is that, by lowering the talus and navicular bone, the tendons on the medial side of the foot (particularly that of the tibialis anterior muscle) are constantly overstretched, thus preventing them from performing their postural function at all. Our prescription for the insert is as follows: *Derotation insert with central medial arch support and a supination wedge*. The supination wedge places the heel in a varus position (■ Fig. 3.413).

Even more effective is an insert with its own heel support. The effect can be enhanced still further by incorporating the corrective function directly in the shoe. Such a shoe modification can control the foot more precisely than a loose insert. On the other hand, a shoe modification is much more expensive than an insert since it must be produced specifically for a particular shoe. Children with flatfeet have a high shoe consumption rate, i.e. they wear out their shoes quickly. The heel in particular wears

down quickly on the lateral side. Hopes that the insert or shoe modification will reduce the shoe consumption rate will be disappointed. The varization wedge, for example, has no effect in this respect [12]. If the heel is in a very extreme valgus position, a so-called inner shoe, i.e. a lower leg orthosis with heel control can prove useful.

### Surgical treatment

Operations for cases of flexible flatfeet may be indicated in very severe forms (in which weight-bearing is greater on the medial than on the lateral side) and if symptoms are present. The desire for cosmetic improvement can also be taken into account to a certain extent, although considerable caution is required here since the correction of the appearance should not be achieved at the expense of pain. If surgery is indicated the operation should not be performed before the age of 8, or preferably 10.

### Talar reduction

In the severest forms, in which the weight-bearing of the foot occurs predominantly, or exclusively, on the medial side, treatment is often required even during early childhood. As with vertical talus (► Chapter 3.4.4), the oblique talus and navicular are reduced, the talonavicular joint is transfixed, the triceps surae is lengthened and the dislocation pouch is closed on the medial side. Consistent, long-term medial support (initially with casts and later with orthoses) is required as follow-up treatment.

### Navicular suspension

In this operation, which was first proposed by Lowman in 1923 [10], the anterior tibial tendon is looped around the navicular bone. This makes the tendon too short relatively, thereby enhancing the tensioning effect. The relocation of the attachment dorsally also causes the tendon to exert a more direct effect on the medial arch of the foot. The problem with this operation is that the underlying condition of these patients tends to involve pronounced ligament laxity and consequently the greater tensioning effect soon starts to decline. Recurrences are frequent after this operation. Lowman himself was also aware of this and proposed an additional arthrodesis of the talonavicular joint. However, since this completely eliminates the mobility in the lower ankle, it is probably an inadequate measure in most cases of flexible flatfoot.

A less drastic, and apparently equally effective, procedure is arthrodesis of the joint between the navicular and medial cuneiform bones. The combination of navicular suspension and naviculocuneiform arthrodesis is practiced in some places in patients with an almost fully-grown foot (i.e. from the age of 12). We do not have any experience with this procedure. If the calf muscles are shortened, this operation should be combined with an aponeurotic lengthening of the triceps surae muscle.



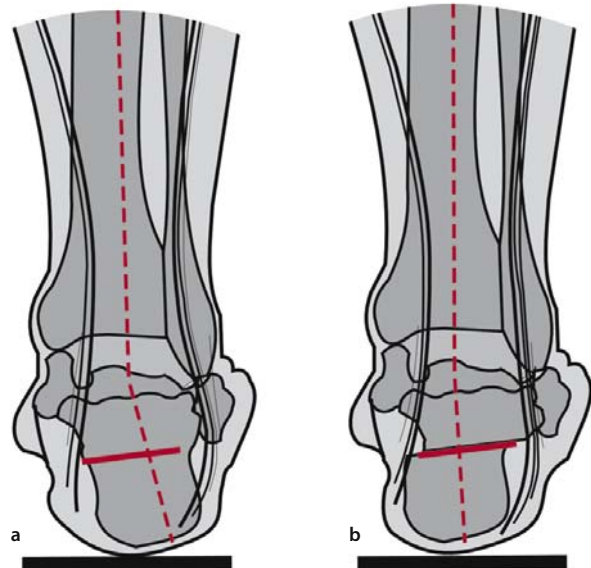
### Extra-articular subtalar arthrodesis according to Grice and dowel implant operation according to Giannini (arthrorisis)

As we have already mentioned under »Etiology«, the pathological mechanism in flatfoot involves a tilting of the talus over the calcaneus in a medial and caudal direction. The calcaneus pronates into an extreme valgus position and the calcaneus and talus are angled abnormally in relation to each other. If the rotation of these two bones into this valgus position is prevented, then the medial longitudinal arch of the foot can be preserved. This is the principle underlying the extra-articular subtalar arthrodesis according to Grice [4]. In this procedure a bony union between the talus and calcaneus is created in the tarsal sinus, i.e. outside the lower ankle. This union prevents the valgus tilting of the heel bone. The drawback of this method is that the lower ankle is permanently stiffened. For this reason Giannini proposed the implantation of a plastic dowel into the tarsal sinus [3].

Other authors have used a similar implant [14] or inserted a bone graft taken from the calcaneus into the tarsal sinus. The disadvantage of the bone graft, however, is that it either leads to fusion of the joint or is reabsorbed. The dowel implant permits a certain degree of residual mobility between the talus and calcaneus and can also be removed at a later date if required by the situation. We ourselves have used these dowel implants in limited numbers, but subsequently abandoned their use as they caused problems in most patients over time (e.g. pain, dislocation of the implant). This procedure must likewise be combined with aponeurotic lengthening of the triceps surae muscle if the calf muscles are shortened.

### Calcaneal varus osteotomy

Since the abnormal valgus position of the calcaneus is an important component of flexible flatfoot, a varus osteotomy of the calcaneus, as proposed by Dwyer [1], can correct this aspect. The procedure can be performed as an opening wedge osteotomy from the lateral side (N.B.: caution is required during wound closure) or as a closing wedge osteotomy from the medial side. Since the calcaneus is of normal length in flatfoot – in contrast with clubfoot – the closing procedure is unproblematic. The result can be fixed with titanium staples, thereby allowing early mobilization in a walking cast. An even simpler surgical procedure is the method described by Koutsogiannis [8] in which the dorsal part of the calcaneus is transferred medially (■ Fig. 3.414). However, the postoperative stability is not as good after this procedure, the result must be fixed with a Steinmann pin and a prolonged period of immobilization is required. The drawback associated with the calcaneal osteotomies is that the abnormal tilt between the talus and calcaneus is not corrected, which means that a significant part of the deformity remains. This is where



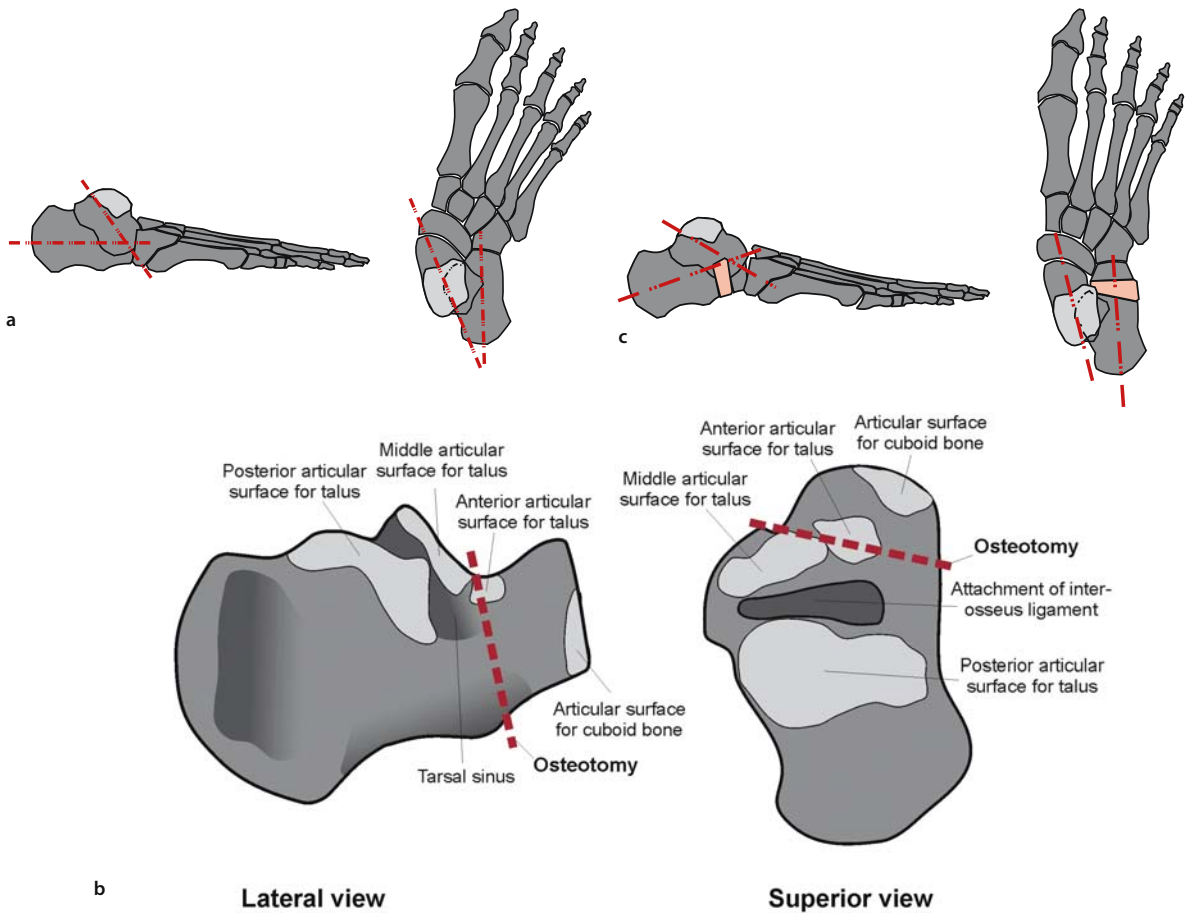
■ Fig. 3.414a, b. Principle of the *medially transferring calcaneal osteotomy* in a pronounced case of calcaneus valgus

the calcaneal lengthening osteotomy according to Evans offers clear advantages.

### Calcaneal lengthening osteotomy according to Evans

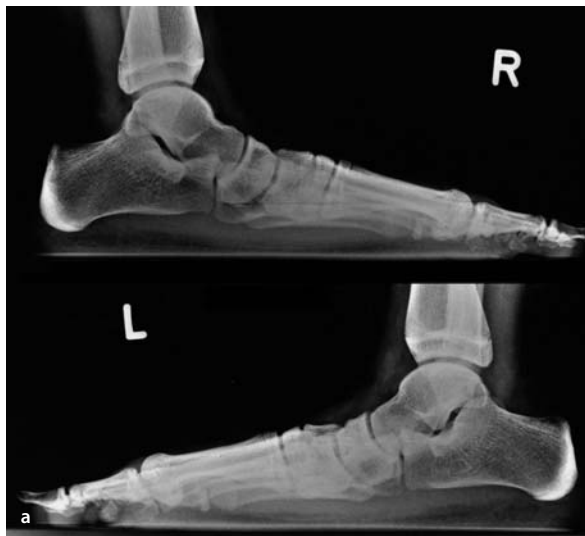
The principle involves the correction of a pes planovalgus or skewfoot by the insertion of an autologous or allogeneic bone wedge with a lateral base in the neck of the calcaneus behind the calcaneocuboid joint. We consider that the lengthening osteotomy described by Evans [2] is indicated on the one hand for severe neuromuscular pes planovalgus (► Chapter 3.4.10) or for idiopathic flexible flatfoot, particularly if weight-bearing under the talus is greater on the medial than on the lateral edge of the foot [5, 6]. Occasionally, the operation may also be useful for managing a case of skewfoot. The operation should not be performed before the age of 8.

The Evans procedure involves a calcaneal osteotomy at the level of the tarsal sinus in the frontal plane from the lateral side and the insertion, on the lateral side, of an allogeneic or autologous bone graft (■ Fig. 3.415). The graft should be slightly less wide on the medial side than on the lateral side. This not only lengthens, but also adducts the calcaneus and places it in a slightly more varus position. The lengthening of the calcaneus anteriorly causes the plantar aponeurosis to be placed under tension and thus promote the formation of the foot arch (■ Fig. 3.416 and 3.417). This operation effectively prevents the tilting of the talus over the calcaneus and, in our view, is currently the most useful surgical procedure for a case of severe flatfoot that is not based on a congenital deformity or a tarsal coalition.



**Fig. 3.415a–c.** Principle of the calcaneal *lengthening osteotomy according to Evans*. **a** Schematic view of a flatfoot from the top and laterally with lacking upward slope of the calcaneus and steeply sloping talus, abduction of the forefoot. **b** Joint surfaces of the calcaneus. The

osteotomy is performed from the lateral side between the middle and anterior talar articular surfaces. **c** Flatfoot after extension and insertion of a bone graft from the lateral side. The position of the talus and calcaneus and the abduction of the forefoot are corrected

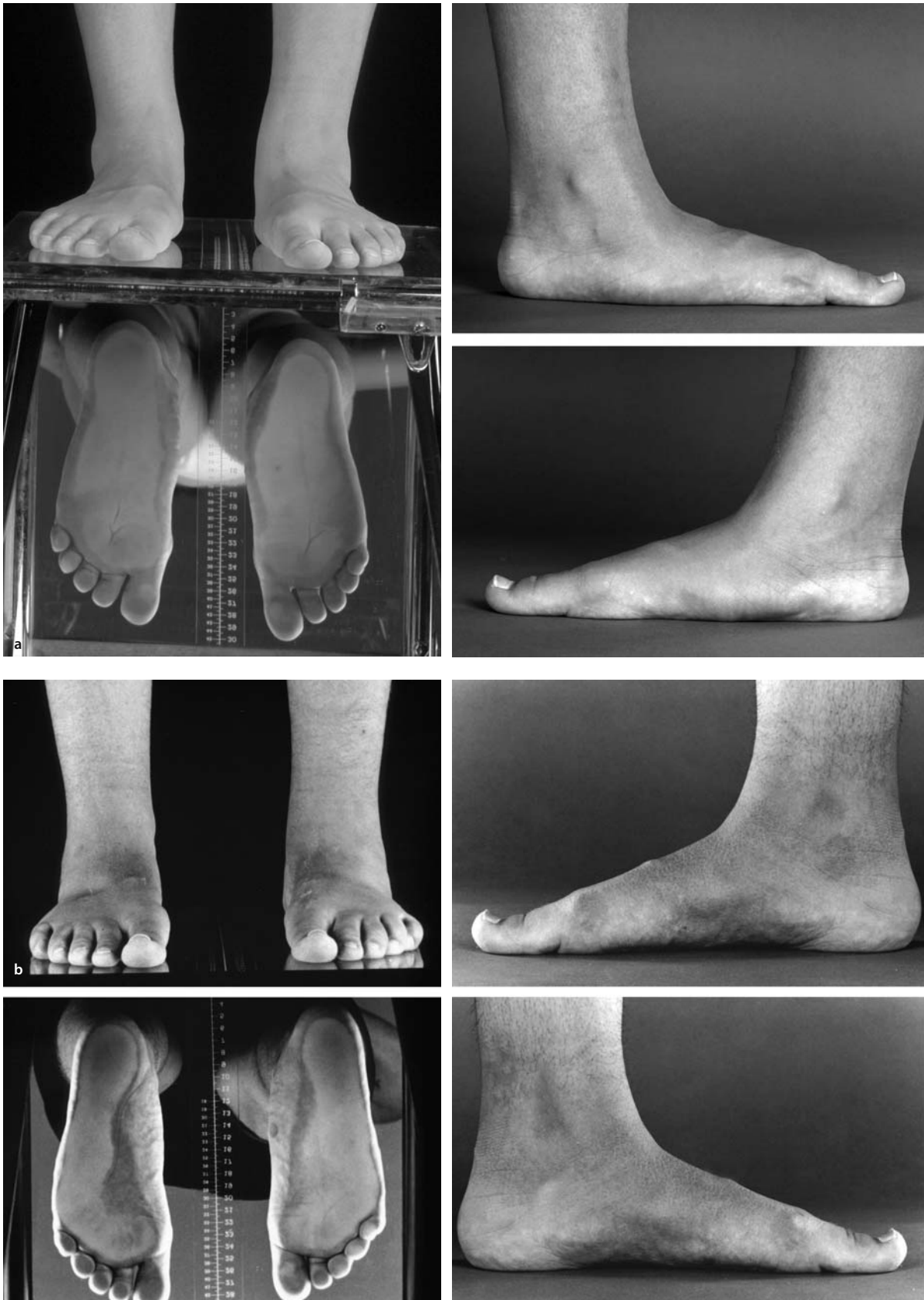


**Fig. 3.416a, b.** Lateral x-rays in a 13-year old girl with *flexible flatfoot*. **a** Preoperative situation with flattened longitudinal arch. **b** After calcaneal *lengthening osteotomy according to Evans*. The talus is no longer



able to tilt medially, the foot arch is restored as a result of tensioning of the plantar aponeurosis. Mobility is preserved in the lower ankle

## 3.4.7 · Flatfoot Indians – which ones must be treated so that they can later become chiefs?



■ Fig. 3.417a, b. Flexible flatfeet on the *podoscope* (same patient as in Fig. 3.416). a Preoperative situation with medial weight-bearing and

no arch. b Situation 6 months postoperatively. The feet now show a largely normal weight-bearing pattern

### Triple arthrodesis

The triple arthrodesis involves stiffening of the subtalar joint and the talonavicular and calcaneocuboid joints. In German-speaking countries this operation is known as a »double arthrodesis« to avoid confusion with a triple arthrodesis that also involves the ankle joint. In the triple arthrodesis the joint surfaces in the anterior part of the subtalar joint and the surfaces of the talonavicular and calcaneocuboid joint are resected and a large autologous bone graft is inserted. This operation is generally not indicated until adulthood, i.e. for flatfeet that have become painful and rigid. We have never performed this procedure on children or adolescents.

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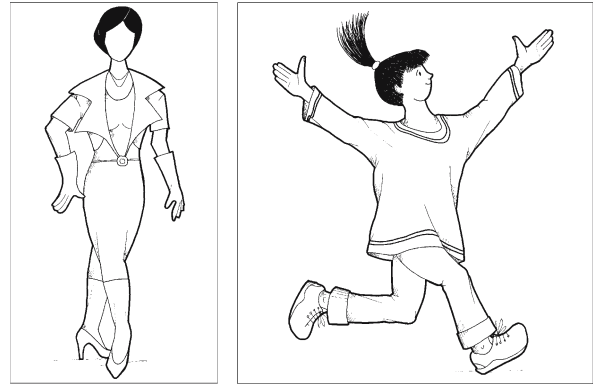
### 3.4.8 Juvenile hallux valgus

» If a crooked toe is the child's fate, an insert won't make it straight «

#### Definition

Valgus deviation of the great toe in adolescents due to an adduction deformity of the 1st metatarsal.

— **Synonym:** Adolescent hallux valgus



The hallux valgus is not least a question of fashion. Some mothers are skeptical of today's trainer generation, but the shoes into which they squeezed their feet as teenagers were much more harmful...

#### Occurrence

In an epidemiological study involving 6000 schoolchildren, unilateral and bilateral hallux valgus were found in 36 and 60 cases respectively. This corresponds to an incidence of 1.6% [8]. Girls are 5 times more frequently affected than boys [8].

#### Etiology

The following *etiological factors* have been discussed:

- **Constitutional metatarsus varus:** The varus deformity, i.e. an angle between the 1st and 2nd metatarsals greater than 14° [8] results in increasing valgus deviation of the great toe. A contributory factor is a slanting position of the joint between the medial cuneiform and the 1st metatarsal.
- **Hypermobility in the 1st metatarsal-medial cuneiform joint:** This also promotes the increasing adduction and varus position of the 1st metatarsal [6].
- **Metatarsus adductus in infancy:** Metatarsus adductus in early childhood is considered by some to be a contributory factor in juvenile hallux valgus, although the scientific proof for this is lacking.
- **Peak pressure under the hallux and the head of the 1st metatarsal:** Measurements in 61 children between the ages of 5 and 16 years were made on a pressure platform. Girls were found to have significantly greater



### 3.4.8 · Juvenile hallux valgus

peak pressure under the hallux, faster timing of heel contact to first metatarsal head loading, and a more medially placed center of pressure trajectory [5]. The association with the development of hallux valgus, however, remains unclear.

The following factors have been discussed but their *causal significance* for juvenile hallux valgus is now *rejected*:

- **Flexible flatfoot:** One investigation has shown that the height of the foot arch is not a relevant factor for the development of juvenile hallux valgus [9].
- **Shape of the head of the 1st metatarsal:** An analysis of 50 patients has shown that the shape of the head of the 1st metatarsal has no pathogenetic significance for the development of hallux valgus [7].
- **Splayfoot:** The flattening of the transverse arch of the foot due to chronic overloading and connective tissue weakness does not play a major role in juvenile hallux valgus, in contrast with the situation for degenerative hallux valgus.

#### Diagnosis

Clinical examination reveals a – generally clearly visible – valgus deformity of the great toe. The medial side of the head of the 1st metatarsal appears inflamed and swollen and is often painful, particularly when tight-fitting shoes are worn. The mobility of the metatarsophalangeal joint of the great toe is not restricted in adolescents. The great toe is usually slightly pronated. If the deformity is very pronounced, the toe can slide under the 2nd toe, causing the latter to be superducted. This often leads to additional inflammation and sore points on the toes. Although varus of the 1st metatarsal can occur even if the great toe is not in a valgus position, this situation almost never leads to symptoms and does not therefore require treatment from the functional standpoint [4]. Hammertoes do not form part of the clinical picture of juvenile hallux valgus but are the typical consequence of splayfoot in adults.

The DP *x-ray* of the foot of the standing patient shows medial deviation of the 1st metatarsal, valgus deviation of the proximal and distal phalanges of the great toe and often a slanting position of the joint between the medial cuneiform and the 1st metatarsal. The following angles can be measured to produce a more objective result: between the 1st and 2nd metatarsals, between the 1st metatarsal and the 1st proximal phalanx I, between the 1st proximal phalanx and the foot axis, which usually corresponds to the axis of the 2nd metatarsal (■ Fig. 3.418). Of course, the measuring error associated with these calculations on the foot x-ray is considerable [2].

#### Differential diagnosis

A special form of hallux valgus is *valgus deviation of the 1st distal phalanx* in children. This is a constitutional de-



■ Fig. 3.418. DP x-rays of both feet of a 14-year old girl with *juvenile hallux valgus*. The joint between the 1st metatarsal and the medial cuneiform is at an oblique angle, the 1st metatarsal is in an exaggerated varus position and the great toe is in a valgus position. The angle between the 1st and 2nd metatarsal is increased (left 17°, right 14°)



■ Fig. 3.419. DP x-rays of both feet in a 12-year old boy with *valgus deviation of the distal phalanx*

formity that generally causes no symptoms and does not require treatment (■ Fig. 3.419).

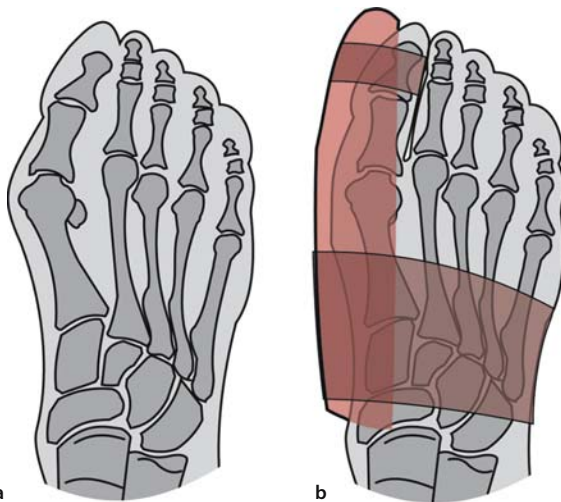
#### Treatment

In contrast with hallux valgus due to splayfoot in adults, juvenile hallux valgus is rarely accompanied by symptoms. Caution is therefore required in deciding whether therapeutic measures are indicated.

The following *treatments* are available:

- splints,
- inserts,
- soft tissue operations,
- osteotomies.





**Fig. 3.420a, b.** *Hallux valgus nocturnal splint:* In contrast with splay-foot-induced hallux valgus in adults, the use of a nocturnal splint can be useful in adolescents. The splint must extend sufficiently far back so that, in addition to the great toe, the deviating 1st metatarsal is also corrected. **a** Situation without, **b** with splint

### Conservative treatment

The value of *insert treatment* for juvenile hallux valgus is extremely dubious since the cause of hallux valgus is not a splayed foot, i.e. flattening of the transverse arch, but rather a malposition of the 1st metatarsal. Consequently, raising the transverse arch with a retrocapital support will not resolve the problem. The inefficiency of such an insert has also been demonstrated in a randomized study [10]. A more promising option is a *splint* worn at night (Fig. 3.420). In contrast with the situation for inserts, the efficacy of such splints has been proven [7]. They cannot be worn during the day, however, since they are too unwieldy to be worn with shoes. Like all orthoses, the efficacy depends greatly on the level of compliance. In some cases we have found them to produce remarkably efficient effects. Treatment with splints is not adequate, however, for severe forms of hallux valgus.

### Surgical treatment

The following *operations* are commonly performed for varus of the 1st metatarsal in juvenile hallux valgus:

- base osteotomies of the 1st metatarsal,
- subcapital osteotomy of the 1st metatarsal,
- scarf osteotomy of the 1st metatarsal (»scarf« is a carpentry term used to describe a joint made by notching the ends of two pieces and fastening them together so that they overlap)
- opening wedge osteotomy of the medial cuneiform,
- transfer of the adductor hallucis muscle from the proximal phalanx to the 1st metatarsal (operation according to McBride [12]),

- combined subcapital, chevron osteotomy and varus (Akin) osteotomy of the proximal phalanx according to Mitchell [13, 15],
- arthrodesis with valgus correction in the joint between the 1st metatarsal and medial cuneiform according to Lapidus.

Since the main problem in juvenile hallux valgus is the deformity of the *1st metatarsal*, an *osteotomy* at this point is a »logical« operation. It should be noted that the joint between the medial cuneiform and 1st metatarsal also shows a pronounced slant. Moreover, since a growth plate exists in the proximal area of the 1st metatarsal, an osteotomy at this level is not possible in adolescents until growth is complete.

What is feasible, however, is an opening wedge *osteotomy of the medial cuneiform bone* that corrects both the slanting position of the joint and that of the 1st metatarsal. Although no epiphyseal plate needs to be considered at this level, the correction options are limited. A recently developed procedure is the combined V-shaped, or chevron, osteotomy of the 1st metatarsal with an osteotomy at the 1st proximal phalanx (Mitchell procedure or chevron Akin osteotomy) [13, 15, 16], or a double osteotomy of the 1st metatarsal at the base and the subcapital level [1, 14]. The scarf osteotomy has gained widespread acceptance in recent years [3, 11] and can likewise be combined with the Akin osteotomy on the 1st proximal phalanx (Fig. 3.421). The advantage of the scarf osteotomy is that the correction angle can be adjusted very precisely and that healing is promoted by the stable screw fixation of large bone areas.

In adolescents we combine this operation with a *soft tissue operation according to McBride* [12] (Fig. 3.421 and 3.422), in which the tendon of the adductor hallucis muscle at the base of the 1st proximal phalanx is detached, passed through a transosseous tunnel in the metatarsal head and fixed on the medial side. This procedure changes this muscle from an adductor of the great toe into an adductor of the 1st metatarsal, thereby producing a contrary effect. At the same time, the pseudoexostosis on the medial side of the head of the 1st metatarsal must also be resected.

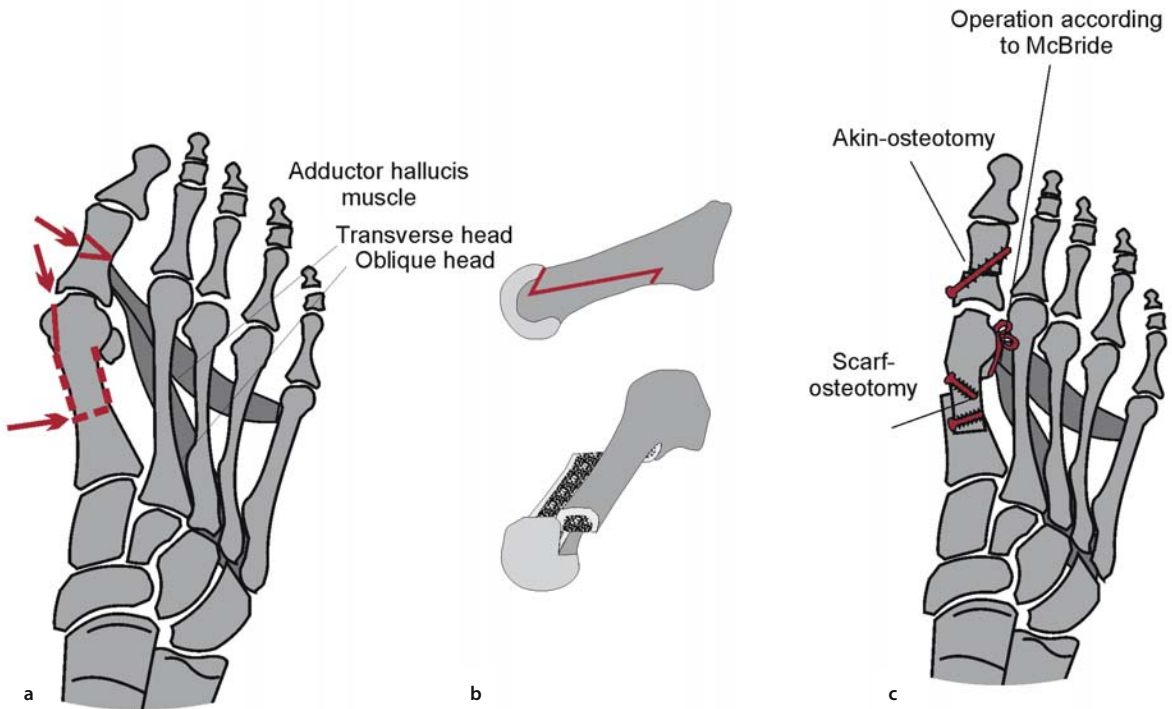
The arthrodesis according to Lapidus, with valgus correction in the joint between the 1st metatarsal and medial cuneiform, was also developed specifically for use in adolescents [6] and is based on the idea that the main problem is the hypermobility in this joint. Even though the results described in a comparative study by these authors were better than with other procedures, we remain very reserved about a method that involves the stiffening of a joint in adolescents. On the other hand, the scarf osteotomy and the Mitchell procedure are becoming increasingly popular [3, 11, 13, 14, 15]. There is no doubt that very impressive anatomical corrections can be achieved with these methods.

## 3.4.8 · Juvenile hallux valgus

Postoperatively, we apply an immobilizing cast for a few days until the swelling subsides. We then prescribe a hallux valgus shoe with a rigid sole. This allows the patient to walk without moving the toe joint during the heel-to-toe roll. After 3 months, a normal roll is again permitted.

We would specifically warn against overcorrection. A hallux varus is a much more handicapping deformity than a hallux valgus since the affected patients are always

stubbing their great toe against objects and find it difficult to slip their foot into a shoe. Moreover, it is difficult to recorrect a hallux varus by surgical means. Occasionally this complication ends in arthrodesis of the metatarsophalangeal joint. It should therefore be borne in mind that the great toe should be in a slight (physiological) valgus position at the end of the operation and not overcorrected in the varus direction.



■ Fig. 3.421a–c. Schematic view of the *surgical correction of juvenile hallux valgus*. The adductor hallucis muscle is detached from the base of the proximal phalanx and fixed to the head of the 1st metatarsal (operation according to McBride). In addition, the pseudoexostosis on the medial side of the head of the 1st metatarsal is resected, and a z-shaped

valgization osteotomy of the metatarsal is performed in the horizontal plane (*Scarf osteotomy*). If the base of the proximal phalanx is very slanting or the distal phalanx shows pronounced valgus deviation, an *Akin osteotomy* of the proximal phalanx is also performed. a Preoperatively, b lateral and oblique view of the scarf osteotomy, c postoperatively



■ Fig. 3.422a, b. DP and lateral x-rays of the left foot of a 15-year old girl with *juvenile hallux valgus*. a Preoperatively. b 1 year after surgical correction by the methods shown in Fig. 3.421

**!** The most serious *complication* is **hallux varus**, i.e. **overcorrection**.

Operations that are usually performed on adults, with resection of the base of the proximal phalanx or the metatarsal head, are never indicated for adolescents and are reserved for conditions in which substantial arthrosis already exists in the metatarsophalangeal joint.

**Our therapeutic strategy for juvenile hallux valgus**

Our therapeutic strategy for juvenile hallux valgus is shown in **■** Table 3.70.

**■** Table 3.70. Our therapeutic strategy for juvenile hallux valgus

During growth	Observe, possibly nocturnal splint
At or shortly before completion of growth	Symptomatic scarf osteotomy of the 1st metatarsal, possibly combined with Akin osteotomy of the proximal phalanx and McBride operation
A varus 1st metatarsal without hallux valgus does not require treatment	

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**3.4.9 Does one have to walk one's feet off before the cause of foot pain can be established? – or: Osteochondroses and other painful problems of the feet**

Foot pain in children and adolescents is not particularly rare. It is most commonly located in the heel area, slightly less frequently in the forefoot area, while the midfoot is affected only in exceptional cases. Severe *deformities* can cause pain as a result of the concentration of stresses at a non-physiological site (see also **►** chapters 3.4.3–3.4.5). Diffuse pain can be induced by *neuromuscular disorders* (**►** Chapter 3.4.10). Pes cavus is often painful, and the possibility of a neurological cause should always be considered if this foot shape is present. This chapter addresses the causes of pain in those feet that appear outwardly normal. The *differential diagnosis* of the aforementioned disorders should always take into account the possibility of a tumor (**►** Chapter 3.4.13) or an infection (**►** Chapter 3.4.12) in a case of severe pain on one side.

**!** Tarsal coalition is one of the most important and most frequently overlooked causes of foot pain in children and adolescents. Tarsal coalitions are described in **►** chapter 3.4.5.2.

**3.4.9.1 Köhler's bone disease (of the tarsal navicular)**

**►** Definition

Temporary circulatory disturbance with aseptic osteonecrosis of the tarsal navicular, usually occurring during early childhood.

— **Synonyms:** Aseptic bone necrosis, juvenile osteonecrosis

**Historical background**

Primary aseptic necrosis of the tarsal navicular was first described by Köhler in 1908, who subsequently reported on 26 cases in 1913 [9].

### 3.4.9 · Does one have to walk one's feet off before the cause of foot pain can be established?

#### Occurrence

This is a very rare disease that occurs 4 times more often in girls than in boys. It typically manifests itself between the ages of 3 and 8 years. Bilateral involvement is observed in 30% of cases.

#### Etiology

There is evidence to indicate that the disease develops as a result of repeated mechanical compression forces. The navicular is the last of the tarsal bones to ossify, the ossification center appearing between 18 and 24 months in girls and between 24 and 30 months in boys. Irregularities of ossification are common. A circulatory problem can occur as a result of the compression forces.

#### Clinical features, diagnosis

The affected children complain of load-related pain in the midfoot and over the back of the foot, and walk with a protective limp, rolling from heel-to-toe over the lateral edge of the foot. Swelling and tenderness may be present over the navicular. The x-ray shows condensation, and possibly fragmentation and flattening of the navicular (■ Fig. 3.423). The radiographic appearance can take several years to return to normal, and residual deformation of the bone may persist. The differential diagnosis should particularly consider tumors and inflammation and rule out the possibility of ossification disorders as normal variants.

#### Treatment

If the symptoms are severe, a below-knee cast may need to be fitted. This should be left in place for around 6 weeks. Accurately-shaped relieving inserts can also prove useful.



■ Fig. 3.423. Lateral x-ray of the foot of an 8-year old boy with osteonecrosis of the navicular (Köhler's disease)

Surgical measures are not appropriate. Two well-documented studies with observation periods of over 30 years have demonstrated the excellent long-term prognosis of this disease. While conservative measures can influence the symptoms, the result is equally good with or without treatment [3, 9].

#### 3.4.9.2 Freiberg's disease (infarction)

##### ► Definition

Temporary circulatory disturbance with aseptic bone necrosis of a bone in the forefoot, usually occurring during adolescence, typically affecting the heads of the 2nd or 3rd metatarsals, although the osteonecrosis can occur in almost any other bone of the foot.

■ **Synonyms:** Osteonecrosis of the metatarsal heads, Osteochondrosis of the metatarsal heads aseptic bone necrosis, juvenile osteonecrosis

##### Historical background

Osteonecrosis of the metatarsal heads was first mentioned in 1914 by Freiberg [5]. Köhler subsequently published a more detailed description of the disease [11]. In German-speaking countries it is commonly known as »Köhler II« disease, whereas the term »Freiberg's disease« is preferred in English-speaking countries.

##### Occurrence

This condition affects the 2nd–4th metatarsal heads, predominantly in girls between 10 and 18 years old, and typically occurs in combination with splayfoot. Not infrequently both feet are involved. The ratio of girls to boys is 3:1.

##### Etiology

Likewise in this disease, mechanical causes play a role in addition to genetic factors. During the development of splayfoot deformity, non-physiological stresses arise in the area of the metatarsal heads of the central rays.

##### Clinical features, diagnosis

Severe, load-related pain that hinders the heel-to-toe roll may be present in some cases, resulting in an unharmonious gait pattern or a protective limp. Clinical examination reveals tenderness in the area of the necrotic metatarsal head. Flattening and a cup-shaped deformation of the metatarsal head are observed on the x-ray as well as widening of the distal diaphysis. As is typical of all osteonecroses, however, such findings only occur after a certain time lag (■ Fig. 3.424).

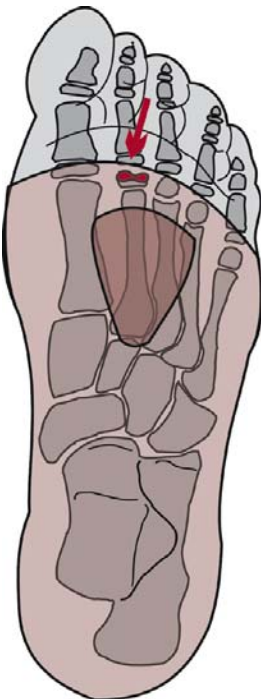
##### Treatment

A lower leg relieving cast is occasionally required in the florid stage. Inserts with retrocapital support are useful (■ Fig. 3.425), at least until metatarsalgia occurs at a later





■ Fig. 3.424. DP x-ray of the forefoot of a 13-year old boy with osteonecrosis of the 3rd metatarsal (Freiberg's disease)



■ Fig. 3.425. Principle of relieving the head of the 2nd metatarsal in Freiberg's disease by means of an insert with retrocapital support

stage because of incipient arthrosis. In such cases, head resection is occasionally required, although this is generally indicated only in advanced adulthood. A shortening osteotomy of the metatarsal may sometimes be useful [15]. A dorsal subcapital wedge resection has also produced successful results [4].

### 3.4.9.3 Other rare forms of osteonecrosis on the forefoot

Osteonecrosis is also observed, from time to time, on the head of the 1st metatarsal (■ Fig. 3.426). This rare phenomenon poses particular problems for treatment since the 1st ray is subjected to greater loads than the central rays. Osteonecrosis of the *sesamoid bone* is also difficult to treat.



■ Fig. 3.426. Oblique view of the forefoot of a 7-year old boy with osteonecrosis of the 1st metatarsal



### 3.4.9.4 Osteochondritis dissecans of the talus

#### Definition

Necrotic focus located on the lateral or medial edge of the trochlea of the talus surrounded by sclerotic bone which can then break off and remain in the joint as a loose body (»joint mouse«).

#### Occurrence

After the knee and elbow, the body of the talus is the third commonest site for osteochondritis dissecans (OD), i.e. around 4% of all cases of OD involve the talus [16].

#### Etiology, pathogenesis

Traumatic and genetic factors predominate. Trauma, in particular, appears to play a major role. Biomechanical studies have shown that the highest shear forces are produced in areas that are most commonly affected by OD,

i.e. on the lateral or medial edge of the talar trochlea [16]. An initial trauma can be followed by the development of a subchondral necrosis. Repetitive mechanical loading can deepen the lesion and lead to separation of this part from the surrounding bone. Finally, a discontinuity occurs in the cartilage surface and the loose body then has no connection with its surroundings. In contrast with the situation in the knee joint however, the loose body very rarely leaves its bed since the lesion is usually covered in all positions by the tibial joint surface and has no room to maneuver. Since, apart from a genetic disposition, traumatic factors are always involved, the former distinction suggesting that the traumatic form is usually located laterally, while the idiopathic form occurs medially and near the edge is no longer useful.

Another possible etiological factor which, to our knowledge, has not previously been mentioned in the literature, seems to be perfectly conceivable according to the case shown in Fig. 3.427. An x-ray of the ankle joint



Fig. 3.427a–d. Osteochondritis dissecans of the talus medially in a 12-year old boy. AP (a) and lateral x-ray (b) and CT sections (c, d) of the upper ankle. Since no symptoms had been present at any time this may be a separate ossification center rather than a secondary separation

was recorded in this patient following a recent episode of trivial trauma. The findings on the talus must have been present for a long time even though the patient had never reported any symptoms at all. This may involve a separate ossification center, resembling the situation in bipartite patella (► Chapter 3.3.3.5). The absence of any linking of separate ossification centers is also seen in talus partitus (► Chapter 3.4.5.1), although the separated ossification center is larger in the latter condition.

### Clinical features, diagnosis

Load- and movement-related pain in the upper ankle is the key symptom. The lesion usually occurs during adolescence and has only rarely been observed to date in children in isolated cases [8]. Plain x-rays usually show the lesion on the edge of the talus very clearly (■ Fig. 3.428). An MRI scan can provide important information about the status of the loose body [2]. Stage I lesions show an intact cartilage surface and contrast enhancement around the edges.

Typical features of stage II lesions include the accumulation of fluid around the fragment, discontinuity of the cartilage surface, separation of the fragment and possibly cyst formation. ► Chapter 3.3.4 provides further details of the stages on the basis of MRI findings. The previously recommended procedure of scintigraphy has been superseded by MRI and is no longer useful for this indication.

### Treatment

As with OD of the knee, no consensus exists on the optimal therapeutic course of action. A meta-analysis of 39 studies on this subject [17] has indicated that the prognosis is better after surgery than after conservative treatment, although most of these studies originate from orthopaedic hospitals. One study originating from a radiological department and involving the MRI follow-up

of 76 patients showed a higher remission rate for those treated conservatively compared to those undergoing surgery [1]. Adolescents usually have a better prognosis than adults. The initial findings certainly play an important role.

In stage I lesions we always try conservative treatment in the form of a below-knee cast for 6–12 weeks. We consider that surgical treatment is only indicated for MRI stage II lesions. Since the fragment has already separated by this stage, drilling of the lesion is no longer sufficient, and we perform an open procedure. The overlying malleolus usually has to be osteotomied in order to afford a complete view of the lesion. If possible we refix the loose body (ideally with resorbable screws, which do not need to be removed). If defect formation has occurred, various reconstructive procedures are possible: periosteal flap reconstruction, mosaicplasty [7], allogeneic bone graft (allograft) [6]. The current value of these methods is discussed in ► chapter 3.3.4.

### 3.4.9.5 Calcaneal apophysitis

Heel pain is common in children aged between 5 and 12, and the cause is almost always *calcaneal apophysitis*. It frequently occurs bilaterally and boys are more often affected than girls. The pain is located at the back of the heel over the apophysis. This is a protracted problem that can cause troublesome symptoms for years. The triggering factors are predominantly mechanical (sporting activity, obesity). No radiologically detectable osteonecrosis is present in the majority of cases. In fact, the radiological distinction between normal and pathological findings can be problematic. The calcaneal apophyseal center is visible on the x-ray from the age of 5 and synostoses at the age of 12. Initially, the ossification center appears denser on the x-ray than the body of the calcaneus. At a later stage the apophyseal center is often fragmented but without concomitant symptoms. Since there is no correspondence between the occurrence of pain and the fragmentation of the apophysis, the attempt to define pathological radiological findings by comparison with the other foot usually proves unfruitful. The only important differentiation is in relation to tumors or osteomyelitis (■ Fig. 3.429).

### Treatment

Treatment consists of relieving measures. The incorporation of a soft heel in the shoe can be useful. The patient should be advised to reduce his or her sporting activity. Occasionally, the wearing of a below-knee cast with heel relief for 4–6 weeks is required.

! Since the symptoms disappear at the latest on completion of growth, surgical treatment is not indicated, even in cases of persistent pain.



■ Fig. 3.428. AP and lateral x-rays of the upper ankle of a 12-year old boy with *osteochondritis dissecans of the talus medially*

### 3.4.9 · Does one have to walk one's feet off before the cause of foot pain can be established?



■ **Fig. 3.429.** *Calcaneal apophysitis* bilaterally on the lateral x-ray of the rearfoot of an 8-year old girl. The condensation and slight fragmentation of the apophysis on the right foot corresponds to a normal finding and is not pathological, although both heels were painful. The fragmentation of the apophysis on the left foot extends beyond the normal level and corresponds to an osteonecrosis



Heel pain should primarily be treated by conservative measures only...

#### 3.4.9.6 Other types of heel pain

Other causes of heel pain exist in addition to calcaneal apophysitis, although these are all rare in children and adolescents:

- Sever's disease,
- Haglund's deformity,
- posterolateral exostosis,
- insertion tendinosis of the plantar aponeurosis.

*Sever's disease* involves chronic pain in the hollow alongside the Achilles tendon that occurs particularly in athletic individuals who undertake intensive training. The cause is an insertion tendinosis of the Achilles tendon. Frequently, the pain is triggered by a less than ideal sequence of movements during sport, in which case a minor shoe modification (e.g. the insertion of a slightly valgizing wedge) can sometimes rectify the problem. Often the shoe must be optimized in a series of trials. We would warn against cortisone injections as these can lead to avascular necrosis of the Achilles tendon.

*Haglund's deformity* is a spur-like projection of the bone over the attachment of the Achilles tendon. This is extremely rare in adolescents. A more common condition is *posterolateral exostosis*, in which the bone projects laterally over the calcaneus slightly in front of the Achilles tendon attachment. Rather than a genuine exostosis, this is more of an anatomical variant, although it can lead to symptoms when normal shoes are worn. Widening the footwear is much more useful than surgical chiseling, since the subsequent scar is more irritating than the former »exostosis«. If the pain is located in the anterior part of the heel, then an *insertion tendinosis of the plantar*

*aponeurosis* is usually involved. This occurs particularly in connection with a pes cavus. A soft-bedded heel in the shoe is therapeutically effective.

#### 3.4.9.7 Fatigue fractures

Stress or fatigue fractures arise, particularly in young bones, as a result of repetitive bending loads [12, 14]. In a study of 369 stress fractures in the Finnish army, the metatarsals were the second commonest site, at 13%, after the tibia (► Chapter 3.3.3.7) [13]. The history is usually one of chronic, load-related pain in the forefoot. The symptoms occur principally in adolescents who are very active sports practitioners and can persist for months. Clinical examination reveals pronounced tenderness behind the 2nd or 3rd metatarsal head.

Other metatarsals are only very rarely affected at the subcapital level. Stress fractures can occur at the base of the 5th metatarsal, particularly if the foot is in an abnormal varus or supinated position. The *x-ray* shows thickening of the cortical bone and possibly central osteolysis as well. The fracture itself is not always visible and, if so, rarely as a typical as a fracture gap, but rather as a more or less diffuse osteolysis resulting from repair processes. A *bone scan* shows strong uptake.

The most important *differential diagnosis* is an osteoid osteoma (► Chapter 3.4.13, 4.5.2.1), which also involves cortical thickening and increased uptake on the bone scan. Moreover, the osteolysis of the stress fracture can easily be misinterpreted as a nidus of an osteoid osteoma. The most important distinguishing feature is the fact that the pain in a stress fracture is load-related,

whereas the osteoid osteoma typically causes nocturnal pain that responds well to aspirin. Treatment involves the exclusion of the causative stress, i.e. a temporary ban on the sport that is being practiced excessively. The best way to achieve this is by fitting a below-knee cast, which immobilizes the fracture and effectively stops the patient from practicing sport. A check x-ray is recorded after 4 weeks, by which time the fatigue fracture has usually healed.

**!** In a patient with foot pain one should always think of the possibility of a tumor. Tumors are described in ► chapter 3.4.13.

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### 3.4.10 Neurogenic disorders of the ankle and foot

R. Brunner

The foot represents the lever arm over which the triceps surae muscle applies its force to the ground. This biomechanical system is primarily responsible for controlling the upright posture: An active plantarflexing force causes the lower leg to adopt a backward lean and the knee to extend (»plantar flexion – knee extension couple«). The preconditions for the efficacy of this process:

- a stable lever arm,
- the lever arm must point in the direction of walking,
- the muscle must produce sufficient active force,
- the force should not be uncontrolled or excessive (spasticity).

Functional or structural foot deformities are very common in neurogenic disorders, and can produce widely differing consequences. It is particularly important to distinguish between functionally relevant and cosmetic impairments. Disruptive components of a deformity must also be differentiated from functionally useful ones so that the treatment can be matched to the individual patient's needs. It must be decided, for example, whether functional improvement is required for standing or walking, or whether the patient is prepared to wear braces such as inserts or orthoses.

A common component in neurogenic foot deformities is a functional or structural equinus foot. In a barefoot child with an equinus deformity, the weight-bearing area is small, which makes it more difficult to maintain an upright posture. In most cases, stabilizing aids such as inserts or orthoses are required in any case to control other components of the foot deformities. At the same time, such braces can, through corresponding bedding, help improve dynamic stability during standing and walking. In view of these considerations, the equinus foot position is not usually of any great significance as long as there is at least a heel-floor contact (by an equivalent heel rise). On the other hand:

**!** An equinus foot on its own rarely leads to an inability to walk, but a calcaneus foot is much more troublesome and requires compensatory postural work on the part of the knee and hip extensors (► Fig. 3.430). Caution is therefore required in deciding whether Achilles tendon lengthening is actually indicated.

Foot deformities do not have functional consequences if the feet are not subject to weight-bearing (although this is rarely the case since even severely disabled individuals are placed in standing frames). For small patients a splint may be indicated for cosmetic reasons so that the shape of the foot is preserved in the long term and normal shoes can be worn. For severe foot deformities a decision needs to be





■ Fig. 3.430a, b. Patient with *spastic tetraparesis*. a Fixed equinus feet. b The patient has been able to walk freely for many years wearing corresponding *orthoses in an equinus position*

made as to whether correction is required (which would include an extensive arthrodesis in the rearfoot) or whether the patient should refrain from wearing normal shoes.

❗ **The orthopaedist must be very cautious when deciding whether surgical treatment of the foot aimed at better orthosis provision is indicated. From the technical standpoint, almost any foot can now be secured and stabilized in an orthosis. If surgery is indicated, then it should be instead of an orthosis or at least reduce its complexity.**

If the abnormal foot position is based on the pathological activity of one or more muscles, this hyperactivity can be countered efficiently by the administration of botulinum toxin. Alternatively, their tendons can be lengthened in order to weaken these muscles.

In any case, a foot deformity should not be considered in isolation, but always together with the whole leg and the general abilities of the patient. Patients who are able to walk should therefore undergo a gait analysis preoperatively. For those who cannot walk, the functional restriction produced by the deformity must be clarified. The better the correction of the foot deformity in walking patients, the better will be the functional results.

### Primarily spastic paralyses

#### ➤ Definition

Functional changes in the foot with no structural deformity and caused by spastic muscle activity.

In cases of spastic paralysis, the activity of the triceps surae muscle is a crucial factor in the development of foot deformities. During walking, the hyperactivity of this

muscle leads to clonic contractions that draw the foot into an equinus position, whereas in the absence of a genuine contracture, the heel drops to the floor on prolonged standing. However, the persistent clonic activity of this muscle during walking leads to overstretching of the antagonists, i.e. the dorsiflexors of the foot. As a result, even if the latter muscles are correctly innervated, they become overlong and functionally inefficient over time or appear inactive, producing the combination of a foot dorsiflexor paresis (footdrop) and a functional equine foot [4]. This initially functional situation eventually develops into a structural equine foot with contracture of the triceps surae muscle. The control of the foot muscles required in this position is insufficient, leading to the development of additional deformities of the foot itself and the toes.

### Functional disorders

An overview of the functional problems in primarily spastic locomotor disorders is shown in ■ Table 3.71.

#### Footdrop

##### ➤ Definition

The foot remains in plantar flexion in the absence of dorsiflexor activity and therefore strikes the ground in an equinus position.

In patients with spastic forms of paralysis, the force exerted by some muscle groups can be weakened. The dorsiflexors of the foot are often affected. Although they can be activated voluntarily, in most automated movements, such as walking, the central command is not issued, functionally resulting in footdrop. By way of compensation, increased knee flexion is required during the swing phase to prevent the plantarflexed foot from dragging on the ground. On foot strike the toes contact the ground first. The initial plantarflexion, the first rocker of foot kinematics, in stance, does not occur. Toe strike is followed by a retrograde rocking of the foot from toe to heel (second rocker), followed by the normal forward heel-to-toe roll (third rocker). One would therefore expect the functional effects of a pure case of footdrop to be slight. However, since there is an underlying spastic condition and the triceps surae is also affected in most cases of spasticity, the retrograde toe-to-heel roll of the foot tenses the Achilles tendon and the triceps surae, which then undergoes spastic contraction and draws the foot into a functional equinus position (see below).

#### Functional equinus foot position

##### ➤ Definition

An equinus foot position is present during functions such as walking and/or standing, but neither a structural equinus foot nor a contracture of the triceps surae is observed on clinical examination at rest.



■ Table 3.71. Functional problems in primarily spastic locomotor disorders

Deformity	Functional benefit	Functional drawbacks	Treatment
Footdrop	–	Equinus gait due to Achilles tendon reflex Hindrance during swing phase	Functional orthosis (muscle transfer)
Functional equinus foot	Indirect knee stabilization/extension (slight equinus foot)	Instability due to reduced weight-bearing area Crouch gait	Functional orthosis Cast correction Lengthening of the triceps surae muscle
Functional clubfoot	–	Unstable stance Hindrance during swing phase	Functional orthosis Lengthening/transfer of the tibial muscles
Functional abducted pes planovalgus	Compensates for increased internal rotation of the leg	Walking/standing aggravated Risk of dislocation in the tarsal bones (pain)	Functional orthosis Cast correction Lengthening of the triceps surae and/or peroneal muscles Calcaneal lengthening Arthrorisis Arthrodesis

Before therapeutic measures are initiated, the functional equinus foot must be differentiated from an equinus gait based on inadequate knee extension at the end of the swing phase, and also from hyperesthesia or pain in the foot area that causes the patient to switch actively to an equinus foot position.

A functional equinus foot must be differentiated from a structural form. While an equinus foot position occurs during walking in both cases, during the examination at rest of a patient with the functional form the foot can be placed in a right-angled position or even in dorsiflexion, at least when the knee is flexed. If this maneuver is performed rapidly, however, a clonus of the triceps surae muscle that prevents dorsiflexion is elicited. The functional equinus foot results from a spastic activity of the triceps surae even though this muscle is not structurally contracted. As with a structural case of equinus foot, an equinus foot position involves only a small weight-bearing area, and both standing and walking are difficult in this position even in a patient with normal, unrestricted body control and balancing reactions. If the control of muscular function is restricted (as in patients with neurological disorders), balancing on tiptoe will be difficult or even impossible. This explains the frequent development of an abduction flat valgus foot or clubfoot (see corresponding section).

The simplest *conservative* treatment for a functional equinus foot position is a functional orthosis. The ankle foot orthosis guides and stabilizes the foot, preventing the equinus foot position and reducing the amount of energy used during walking [7]. Wearing an orthosis also improves walking without the orthosis because deformities of the foot skeleton are prevented and muscle length is preserved. If the patient does not have a stiff form of

spasticity but merely shows clonic activity of the triceps surae, a dynamic lower leg orthosis can be used, otherwise a *rigid orthosis* will be needed. Gait function is better with a mobile orthosis. A short orthosis shaft is functionally equivalent to a mobile orthosis but involves a loss of the additional control. The orthosis shaft must extend up to the knee if the position in the upper ankle is to be controlled (as with an equinus foot) [9].

The functional equinus foot also constitutes the ideal indication for the administration of botulinum toxin [9, 11]. If it is injected on one side the soleus muscle can be included as the patient can use the muscles on the other side for postural control. If the botulinum is injected on both sides, however, the soleus muscle should be spared, otherwise the knee may give way in the absence of triceps function on both sides. An increased varus position is usually present in spastic equinus feet. In such cases, the botulinum toxin is also injected into the tibialis posterior muscle. Botulinum can also be injected to check the functional therapeutic concept before a surgical procedure.

As a *surgical* alternative option, the *lengthening of the triceps surae muscle* is often performed. This tendon lengthening procedure results in a reduction of the stretch reflex on foot strike and a diminution in muscle power, which manifests itself as a reduction in the Achilles tendon reflex. In other words, the spasticity is treated by muscle weakening. While heel contact during walking is achieved as a result of this procedure, the concomitant footdrop is not corrected. The toe walking is changed to a toe-heel-ball gait, but a physiological heel-ball gait is not achieved.

Surgical lengthening procedures (primarily of the Achilles tendon) are much more hazardous in respect of functional insufficiency during walking in cases of purely functional equinus foot than in cases of a contracted

muscle. At best, the loss of power in the triceps surae only hinders the foot push-off during walking. Overcorrections of equinus foot after (Achilles) tendon lengthening procedures are not infrequent and result in a pes calcaneus position and thus a functional deterioration. The consequence can be secondary contractures of the knee and hip flexors.

Whereas an equinus foot can be managed with a functional spring orthosis, which facilitates an almost normal gait, a rigid orthosis, which hinders walking, is required for pes calcaneus. Gait function with the latter is therefore much worse than with an equinus foot. A safer method than tendon lengthening is the aponeurotic lengthening of the triceps surae according to Baumann [1]. Although this procedure does not lengthen the muscle as much, it also involves less of a loss of muscle power [3]. If dorsiflexor activity fails to recover, additional measures will need to be considered: A conservative option is a foot-lifting orthosis. A surgical alternative is to tension the dorsiflexors or transfer the muscles to compensate for the deficit.

### Functional clubfoot position

#### ➤ Definition

Supination, forefoot adduction, varus of the calcaneus and an equinus position are present in varying degrees during functional use of the foot. At rest and without weight-bearing the shape of the foot is normal.

Particularly with hemiparesis, and slightly less frequently with diplegia or tetraparesis, the spasticity of the tibial muscles draws the foot into a clubfoot position (■ Fig. 3.431). The fixation of the deformity must be prevented by bringing the foot into an anatomically correct position, otherwise a progressive clubfoot deformity will result in a deterioration in walking ability. The foot is supinated during the swing phase and strikes the ground in this position, resulting in instability in the stance phase.

Here, too an orthosis is available as a *conservative* therapeutic option. Since, in addition to the clubfoot position, an equinus foot is also usually present at the same time, both deformities can be corrected with the one orthosis.

The increased supination of the foot is generally based on tibialis posterior muscle hyperfunction. Provided no bone deformities have developed, the injection of botulinum toxin is an elegant method of inactivating this overactive muscle. Since this muscle is located deep in the tissues and is relatively thin, we prefer to perform this injection under ultrasound control. If the tibialis anterior is also producing a deforming effect then this muscle can be included in the injection treatment.

*Surgical* treatment is often useful in walking patients, and may also be indicated in those who cannot walk in order to alleviate pain, resolve major problems or possibly to simplify the provision of footwear. The lengthening of all contracted muscles is a simple surgical option. Since, as



■ Fig. 3.431. Foot of a female patient with *spastic hemiparesis*. Clearly visible through the skin is the tendon of the *spastic anterior tibial muscle*, which draws the foot into a clubfoot position

with all muscle lengthening procedures, this involves the risk of recurrences, consistent follow-up orthotic management is required. The foot position can also be corrected with a muscle transfer with the aim of balancing the muscle forces. If the spasticity is strongest in the tibialis anterior, the best option is to transfer the whole muscle distally to the middle of the back of the foot or just laterally of this position. The tendon is long enough to be pulled through a bone tunnel and then sutured back on itself. However, the tibialis posterior should not be transferred completely, otherwise there will be a risk of overcorrection.

A more suitable procedure is the split transfer: The tendon is exposed at its distal end and divided, and the lateral half is pulled laterally behind the tibia and sutured to the peroneus brevis tendon. A problem arises, however, if the muscle has already become contracted by this stage: In this case, one half of the tendon would need to be lengthened and the other half transferred. But since the tendon of the tibialis posterior muscle is usually too thin for this procedure, we only perform the tendon lengthening with the sliding technique. While good results are reported in the literature for both muscle transfers, our own experience has dampened our expectations of this operation. Nevertheless we have also used the tendon transfers in patients with structural foot deformities. Better results

can be expected if the foot skeleton is still anatomically unchanged, although this condition is only satisfied in early childhood in many patients.

If bone deformities already exist, an improvement, but not a full correction, can be achieved with purely soft tissue procedures.

### Functional abducted pes planovalgus position

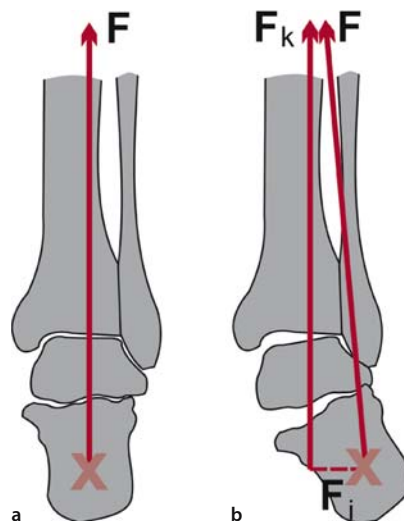
#### ➤ Definition

The foot gives way under load as a result of the absent, or insufficient, activity of the muscles that stabilize the foot. The arches are flattened out, the heel is in a valgus position and the forefoot is abducted. The skeleton is normal when the foot is not weight-bearing.

This foot position progressively leads to subluxation and eventually to dislocation between the talus and navicular or between the navicular and the distal tarsal bones, as well as subluxation in the lower ankle. As the valgus position of the calcaneus progresses, the 5th ray is pulled laterally, in turn pulling the 1st ray as well. The triceps surae muscle becomes contracted since the deviation of the calcaneus into the valgus position brings the origin and insertion of the triceps surae muscle closer together. The contracture of the triceps surae muscle leads, in turn, to a progressive valgus position, since the muscle is evidently not as extensible as the joint capsules in the tarsal bones and because the direction of traction of the muscle with a valgus calcaneus runs laterally relative to the center of the upper and lower ankle (■ Fig. 3.432).

Moreover, as the foot of a patient with abducted pes planovalgus rolls over on its medial aspect it is pushed away laterally with each step, thereby exacerbating the overstretching of the medial capsular ligament structures. As a result, although the whole foot (including the heel) may strike the ground, the triceps surae muscle can, in severe abducted pes planovalgus, be greatly contracted. If the plantarflexed foot is placed in maximum supination and adduction (thereby reducing the tarsal bones) and then dorsiflexed, the extent of the shortening of the triceps surae becomes clear (■ Fig. 3.433). If this deviant position of the foot is present permanently or for most of the time, the foot skeleton will grow into this abnormal shape (with shortening of the 5th ray and an excessively lengthened 1st ray). The subluxation or dislocation in the rearfoot will become fixed. In extreme cases the patient will be standing on the talus and the foot folds out sideways.

In functional terms the foot deformity leads to a loss of stability. The triceps surae loses its lever arm, partly as a result of the flattening of the foot arch and partly as a result of the outward rotation. This loss of stability leads to a functional pes calcaneus position with increased flexion of the knees. At the same time, the foot skeleton twists as the talus slides down off the calcaneus in a medial



■ Fig. 3.432a, b. Schematic view of the direction of traction exerted by the Achilles tendon in the anatomical position (a) and with subluxation in the lower ankle (b).  $F$  Direction of the Achilles tendon which, in the case of subluxation in the lower ankle, exerts a force in line with the lower leg  $F_k$  and a dislocated force component  $F_i$ .

direction, resulting in an internal rotation of the leg. This internal rotation combined with a flexed knee simulates a pronounced valgus position.

The aim of *conservative treatment*, on the one hand, is to preserve the muscle length relationships and prevent abnormal growth of the skeleton and, on the other, to restore the biomechanically important lever arm of the foot. To this end, the foot skeleton must be held in an anatomically correct position in an orthosis and realigned so that it points in the direction of walking. If the patient's body control is good enough to benefit from mobility, a spring orthosis may be used as this is less irksome. Otherwise a rigid orthosis is indicated.

If there is minimal or no spasticity and adequate dorsiflexion in the upper ankle, a good insert in a shoe with medial and lateral stabilization will suffice. This insert must incorporate a medial, plateau-shaped support whose dorsal section rests under the medial part of the calcaneus so that the calcaneus is placed in a sufficiently varus position that it remains under the talus even during weight-bearing. This reduction of the calcaneus will also correct the abnormal posture of the forefoot, the flattening of the longitudinal arch and the abduction. The correctly functioning insert will also improve the foot position, compared to the barefoot position, as regards the direction of walking. Instead of shoe adaptation, we have started employing foot orthoses consisting of an insert and stabilizer, and which cover the hindfoot up to the malleoli, as these provide greater stability and fit in normal shoes. If a slight contracture of the triceps surae muscle is also present, it can be lengthened by means of a spring orthosis



■ Fig. 3.433a, b. Extreme *abducted pes planovalgus* with subluxation in the lower ankle. The patient is able to stand in a plantigrade position on the ground, but only on the talus (a). Same foot in maximum dorsiflexion after reduction in the lower ankle (b). The contracture of the triceps surae muscle of approx. 50° is apparent

alone. To this end, the orthosis is adapted according to the existing muscle contracture and the equinus foot position is progressively reduced over several months. At the same time all the other muscles can gradually adapt themselves to the new length relationships and provide the ideal pre-conditions for a functionally good result.

A faster way of stretching the triceps surae muscle is by corrective casts (■ Fig. 3.434), although this method regularly leads to a muscle atrophy of varying severity. Load-related pain can occur after removal of the cast, possibly partly as a result of centrally induced hyperesthesia (► Chapter 4.7). However, since the length adaptation of the antagonists takes longer than the stretching of the triceps surae, the cast treatment must be followed by the fitting of an orthosis, since recurrences can develop very quickly. Alternatively, the cast correction can be repeated at short intervals (every 4 to 6 months). Unfortunately there is a high risk for relapses while muscle strength recovers.

The stretching treatment can be facilitated by the injection of botulinum toxin, in which case the corrective cast is applied when the toxin starts to take effect after 10–14 days.

The *surgical treatment* must, on the one hand, take account of the length relationships of the triceps surae muscle, and thus include an extension, and, on the other, restore anatomically correct skeletal conditions where necessary. Because it produced unsatisfactory results, we no longer perform the procedure of arthrorisis (stabilization in the tarsal sinus with dowel implants or a bone graft), which is designed to preserve mobility in all joints. In order to retighten and realign the leaf-spring-like arch of the foot, the calcaneus can be lengthened (operation according to Evans). This is the best way of restoring the anatomical relationships without sacrificing any joints.



■ Fig. 3.434. Corrective casts are an efficient means of correcting contractures. Left the first fitted cast, which is left in place for 2 weeks, right the following cast, which was only fitted as a temporary measure until the orthosis was ready

But even this procedure loses its efficacy when the deformity becomes less flexible and then irreducible. In older children a calcaneo-cuboidal arthrodesis is helpful to correct the hypermobility of this joint. In these cases the bone block is inserted into the arthrodesis, producing an effect similar to that of the Evan's procedure.

Hyperfunctioning of the peroneal muscle group can also lead to an abducted pes planovalgus. Conservative treatment involves physical therapy with stretching exercises and orthoses or weakening with botulinum toxin. The surgical treatment of choice is lengthening of the relevant peroneal muscle (particularly the peroneus tertius).

### Structural deformities

Structural deformities in primarily spastic locomotor disorders are shown in ■ Table 3.72.

#### ► Definition

Structural deformity of the foot caused by spastic muscle activity.

#### Structurally fixed equinus foot

#### ► Definition

The cause is structural shortening (contracture) of the triceps surae muscle. In structural equinus foot, dorsiflexion to the neutral position is not possible, even if the triceps surae is inactive. Bony obstacles are not present however.

As with the functional form, the weight-bearing area is reduced in structural equinus foot, resulting in dynamic instability. In contrast with a purely functional equinus foot, the foot drops during weight-bearing while standing, but not onto the heel and without any additional deformation in the form of an abducted pes planovalgus or a clubfoot.

As a *conservative* measure, the triceps surae muscle can be stretched by cast correction, although this treatment regularly results in muscle atrophy, and load-related pain occurs after its completion. It must likewise be followed by orthotic management. In fact, orthotic treatment over several months on its own can also prove successful (see above: »Functional equinus foot position«) and more effective in maintaining the balance between agonists and antagonists.

A *surgical* option is the *Achilles tendon lengthening* procedure in which the tendinous portion is lengthened

while the shortened muscle belly is left as is. Consequently, the muscle loses less power. Adjusting the lengthened tendon to achieve the optimal tension is important for the subsequent functioning of the muscle. The aim is to keep the length range over which the muscle can produce its power within the range of motion of the joint (► Chapter 4.7). The procedure can either be performed openly with Z-plasty lengthening or percutaneously by an incomplete transverse incision of the Achilles tendon at various levels. As the dorsiflexors have been stretched out for a long time in these feet, a dropfoot may result. To overcome this dorsiflexor weakness we have started to add a shortening of the tibialis anterior at its distal insertion, with favorable initial results.

Equinus foot overcorrections are not infrequently observed after tendon lengthening procedures. These can result in muscle insufficiency with a pes calcaneus position that ultimately causes the patient to end up with poorer function than with the equinus foot position. In the *operation according to Strayer* the efficiency of the gastrocnemius muscles is weakened by proximalization of the distal insertion of the heads on the Achilles tendon. Since the involvement of these muscles in the development of an equinus foot is greater than that of the soleus muscle, this operation produces a functionally positive result and overcorrections are rare.

However, it can also result in a functionally relevant loss of power as a consequence of the partial failure of the gastrocnemius muscles. The intramuscular division of the aponeurosis can stretch the muscle belly and thus lengthen its tendon, which was not shortened in the first

■ Table 3.72. Structural deformities in primarily spastic locomotor disorders

Deformity	Functional benefit	Functional drawbacks	Treatment
Equinus foot	(Knee extension)	Dynamic instability due to small weight-bearing area Deformation of the feet	Functional orthosis (in equinus foot) Cast correction Lengthening
Clubfoot	–	Dynamic instability in the stance phase Skin problems	Functional orthosis Calcaneal osteotomy (Dwyer) Cuneiform/cuboid osteotomy Arthrodesis
Abducted pes planovalgus	Compensates for increased internal rotation of the leg	Dislocation in the tarsal bones Hyperactivity of the peroneal muscles Instability of leg in stance	Functional orthosis Cast correction Arthrodesis Orthoses, cast correction Surgical lengthening of lateral column of foot
Pes cavus	–	Overloading due to stiffness	Padded insert Release of the plantar fascia Corrective osteotomy



place [1]. The operation can be performed on the gastrocnemius muscles, the soleus muscle or at both sites. Intensive postoperative stretching by physical therapy is essential for a successful result. Although the effect of this procedure is usually inferior to that of the tendon lengthening procedure, overcorrections are very rare and muscle power is largely preserved [3, 10]. While the risk of recurrence is high, the operation can be repeated if necessary.

The triceps surae muscle can also be lengthened by means of an external fixator (Ilizarov-type apparatus) that incorporates the lower leg and calcaneus with forefoot, i.e. extending across the joint. The procedure is time-consuming and mentally stressful but, on the other hand, does produce good correction of the length relationships without scar formation. Here too, the risk of recurrence is high. This method is only recommended for previously operated feet and feet with complex deformities.

! **All lengthening measures, both conservative and surgical, are associated with a high risk of recurrence, particularly during growth. They can however be repeated.**

### Structural clubfoot

#### Definition

Clubfoot based on defective muscle function as a result of the underlying neurological disorder. The typical components, e.g. varus position of the heel, contracture of the triceps surae muscle, pes cavus and supination and adduction in the forefoot, are present in varying degrees at rest and skeletally fixed.

The spastic activity, i.e. usually of the tibial muscles and the triceps surae, initially draw the foot into the abnormal position during standing and walking. Weight-bearing produces an additional deforming effect on the foot skeleton. Finally, what was initially a functional deformity becomes a fixed bone deformity.

As a *conservative* measure, a clubfoot can be treated with a functional orthosis that prevents supination, takes account of the contracture of the triceps surae and stabilizes the foot. This is a functionally efficient option. In principle, almost any type of clubfoot can be managed with an orthosis. If the muscle contractures are severe enough to make the placement of the foot in a sufficiently correct position impossible, cast correction can remedy the situation and facilitate the orthotic management (■ Fig. 3.434). Serious cases of clubfoot are problematic however, since they can lead to excessive stresses on the lateral edge of the foot with the risk of pressure ulcers. The efficiency of stretching exercises can be enhanced by the injection of botulinum toxin into the contracted muscles.

Surgical correction may be required in some cases. The *Dwyer* or *Mitchell* osteotomy of the calcaneus is a simple procedure for correcting the varus position of the

heels (► Chapter 3.4.3). The foot is then immobilized for 2 weeks in a non-weight-bearing cast and then for 4 weeks in a below-knee cast. An opening-wedge *cuneiform* and subsequent *cuboid osteotomy* is an appropriate procedure for correcting the adduction position (► Chapter 3.4.3).

Correction with the Ilizarov external fixator has proved effective for severe deformities that have been present for a prolonged period. When a position of slight overcorrection has been reached, the fixator is removed and the corresponding corrective osteotomies performed. This procedure is easier and more efficient after the soft tissues have been prestretched with the fixator. If no osteotomy is performed, the abnormal position will recur within a short period. Severe cases of clubfoot, on the other hand, require a corrective *arthrodesis* of various joints in order to place the foot in a plantigrade position. Since such patients had previously been reliant, usually permanently, on a rigid, functional orthosis for walking and standing, and have therefore become accustomed to rigid foot joints, they suffer no functional deficit as a result of this procedure. Quite the opposite in fact, as they are placed to be free of the orthosis after the operation. The ankle joint however should be spared.

### Structural abducted pes planovalgus

#### Definition

A foot deformity with a valgus calcaneus, flattening of the medial longitudinal arch and forefoot abduction that develops as a result of the underlying neurological disorder. If the foot remains in this position permanently or for most of the time, this abducted pes planovalgus becomes skeletally fixed.

The skeletally fixed form of abducted pes planovalgus can be treated *conservatively* in the same way as the functional form (see above: »Functional disorders«) with orthoses.

Severe cases of abducted pes planovalgus (■ Fig. 3.435), however, require *surgical* stabilization and correction, for which an arthrodesis is the most appropriate procedure. At the same time, the arthrodesis dispenses with the



■ Fig. 3.435. Structurally fixed extreme abducted pes planovalgus with pes cavus component

need for subsequent orthoses. On the other hand, such operations do involve the problem of osteoporosis. Although the skeletal configuration can be fixed in the ideal position at operation after correction, the osteosynthesis material gradually loosens during the healing period, and even a cast cannot preserve this position sufficiently. As a result, a deterioration in the foot position often occurs during the healing phase, although this does not usually have any functional significance. While soft tissue lengthening procedures can place the foot in a better position temporarily, an orthosis will again be required after surgery and recurrences are common. However, if an arthrodesis is not possible, or if such an operation is rejected, this simple measure can place the foot in a better position, at least for a certain period of time.

Severe cases of pes planovalgus can be protected against subluxation laterally with an extra-articular arthrodesis (according to Grice) [5]. But even the extra-articular arthrodesis of the lower ankle may require a triple arthrodesis at a later date to correct deformity and instability. When skeletal growth is complete, the subtalar arthrodesis produces the best results for fixed deformities. A patient who has become accustomed to orthoses does not lose any function as a result of this procedure. A corrective pan-talar arthrodesis may be appropriate for severe subluxations or dislocations of the tarsal bones in patients who are only able to stand. Since the loss of mobility makes no difference in these patients, the arthrodesis can replace splint treatment.

### Structural pes cavus

#### ► Definition

The abnormal muscle activity resulting from the underlying neurological disorders draws the foot into a deformed position with an excessively pronounced longitudinal arch, in particular, which then becomes skeletally fixed. The extent of the transverse arch varies. In functional respects, dynamic and static overload occur as a result of the excessive foot stiffness.

The pes cavus develops as a result of hyperactivity of the peroneus longus and tibialis anterior muscles in relation to their antagonists. The accentuation of the arch reduces the cushioning function of the foot. A self-supporting arch (similar to a Roman archway) that no longer permits cushioning develops from the longitudinal foot arch. As a result, the heel and the ball of the foot are overloaded particularly during walking, leading to increased callusing and subsequently to local painful conditions. Symptoms are rare, therefore, for as long as the pes cavus remains mobile.

Inserts should not be used as *conservative* treatment while the foot still shows some residual mobility and if not required for the management of callusing. Without inserts, the foot is functionally stretched and flexed during walking and thus remains mobile. Inserts, on the

other hand, support the arch, preventing it from wanted sinking and thus increasing the risk of premature stiffening. If excessive callus forms on the overloaded areas of the sole, a soft *insert* with shock-absorbing pads beneath the pathological calluses will provide symptomatic relief. Shoes with soft soles and heels are beneficial. As a further measure it may be useful to mobilize the foot several times a day in order to preserve mobility.

As *surgical* treatment, a *fasciotomy of the plantar aponeuroses according to Steindler* can restore some flexibility to the foot, provided the bones of the foot are not yet wedge-shaped.

If the foot is fixed, the *conservative* measure of padding the whole sole with shock-absorbing pads can alleviate the symptoms of excessive loading. The only remaining surgical options are a *corrective osteotomy* in the area of maximum arching, with excision of a wedge (producing a shorter stiff foot with a poor roll function) or an angular correction by lengthening with the *Ilizarov fixator* (producing a longer stiff foot with a poor roll function, but a better cosmetic appearance). In functional respects, the shorter foot is better. Shoe rockers will then be required in such cases.

### Primarily flaccid paralyses

The main problem in flaccid paralyses and myopathies is the loss of power in the muscles required for walking and standing. Functional orthoses that replace the missing muscle power are therefore required. These orthoses must be worn whenever muscle activity is required, i.e. usually throughout life for standing and walking.

### Functional disorders

Functional deformities in primarily flaccid locomotor disorders are shown in ■ Table 3.73.

#### ► Definition

Changes in the functions of a foot without a structural deformity and caused by reduced or absent muscle activity.

### Pes calcaneus position

#### ► Definition

The triceps surae muscle controls the forward motion of the lower leg in relation to the foot on the ground during standing and brakes this motion in the stance phase during walking. The pes calcaneus results from a weakness of this muscle. The main load is transferred to the heel and the lower leg stands in a forward lean. By way of compensation, the knees and hip must be held in flexion to ensure that the whole sole strikes the ground, otherwise the knee will be actively straightened by the knee extensors and all the weight on the foot will be borne by the heel.

The triceps surae muscle normally acts to stabilize the ankle and thereby prevent the lower leg from sinking

■ **Table 3.73. Functional deformities in primarily flaccid locomotor disorders**

Deformity	Functional benefit	Functional drawbacks	Treatment
<b>Pes calcaneus position</b>	–	Crouch gait	Functional orthosis (muscle transfer) Arthrodesis
<b>Functional abducted pes planovalgus</b>	Compensates for increased internal rotation of the leg	Walking/standing aggravated Risk of dislocation in the tarsal bones (pain)	Functional orthosis Cast correction Lengthening of the triceps surae and/or peroneal muscles Lengthening of lateral column of foot Arthrodesis

forward during standing and walking. This muscle can suffer a loss of power during a paresis or myopathy (the same symptoms are also observed after excessive tendon lengthening). The affected patients have to stand on their heels only to offset this weakness. But since the resulting ground contact area is too small and the stability inadequate, some patients will sink forward with the foot and lower leg, causing them to stand and walk in a crouch position, i.e. with flexed knees and hips. This position requires more power and energy for walking and standing, and the flexed positions of the joints become fixed as the muscles contract because they are no longer extended to their full extent. The ability to walk and stand is then jeopardized with increasing age, weight and height.

A pes calcaneus must therefore be managed *conservatively* with a *functional orthosis*. The orthosis must be of a rigid design since it has to replace the absent muscle activity. During walking, the orthosis prevents the premature forward movement of the tibia in relation to the foot in contact with the ground and ensures adequate knee extension [6]. For growing children there is no alternative to an orthosis, since an arthrodesis will inhibit foot growth and leave the feet smaller than normal. Only on completion of growth can the orthosis be replaced *surgically* with an *arthrodesis*, which must incorporate the upper and lower ankle. Due to a lack of mobility, and hence of compensatory movements, pressure ulcers may occur and are difficult to treat. Maintaining mobility is therefore favorable in functional feet, especially if sensation is not normal. A muscle transfer procedure to replace the absent plantar flexion may be tried, specifically by *transferring tibial muscles* to the calcaneus. Although good results have been described, our everyday experience with our patients has shown that the transferred muscle is often unable to exert the necessary force to serve as an efficient replacement for the triceps surae.

The shortening of the Achilles tendon represents a logical alternative. However, this procedure is reputed to produce poor results. Although it can prove helpful in extreme cases, the chances of a good result in neuro-

thopaedic patients are slim. A precondition appears to be adequately free plantar flexion, which is no longer present in most cases.

### Functional abducted pes planovalgus

#### ➤ Definition

The foot gives way under load as a result of the absent, or insufficient, activity of the muscles that stabilize the foot. The arches are flattened out, the heel is in a valgus position and the forefoot is abducted. The skeleton is normal when the foot is not weight-bearing.

For the functional form of abducted pes planovalgus in muscle weakness due to a paresis or myopathy, the same therapeutic ideas and principles associated with primarily spastic paralysis apply (see above). The necessary stability can be achieved only by means of an external appliance (orthosis) or a surgical procedure. In many cases an *arthrodesis of the lower ankles* (usually an extra-articular Grice operation) is performed to stop the foot from going over. However, this procedure can only rectify the deviating valgus component of the foot. An excessive dorsiflexion, as also observed in insufficiency of the triceps surae, remains, and this is much more disruptive from the functional standpoint. Since an orthosis will still be required the benefit for the patient from a procedure such as the Grice arthrodesis is minimal.

### Structural deformities

Structural deformities in primarily flaccid locomotor disorders and muscular dystrophies are shown in ■ Table 3.74.

#### ➤ Definition

Structural deformity of the foot caused by reduced or absent muscle activity.

### Structural equinus foot

#### ➤ Definition

A contracture of the triceps surae muscle is present, regardless of the muscle activity and power, which prevents dorsiflexion even with a flexed knee.

In flaccid paralyses, a pronounced equinus foot makes it difficult for the body to keep in balance over the flaccid leg. Usually the foot gives way and drops into an abducted pes planovalgus or clubfoot position because the dynamic stabilizers that would have to keep the foot on tiptoe are also insufficient. The foot skeleton becomes deformed and fixes what was initially just a functional deformity. The ability to walk and stand can be further impaired as a result.

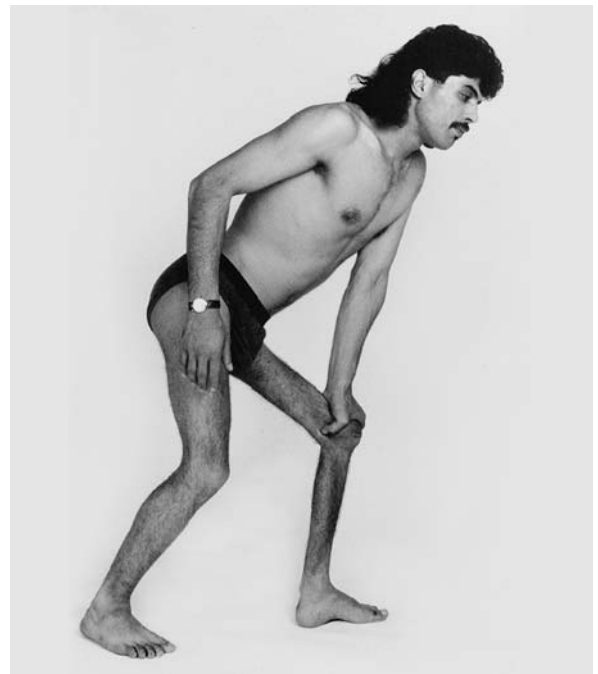
As a *conservative* measure, a *functional orthosis*, adapted to an equinus foot position, stabilizes the foot and permits weight-bearing without deformation of the foot skeleton. If a functionally disruptive contracture is present, i.e. if the condition is extreme, the *triceps surae muscle may be lengthened surgically*, but only sufficiently enough to produce the neutral position. An overcorrection will lead to a pes calcaneus position with corresponding flexion at the knees and hips, thereby compromising walking and standing. If the knee and hip extensors are not available for compensation (as in muscular dystrophies), a slight overcorrection will result in the loss of the ability to walk and stand. Since the lengthening procedure does not need to take account of the muscle power, it can be implemented in the form of tendon lengthening. One surgical technique for correcting the equinus foot in flaccid paralyses is the *rearfoot arthrodesis according to Lambrinudi* (■ Fig. 3.436). This procedure is risky to the extent that dorsiflexion is not blocked at the ankle. If the knee and hip extensor muscles are not strong enough to compensate for the lack of power in the triceps surae, a crouch gait will result. Dorsiflexion should hence be set at  $-5^{\circ}$  to  $0^{\circ}$  during surgery.

❗ **The equinus foot is an important aid to stabilization during standing and walking, particularly in muscular dystrophy patients and patients with post-polio syndrome.**

A slight case of equinus foot blocks the upper ankle and prevents dorsiflexion. As a result, the knee is indirectly extended and the patient is able to remain upright passively (»plantar flexion – knee extension couple«, ► chapter 4.7). Neither orthosis management nor an operation is indicated to correct this type of equinus foot. On the contrary, a foot with free dorsiflexion should be secured *conser-*

*vatively* in a functional orthosis. This must be prepared with a slight backward lean (= equinus foot position) so that the knee is stabilized indirectly, thereby achieving the same effect as a slight equinus foot. Otherwise the only option for protecting the knee from giving way in flexion is by supporting it with the hand (► Chapter 4.7.4).

A slight hyperextension of the knee of up to  $5^{\circ}$  is acceptable, whereas a higher angle will overstretch the knee capsule and can lead to knee problems with anterior pain at a later date. Ideally, the hyperextension should be prevented indirectly by a corresponding orthosis for the lower leg and foot with an integrated heel. Shells designed to stabilize the knee in the sagittal plane hardly achieve the desired goal at all if the knee is not blocked towards



■ Fig. 3.436. Patient with left-sided *poliomyelitis* after a dorsally extending talar osteotomy (*Operation according to Lambrinudi*) to reduce the footdrop tendency. Since dorsiflexion is not blocked at the ankle by this procedure and the extensors at the knee and hip are not strong enough to compensate for the lack of power in the triceps surae, a crouch gait will result

■ Table 3.74. Structural deformities in primarily flaccid locomotor disorders and muscular dystrophies

Deformity	Functional benefit	Functional drawbacks	Treatment
Equinus foot	Knee extension	Dynamic instability due to small weight-bearing area Deformation of the feet	Functional orthosis (in equinus foot) Cast correction Tendon lengthening (beware of overcorrection)
Clubfoot	Compensates for increased external rotation of the leg	Walking/standing aggravated	Functional orthosis Tendon lengthenings Arthrodesis

extension. Regular stretching of the triceps surae by the physical therapist or splint treatment (possibly with a postural splint) is indicated for preventing severe cases of equinus foot that interfere with standing and walking. The alternative options of a functional orthosis in an equinus foot position or *surgical* lengthening of the contracted muscle apply only to patients with a pronounced contracture of this muscle who are able to walk. In all cases, a slight residual equinus foot position is needed for functional purposes.

❗ **A powerful triceps muscle or a marginal or slight equinus foot are functionally beneficial since this blocks the lower leg in relation to the foot and prevents it from moving forward. This facilitates the development of the indirect extension moment at the knee, which enables the patient to stand without activating the knee extensors.**

### Structural clubfoot

#### ➤ Definition

Hyperactivity of the medial dorsiflexors of the foot with a lack of power in the lateral dorsiflexors and often in the triceps surae muscle.

A clubfoot position that becomes skeletally fixed over time can occur in patients with myelomeningocele who are paralyzed at the level of L4/L5 as a result of constant traction on the tibial muscles with missing antagonists and a lack of activity on the part of the triceps surae muscle (■ Fig. 3.437). The abnormal loading and overloading of the lateral edge of the foot together with the impaired sensation increases the risk of poorly healing pressure ulcers.

As a *conservative* option, the foot position must be held and stabilized at an early stage in order to prevent deformities as far as possible. Functional orthoses, which are required in any case to replace the lack of triceps surae activity, are suitable for this purpose. On completion of growth, the foot position can be corrected *surgically* and an arthrodesis performed to compensate for the lack of the stabilizing function of the triceps surae.

➤ **A structural club foot deformity is also frequent in Charcot Marie Tooth disease. As loss of muscle strength is progressive, transfers are not indicated. Clinically a short plantar fascia and tibialis posterior are found. It can be helpful to ameliorate the position and mobility of the foot to release the plantar fascia (Steindler) and to lengthen the posterior tibial tendon.**

### Other deformities

The development of a ball-and-socket joint in the upper ankle in patients with myelomeningocele has been described.



■ Fig. 3.437. Clubfoot in myelomeningocele with paralysis level at L4

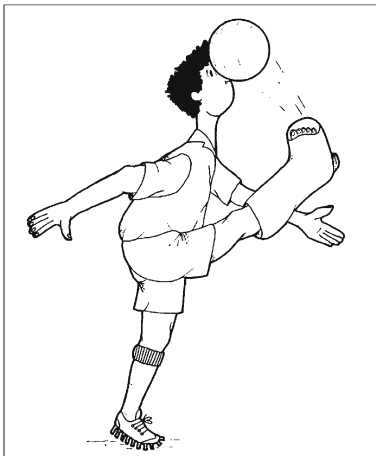
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### 3.4.11 Ankle and foot injuries

C. Hasler



#### 3.4.11.1 Fractures of the distal tibia

##### Occurrence

Fractures of the distal tibial metaphysis account for 5% of all fractures occurring during the growth phase.

##### ! The fracture patterns for the distal tibia and ankle show typical age peaks:

- compression and bowing fractures in younger children,
- medial malleolar fractures around the age of 10,
- epiphyseal separations during puberty,
- transitional fractures during physiological physal closure.

Epiphysiolyses and epiphyseal fractures of the distal tibia are the second commonest fractures that affect the growth plates after distal radius fractures.

##### Diagnosis

##### Clinical features

Localized pain, often accompanied by pronounced perimalleolar swelling. Deformities are readily apparent on clinical examination because of the thin perimalleolar soft tissue covering. If the x-ray shows an undisplaced but impressive epiphysiolysis, the doctor should examine the patient for clinical signs of a rotation deformity.

##### Imaging investigations

Standard AP and lateral x-rays, including the adjacent ankle. Views of the upper ankle in internal and external rotation increase the sensitivity in cases of suspected epiphyseal fracture. The projection in internal rotation provides a view of the distal tibiofibular joint space and enables the examiner to assess the congruence of the

intersection of the fibula, tibia and talus. In complex fracture patterns (e.g. for transitional type II triplane fractures; see below), a CT scan with 3D-reconstruction can be helpful for surgical planning.

##### Fracture types

##### Extra-articular fractures (metaphyseal)

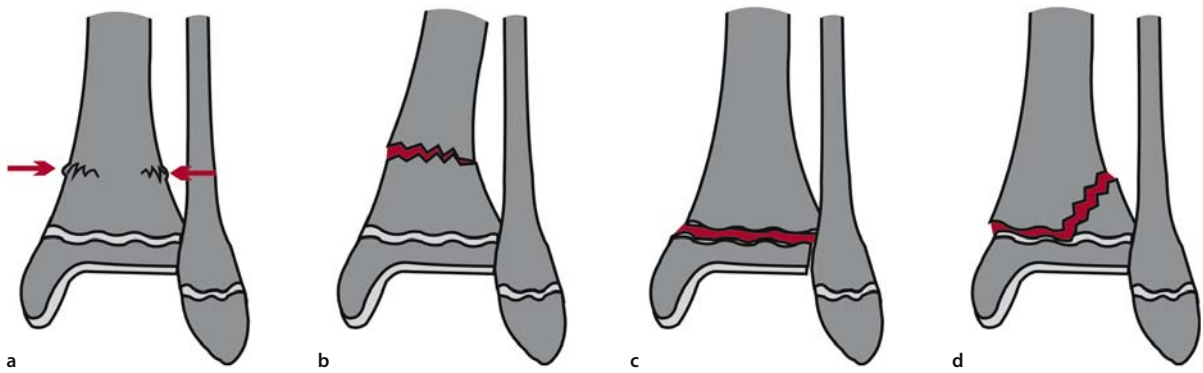
- *Compression fractures* frequently show slight recurvation, but no deviation in the frontal plane. They are stable (■ Fig. 3.438a).
- *Bowing fractures* are associated with the same problem as those of the proximal tibia, i.e. potential partially stimulatory growth disturbances if any deformity in the frontal plane is not consistently corrected. The lower ankle can functionally compensate for slight valgus deformities by inversion, but has only a limited potential for compensation of varus deformities (■ Fig. 3.438b).
- *Epiphyseal separations* (Salter types I and II): separations without a metaphyseal wedge, i.e. type I separations, are rarer and affect younger children than type II separations with a metaphyseal wedge (Thurston-Holland fragment), which primarily occur during puberty (■ Fig. 3.438c).

##### Intra-articular fractures (epiphyseal)

- *Medial malleolar fractures* usually involve the epiphysis (Salter-Harris type III, ► chapter 4.1), less frequently the epiphysis plus metaphysis (type IV), and arise as a result of pressure exerted by the medial edge of the talus against the transition between the distal tibial epiphysis and the medial malleolus (■ Fig. 3.439a, b).

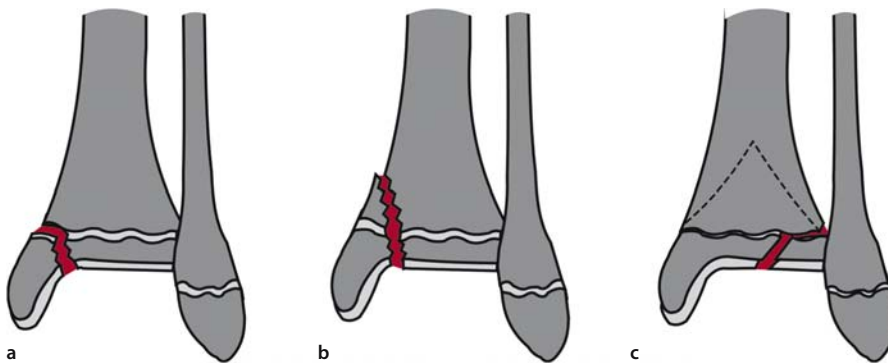
*Transitional fractures* occur as a result of external rotation trauma during physiological physal closure, i.e. in the 1 or 2 year »transitional period« between an open and closed growth plate (■ Fig. 3.439c, 3.440). The ossification front spreads out from the center in a posteromedial, followed by an anterolateral direction. Its asymmetrical advance determines the fracture pattern: the earlier the fracture, the greater the anterolateral fragment, whose horizontal wall runs through the still open physal sections and whose vertical, epiphyseal projections mark the boundary between open and ossifying sections.

- Accordingly, we see very medial, even intramalleolar epiphyseal avulsed fragments or very lateral fragments. The latter lead to the so-called juvenile Tillaux fracture, similar to the bony, anterior syndesmotom disruptions seen in adults. *Twoplane fractures* show the aforementioned basic morphology and are restricted to the epiphysis. *Triplane fractures* additionally show a metaphyseal fracture plane that runs obliquely, with a variable ascent, in a ventral-distal to dorsal-proximal direction. The metaphyseal involvement may



■ Fig. 3.438a–d. *Metaphyseal fractures, distal tibia*: Extra-articular fractures include metaphyseal compression fractures (a), metaphyseal

flexion fractures (b) and epiphyseal separations without (Salter I; c) and with metaphyseal wedge (Salter II; d)



■ Fig. 3.439a–c. *Epiphyseal fractures, distal tibia*: Intra-articular fractures include medial malleolar fractures without (Salter III, a) and

with metaphyseal wedge (Salter IV; b) and transitional fractures (c; see also Fig. 3.440)

be suggested on the AP x-ray by the »Gothic arch« (■ Fig. 3.440). If a continuous epimetaphyseal block is present, the fracture is known as a type I triplane. If the metaphyseal plane continues into the joint at the front, a posterior Volkmann fragment forms, as with malleolar fractures in adults: This rarest and most difficult type of transitional fracture is known as a type II triplane. So-called complex transitional fractures are combined with ipsilateral tibial shaft fractures [9].

### Treatment

*Spontaneous corrections* of varus, valgus and recurvation deformities of up to approx. 10–20° can be expected in patients under the age of 10.

### Conservative treatment

*Non-displaced fractures* and fractures with axial deviations of up to 10° are immobilized in a plaster slab, which is replaced by an encircling cast after the swelling has sub-

sided. The axial deviations are wedged after 7–10 days. (■ Fig. 3.441).

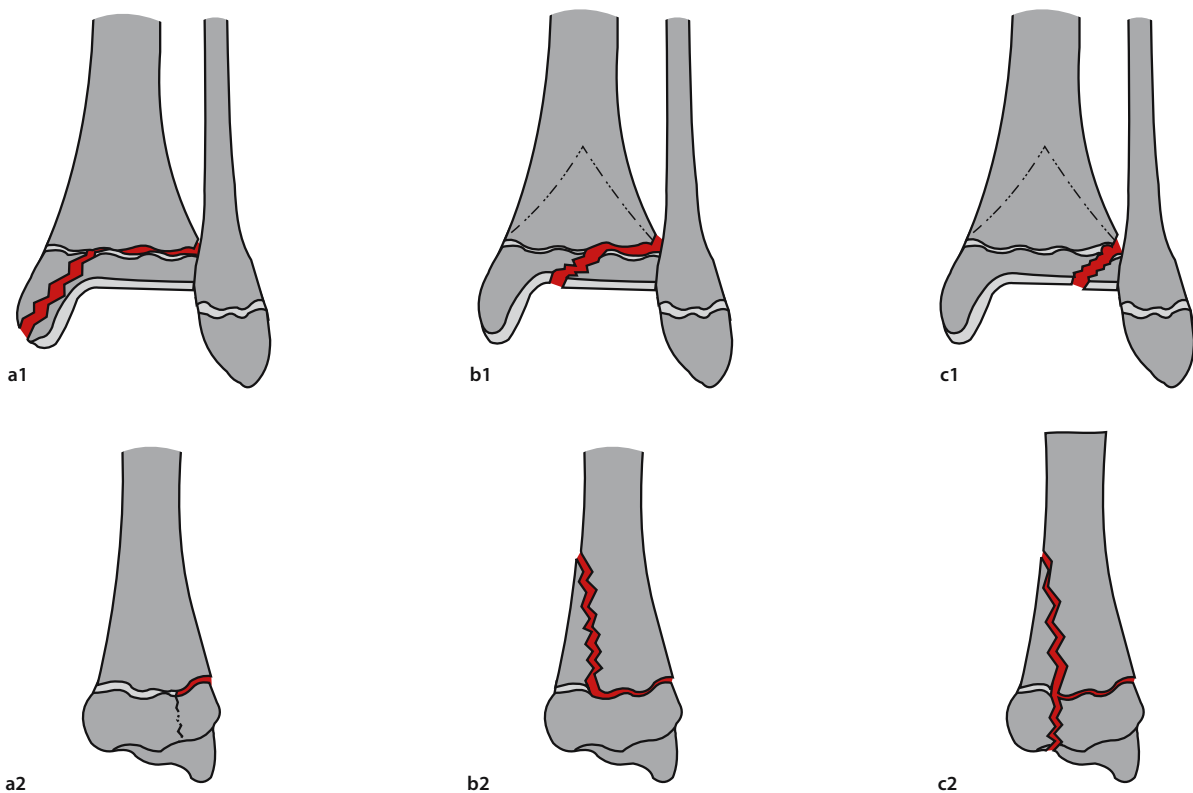
*Greenstick fractures* that are pushed in on the ventral side of the cortical bone and split open on the dorsal side must be fixed in plantar flexion in a below-knee cast in order to achieve dorsal compression and thus timely consolidation.

### Surgical treatment

*Dislocation fractures* require urgent reduction.

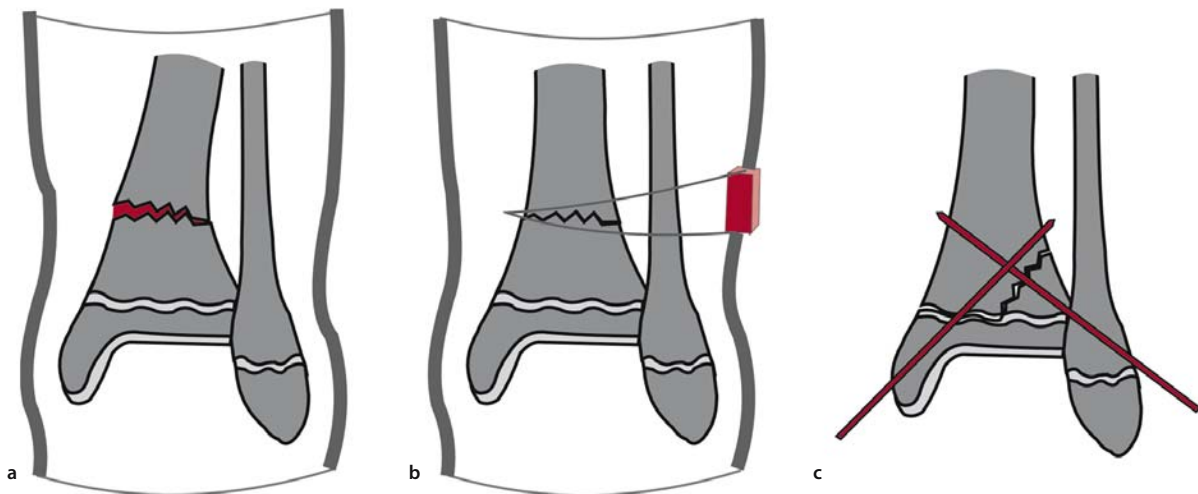
Reducible *epiphysiolyse*s that are not closed in the desired position usually involve the interposition in the plate of a periosteal flap, or even the medial neurovascular bundle, which then has to be released surgically [8].

*Epiphysiolyse*s and *bowing fractures* with axial and/or rotation deformities in excess of 10° are anatomically reduced under general anesthesia, and any instability is fixed with percutaneous, crossed Kirschner wires (■ Fig. 3.441c). If a very large metaphyseal wedge is present, the internal fixation can also be performed using only percutaneously inserted metaphyseal lag screws.



**Fig. 3.440a–c.** Transitional fractures, distal tibia: Top row: A general distinction is made in transitional fractures between twoplane and triplane fractures: In the twoplane fractures one of the fracture planes is in the epiphysis, the other in the epiphyseal plate. In the triplane fractures, an additional metaphyseal fracture forms the 3rd fracture plane. In both types, the fracture gap can appear on the AP view very medially, even involving the medial malleolus, or very laterally. The more physiological physeal closure has already progressed, the more lateral

the fracture. Bottom row: In the lateral or oblique view, two different types of triplane fracture can be distinguished: In a type I triplane fracture, the metaphyseal fracture ends – as with an epiphyseal separation – in the plate. In a type II triplane fracture, the metaphyseal fracture (regardless of the additionally present epiphyseal fracture) crosses the plate and ends in the joint. Both in the epiphyseal separations with a metaphyseal wedge and triplane fractures, the metaphyseal end of the fracture can be seen on the AP view as a »Gothic arch«



**Fig. 3.441a–c.** Treatment of metaphyseal fractures of the distal tibia: The metaphyseal fractures are treated conservatively in a below-knee cast (a). Any primary or secondary axial deviations are corrected around the 8th day with cast wedging (b). In the event of unstable

fractures requiring primary or secondary reduction due to an unacceptable axial deviation the final position should be secured with crossed percutaneous Kirschner wires (c)

## 3.4.11 · Ankle and foot injuries

*Malleolar fractures* with displacement of 2 mm or more should be anatomically reduced and fixed in a »wassertight« manner parallel to the plate with an epiphyseal 4.0 mm cancellous lag screw.

*Transitional fractures* are openly reduced if the intra-articular displacement exceeds 2 mm and then stabilized with epiphyseal or epi- and metaphyseal lag screws (Fig. 3.442). During the open reduction, the joint should be irrigated liberally and checked for concomitant osteochondral lesions and loose fragments. An accompanying fibular fracture usually readjusts itself satisfactorily after the tibia has been reduced.

Depending on the patient's age and the fracture, the *immobilization period* ranges from 2 weeks for compression fractures to 4–5 weeks if weight-bearing with crutches and a below-knee cast is involved.

### Follow-up management

The displacement risk of unstable fractures without internal fixation justifies a visual radiological control after 7–10 days. Patients with bowing fractures, epiphyseal separations and medial malleolar fractures are followed up for at least 2 years after the trauma because of the risk of growth disturbances. Since the risk of a clinically relevant growth disturbance is negligible after transitional

fractures given the imminence of physeal closure, the completion of treatment is based on the restoration of free function and freedom from symptoms.

### Complications

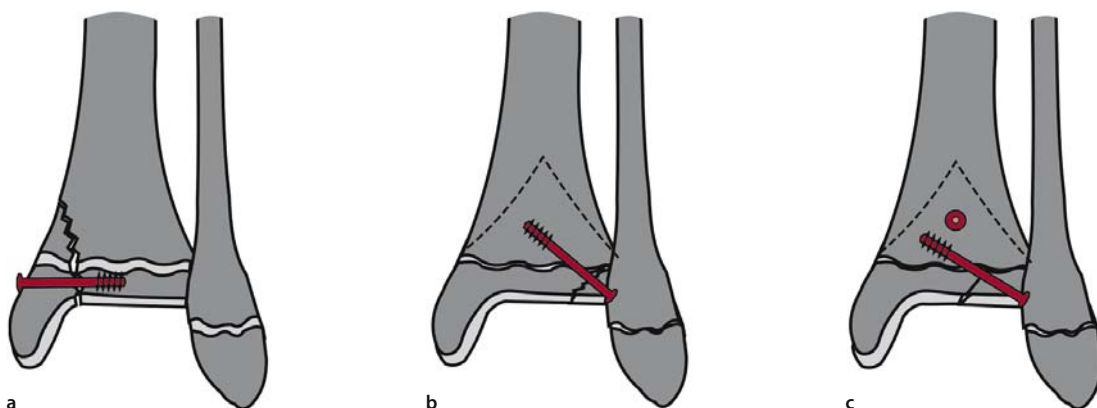
*Pseudarthroses and avascular epiphyseal necroses* are rare.

*Growth disturbances and posttraumatic deformities:* *Premature global physeal closure* is fairly frequently observed in non-anatomically consolidated fractures [1]. Valgus deformities can occur after incorrectly reduced epiphyseal separations with insufficient corrective potential. Varus deformities, on the other hand, are usually the result of an inhibiting growth disturbance.

*Partially stimulatory growth disturbances* after uncorrected valgus deformities in metaphyseal bowing fractures produce a progressive deformity on the tension side until definitive consolidation.

*Partially inhibitory growth disturbances* as a result of physeal bridges are usually located on the medial side and occur after epiphysiolyse and medial malleolar fractures. They are also occasionally observed after fractures with perfect anatomical reduction. Their effect depends on the size and location of the closure and on the amount of residual growth [2]. The resulting varus deformity is only poorly compensated by the lower ankle.

■ **Fig. 3.442a–c.** *Surgical treatment of displaced epiphyseal fractures of the distal tibia:* Displaced epiphyseal fractures – regardless of type – are always reduced openly and secured against movement with small-fragment lag screws. When treating medial malleolar fractures in a patient with open physeal plates, the surgeon must ensure that the screw in the epiphysis runs parallel to the plate, leaving the plate intact. With lateral transitional fractures, the screw may cross the plate since the physiological physeal closure is already well advanced and growth disturbances and relevant consequences are no longer anticipated by this stage. If a triplane fracture is present, the dorsal metaphyseal wedge should be reduced and secured first, followed by management of the epiphyseal fracture



*Rotational deformities* represent a threat particularly after epiphyseal separations. They are clinically evident if the difference from the uninjured contralateral side is more than 10°. An external rotational deformity is usually present. Whether pre-existing rotational asymmetries predispose to fractures remains unclear [14].

### 3.4.11.2 Syndesmotic disruptions and fibular epiphyseal separations

#### Occurrence

Syndesmotic injuries are rare during childhood and adolescence, while fibular epiphyseal separations occur frequently, usually in combination with a fracture of the distal tibia.

#### Diagnosis

##### Clinical features

Local swelling, pain, possibly joint effusion in the upper ankle.

##### Imaging investigation

AP and lateral x-rays

##### Types of fracture

*Epiphysiolyse of the distal fibula with/without metaphyseal wedge (Salter-I or II fracture):*

- in isolation, usually with minimal displacement,
- displaced as a lesion accompanying distal epiphyseal fractures and transitional fractures of the distal tibia.

*Anterior syndesmotic disruptions* are generally equivalent to bony avulsions of the tibial attachment. The size of the avulsed fragment varies from a sliver to a veritable anterolateral quadrant fracture of the tibial epiphysis in the manner of a Salter type III fracture (Tillaux or Kleiger fragment; ■ Fig. 3.443). The latter typically occurs as a transitional twoplane fracture during physal closure.

*Syndesmotic tears*, i.e. purely ligamentous injuries with ankle mortise rupture, are rare and only encountered from late adolescence onward.

#### Treatment

##### Conservative treatment

Lower leg walking cast for 4 weeks for *fibular epiphyseal separation in isolation*, including ad latus deformities of up to 50%. Sarmiento cast for *syndesmotic disruptions with up to 2 mm of displacement*.

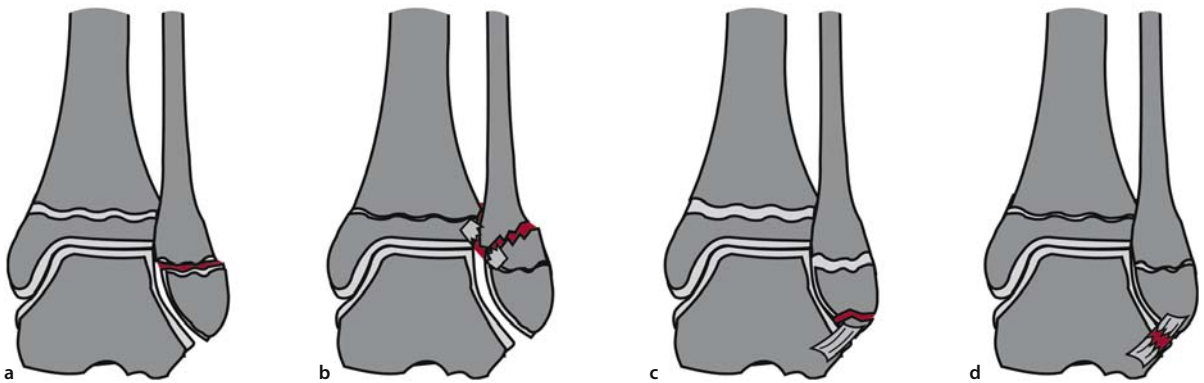
##### Surgical treatment

- *Syndesmotic osseous disruptions with >2 mm of displacement*: Refixation and fibulotibial set screw,
- *Syndesmotic disruptions, ligamentous*: Suturing and fibulotibial set screw.

### 3.4.11.3 Fibulotalar ligament lesions

#### Occurrence

The younger the patient, the earlier a distortion trauma will lead to a bone lesion.



■ Fig. 3.443a–d. *Fibular epiphyseal separation and syndesmotic disruption*: Fibular epiphyseal separations are common, occurring particularly in combination with distal tibial fractures (a). In isolation they can usually be recognized on the lateral x-ray, and occasionally on an AP x-ray, by a metaphyseal wedge of varying size (b). They are often combined with shell-shaped syndesmotic disruptions. Consequently, whenever a fibular epiphyseal separation is suspected, the fibular notch should

be carefully scanned on the 1st AP x-ray for such shell-shaped tears. *Fibulotalar ligament lesions*: The lesion pattern for the fibulotalar ligament is age-related: Below the age of 12, we find periosteal, chondral or bony avulsions with an intact ligament in around 80% of cases (c), whereas intraligamentous ruptures are seen in around 80% of patients older than 12 years (d)



**Diagnosis****Clinical features**

Pain and swelling distally and ventrally to the tip of the fibula. A stability test should not be performed during the first 6–8 weeks, since this is initially painful and does not affect the treatment at all, and even impairs ligament healing during the first few weeks after the trauma.

**Imaging investigations**

AP and lateral x-ray to exclude bone lesions, particularly in the form of ligament avulsions at the tip of the fibula. X-rays with the ankle held in a particular position are obsolete.

**Types of injury**

The relationship between ligament strength and mechanical resistance of the bony attachment determines the injury pattern but does not have any effect on treatment.

*Bony avulsions* occur particularly in the group of *under 12-year olds*.

Ossification on the lateral malleolar tip is either induced by trauma or equivalent to an accessory bone. A shell-shaped fragment suggests a fresh avulsion of the attachment of the anterior fibulotalar ligament anteromedially on the lateral malleolus, which is subsequently rounded down and then no longer distinguishable from a primary accessory ossicle [7]. A differentiated, clearly demarcated, round ossicle is known as an *os subfibulare*. A »genuine« *os subfibulare* is located within the peroneal tendons, i.e. more posterior than attachment tears of the anterior fibulotalar ligament, and is probably far rarer than posttraumatic ossification [6].

*Ligamentous lesions* predominantly involve the anterior fibulotalar ligament after the age of 12. The fibulocalcaneal and posterior fibulotalar ligaments are less frequently affected.

**Treatment**

The treatment of an acute fibulotalar ligament lesion is principally *conservative*: plaster splint for 1 week to alleviate pain and reduce swelling. The findings on inspection after removal of the cast will determine the subsequent course of action. A substantial reduction in swelling and an absence of pain suggest that simple ankle *distortion* has occurred, and prompt transition to full weight-bearing within the limits of pain is permitted. On the other hand, distinct swelling in combination with a hematoma suggest a *ligament rupture*, which is treated functionally with approx. 2-month protection of the lateral ligament complex, e.g. with a pneumatic ankle splint (»Aircast«), a laced ankle brace or a support shoe. In view of the increased risk of trauma recurrence, we additionally prescribe physical therapy with additional proprioceptive and stabilizing training for patients with sporting ambitions or a history of recurrent traumatic sprains.

**Follow-up controls**

History-taking and a clinical assessment of the stability is indicated a few months after the trauma.

**Prognosis**

*Pain*: An *os subfibulare* can cause chronic pain in the vicinity of the malleolar tip. The pain per se or an additional instability are indications for surgical removal of the ossicle, possibly combined with a revision of the ligamentous apparatus.

*Chronic signs of instability* include repeated supination traumas, a feeling of insecurity, perimalleolar swelling, load-related pain and ankle locking. Even if the ligament rupture has been treated appropriately, such signs can be expected in around 10% of cases. Secondary symptoms such as achillodynia or calcaneodynia, and pain or insufficiency of the posterior tibial tendon are common. The clinical examination should include tests for *passive* instability (anterior drawer test for the talus, lateral opening with comparison of both ankles), *static* instability (rear-foot varus) and *dynamic* instability (hyperactivity of the peroneus longus muscle). Surgical treatment is indicated only after several months of unsuccessful proprioception training and muscle strengthening exercises for the lower leg and foot. Possible procedures, depending on the clinical findings, are an anatomical ligament reconstruction and/or a calcaneal osteotomy [16].

**3.4.11.4 Fractures of the foot****Occurrence**

10% of all childhood fractures. Like the hand, the foot is also exposed during play and sport. The midfoot and forefoot are most frequently affected, as this is where distortion, impact trauma and falling objects can lead to fractures. By contrast, only fairly substantial forces will lead to fractures in the rearfoot, hence their rarity during childhood.

**Diagnosis****Clinical features**

The foot skeleton is generally very easy to palpate directly in view of the thin soft tissue covering. Local tenderness and swelling are strongly indicative signs of a fracture.

**Imaging investigations**

Most foot fractures can be clearly demarcated on standard AP and oblique x-rays of the foot. Additional views may be required for the rearfoot area, e.g. axial view of the calcaneus or specific views for the lower ankle. If doubt exists about the indication for surgical correction of calcaneal fractures, a CT scan can be worthwhile in visualizing intra-articular steps.

The objective in differential diagnosis is to distinguish fractures from accessory bones. The latter are clearly de-

marcated from their surroundings and have a roundish appearance.

**!** Often the apophyseal cartilage of the base of the 5th metatarsal or the accessory os vesalianum is confused with an avulsion fracture of the short peroneal tendon. The distinction is simple: The physal plate always runs longitudinally in contrast with the transverse fracture line.

Furthermore, aseptic necrosis of the navicular bone and the 2nd metatarsal head (Köhler's and Freiberg's diseases respectively) or irregularities of the calcaneal apophysis can be confused with fractures or posttraumatic states. Growth plates are present at the proximal end of all phalanges and the 1st metatarsal, and additionally at the distal, subcapital end of the other metatarsals.

### Fracture types

*Calcaneal fractures* in children occur outside the joint in most cases, in contrast with the situation for adolescents and adults, where these fractures are predominantly intra-articular. Since there is usually a history of a fall from a great height, the doctor must always look for additional injuries, particularly in the lumbar spine.

Fractures of the *talus* are the result of forced dorsal extension and therefore run through the talar neck, i.e. are extra-articular. Fractures of the lateral process frequently occur in connection with snowboarding and must be handled according to the principles for the management of intra-articular fractures [11].

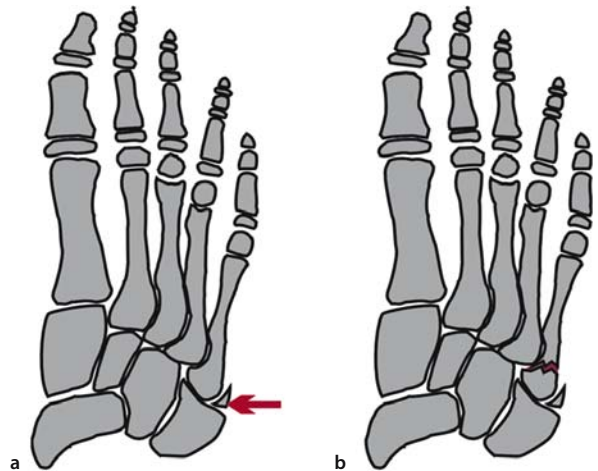
*Navicular fractures* are very rare.

*Cuboid fractures* are rare, but should be considered in the differential diagnosis particularly in children under 4 years of age who refuse to walk or avoid weight-bearing on the lateral edge of the foot (toddler's fracture) [3].

Injuries of the *Lisfranc joint* (tarso-metatarsal joint) occur in plantar flexion trauma [5].

*Metatarsal fractures:* The 5th metatarsal is most frequently fractured in children over 10 years and the 1st metatarsal in children under 5 [13]. Most fractures at the base of the 5th metatarsal show minimal displacement and can be treated conservatively (■ Fig. 3.444). Subcapital and shaft fractures are considered to be non-displaced if the displacement is tolerable or it will correct itself during subsequent growth: no rotational deformity, axial kinks in the frontal plane of less than 10° and sagittal deviations of less than 20° in children under 10 years and correspondingly less with increasing age.

*Toe fractures:* Because of its size and prominence, the great toe is particularly susceptible to trauma induced by kicking and trapping. Fractures of the distal phalanx combined with skin lacerations, bloody tearing of the nail wall or eponychium should be treated as open fractures, otherwise there is a risk of osteomyelitis [10].



■ Fig. 3.444a, b. Apophysis and fracture at the base of the 5th metatarsal: The longitudinally oriented apophysis (a) should not be confused with a fracture. Fractures always run in a transverse direction at this point (b)

### Treatment

#### Spontaneous corrections

While little is known about the remodeling capacity after intra-articular calcaneal and talar fractures in children, the good results obtained after conservative treatment suggest that children have a much wider range of tolerance than adults [4, 12]. Tilt deviations in the area of the *metatarsals and phalanges*, are corrected only in the main plane of movement, i.e. the sagittal plane. The maximum correction is approx. 20° in under 10-year olds, declining thereafter until physiological physal closure (girls: 14 years, boys: 16 years). Rotational defects and deviations in the frontal plane will persist and should therefore be reduced as a primary measure.

#### Conservative treatment

*Calcaneal and talar fractures* in children under 10 years are usually treated with a relieving lower leg Sarmiento cast, which takes the weight off the rearfoot by providing support for the femoral condyles and tibial head.

Because of the precarious blood supply situation we would advise against the open reduction of displaced talar fractures. Such displaced fractures should preferably be reduced indirectly by plantar flexion and percutaneous stabilization.

*Metatarsals and phalanges:* No rotational deformity, axial kinks of less than 10° in the frontal plane and less than 20° in the sagittal plane in under 10-year olds are the requirements for immobilization in a below-knee cast without prior reduction.

#### Surgical treatment

The indication for surgical reduction and stabilization of *talar and calcaneal fractures* in adolescents is based on the

### 3.4.11 · Ankle and foot injuries

guidelines for adult surgery. We also internally fix rare, clearly displaced, extra-articular fractures.

**Metatarsals:** Kirschner wire, internal fixation if instability is present after closed reduction.

**Phalanges:** closed reduction under regional anesthesia for displaced fractures, followed by immobilization with overlapping taping. For fractures of the great toe the fitting of a below-knee splint may be advisable to avoid any stubbing. Displaced intra-articular fractures (particularly condylar fractures) must be reduced openly to an anatomical position and fixed.

#### Immobilization period

Calcaneal and talar fractures: 6 weeks, or possibly longer depending on the radiological result. Metatarsal shaft: 4 weeks. Metatarsal base, subcapital, phalanges: 3 weeks.

#### Follow-up controls

Consolidation may be assessed clinically since the foot skeleton can be palpated directly at any point. If the patient is free of pain, he or she may proceed to full weight-bearing without a cast. Further follow-up controls are only justified in the following situations:

- posttraumatic, troublesome deformities, which may need to be corrected,
- rare, epiphyseal fractures which, because of the possibility of an inhibiting growth *disturbance*, should be followed up for 2 years,
- Talar fractures in view of the risk of avascular necrosis.

#### Complications

**Posttraumatic deformities:** Persisting volar tilts after metatarsal or phalangeal fractures can hinder walking. Varus and valgus deformities and rotational defects can result in the problem of overlapping toes.

Detecting a threatened or established *compartment syndrome* requires considerable alertness on the part of the examiner, particularly in patients presenting with only slight forefoot swelling initially directly after a crush injury or overroll trauma. If doubt exists, immobilization in a below-knee cast and elevation with close, clinical monitoring in hospital is recommended.

**Avascular necrosis** is observed in 15–20% of childhood talar neck fractures, even including undisplaced fractures. Failure to spot the injury initially and an age of under 9 years are risk factors [15]. Since the absence of any radiological displacement on the trauma x-rays does not rule out the possibility of shifting during the trauma followed by spontaneous reduction, the risk of a circulatory impairment is not reduced.

**Osteomyelitis** occurs after trivialized open fractures of the great toe that have received inadequate initial treatment [10].

Premature *posttraumatic arthritic changes* after Lisfranc injuries have been described [5]. Minor radiological changes after calcaneal fractures during childhood are usual. More rarely seen over the long term, however, are signs of arthritis which, in turn, correlate poorly with the clinical findings [4].

We have observed *pseudarthroses* particularly after the conservative treatment of displaced fractures of the base phalanx of the great toe and after displaced condylar avulsions, less frequently after fractures of the base of the 5th metatarsal. If corresponding symptoms are present, open reduction with trimming of the fragments and stable internal fixation is indicated.

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### 3.4.12 Infections of the foot and ankle

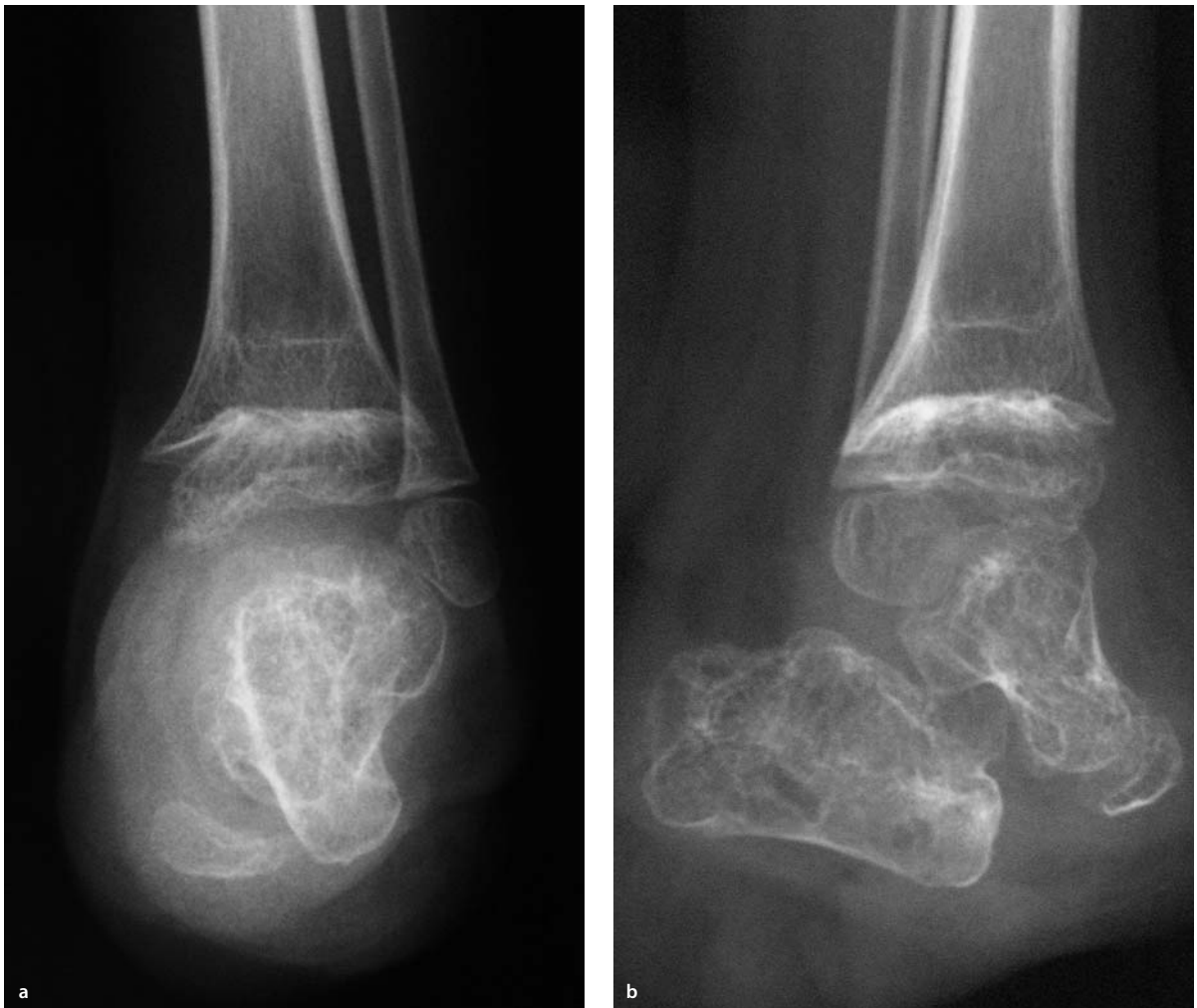
#### **Etiology, frequency and site**

Infections of the foot and ankle have their own distinctive features because of the biological conditions. The circulation is poorer and the temperature lower than in other regions of the body. Hematogenous osteomyelitis is rarer than in other sites and tends to occur in the distal tibial metaphysis. While hematogenous osteomyelitis in the foot is extremely rare, it can occur either in connection with general sepsis (e.g. meningococcal sepsis, ■ Fig. 3.445) or in isolation. The latter type generally involves acute forms of hematogenous osteomyelitis in children under 3 years [3]. The talus is most commonly affected (■ Fig. 3.446), followed by the cuboid and navicular bones [3]. The calcaneus is very rarely affected [4]. Tuberculosis also occurs in the foot, primarily affecting the calcaneus [2]. Children

with sickle-cell anemia can suffer from salmonella osteomyelitis of the feet [1].

The foot is typically susceptible to exogenous infections, e.g. after serious injuries (caused by lawn mowers for example [5]) or if congenital or acquired sensory disorders are present in the lower limbs. Such infections can lead to extensive callus formation, providing a potential portal of entry for insidious bone infections which can then become chronic. Because of the poor circulation and reduced temperature, the course of such conditions is more protracted than in parts of the body closer to the trunk (► Chapter 4.3).

An important differential diagnosis in chronic osteomyelitis is Ewing sarcoma, which can also show a protracted course in the foot, with slow growth and subsequent metastasis to other regions of the body. We have treated two children with Ewing sarcomas that had previ-



■ Fig. 3.445a, b. Neonatal sepsis. AP (a) and lateral (b) x-rays of the foot of a 5-year old girl after an episode of neonatal sepsis with thrombotic vascular occlusions and necrosis of the extremities





■ Fig. 3.446. Acute osteomyelitis of the talus: Left CT scan, right MRI scan of the rearfoot of a 15-year old boy with acute osteomyelitis of the talus. Note the intraosseous abscess

ously been misdiagnosed and treated for years as cases of primarily chronic osteomyelitis. Both have survived despite the greatly delayed diagnosis (currently 10 and 28 years respectively; ■ Fig. 3.448).

#### Clinical features, diagnosis, treatment

The pain often begins after the type of minor trauma that frequently occurs in the foot and ankle area. The diagnosis is often confirmed only at a late stage. If the examiner suspects that the trauma was not severe enough, the possibility of an infection should be considered – even if no fever is present – particularly if the symptoms intensify. Further details of diagnosis and treatment are provided in ► chapter 4.3.

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### 3.4.13 Tumors of the foot and ankle

#### ► Definition

Primary bone tumors originating in the distal part of the lower leg or the bones of the foot or soft tissue tumors arising from the muscles, connective tissue, blood vessels or nerve tissue in the immediate vicinity of the foot (see ► chapter 4.5 for further details about tumors).

#### Occurrence

##### Bone tumors

Primary bone tumors of the distal lower leg and *foot* are relatively rare. In our register we have recorded 285 cases affecting the distal lower leg and 259 cases in the foot. For children and adolescents this corresponds to 6.9% and 5.3%, respectively, of all bone tumors that are not located in the skull. In our patient population, 48% of patients with tumors of the foot or distal lower leg were under 20 years. The distribution of tumor types in the foot in children and adolescents compared to adults is shown in ■ Table 3.75.

The benign tumors that mostly affect the *foot* of the young include osteochondromas, aneurysmal bone cysts, osteblastomas and osteoid osteomas (■ Fig. 3.447). The commonest tumor affecting adults is the enchondroma. Almost the only malignant tumor to affect young people is Ewing sarcoma (■ Fig. 3.448), as confirmed by studies in other countries [6]. By contrast, the chondrosarcoma predominates in adults.



■ Table 3.75. Primary bone tumors of the distal lower leg and foot in children and adolescents (n=264) compared to adults (n=280). (Basel Bone Tumor Reference Center)

	Children and adolescents				Adults			
	Distal lower leg		Foot		Distal lower leg		Foot	
Osteochondroma	30	16.9%	16	18.4%	8	7.4%	16	9.3%
Chondromyxoid fibroma	3	1.7%	0	0.0%	1	0.9%	4	2.3%
Chondroblastoma	–	0.0%	1	1.1%	0	0.0%	7	4.1%
Enchondroma	3	1.7%	7	8.0%	5	4.6%	37	21.5%
Aneurysmal bone cyst	18	10.2%	13	14.9%	2	1.9%	9	5.2%
Non-ossifying bone fibroma	64	36.2%	1	1.1%	9	8.3%	2	1.2%
Giant cell tumor	4	2.3%	2	2.3%	14	13.0%	4	2.3%
Fibrous dysplasia	2	1.1%	1	1.1%	5	4.6%	0	0.0%
Osteofibrous dysplasia (Campanacci)	3	1.7%	0	0.0%	0	0.0%	0	0.0%
Osteoid osteoma/osteoblastoma	15	8.5%	15	17.2%	8	7.4%	15	8.7%
Other benign tumors	7	4.0%	4	4.6%	3	2.8%	11	6.4%
<i>Langerhans cell histiocytosis</i>	5	2.8%	3	3.4%	1	0.9%	1	0.6%
<i>Solitary bone cysts</i>	5	2.8%	10	11.5%	10	9.3%	3	1.7%
<i>Other tumor-like lesions</i>	1	0.6%	3	3.4%	12	11.1%	36	20.9%
<b>Osteosarcoma</b>	<b>5</b>	<b>2.8%</b>	<b>2</b>	<b>2.3%</b>	<b>4</b>	<b>3.7%</b>	<b>8</b>	<b>4.7%</b>
<b>Ewing sarcoma</b>	<b>4</b>	<b>2.3%</b>	<b>6</b>	<b>6.9%</b>	<b>1</b>	<b>0.9%</b>	<b>1</b>	<b>0.6%</b>
<b>Adamantinoma of the tibia</b>	<b>2</b>	<b>1.1%</b>	<b>0</b>	<b>0.0%</b>	<b>1</b>	<b>0.9%</b>	<b>0</b>	<b>0.0%</b>
<b>Chondrosarcoma</b>	<b>2</b>	<b>1.1%</b>	<b>2</b>	<b>2.3%</b>	<b>12</b>	<b>11.1%</b>	<b>15</b>	<b>8.7%</b>
<b>Other malignant tumors</b>	<b>4</b>	<b>2.3%</b>	<b>1</b>	<b>1.1%</b>	<b>12</b>	<b>11.1%</b>	<b>3</b>	<b>1.7%</b>
<b>Total</b>	<b>177</b>	<b>100%</b>	<b>87</b>	<b>100%</b>	<b>108</b>	<b>100%</b>	<b>172</b>	<b>100%</b>

Tumors of the *distal lower leg* are fairly common, particularly in children and adolescents. ■ Table 3.75 shows the distribution of tumor types in young people that affect the distal tibia and fibula compared to adults. Malignant tumors are more likely to occur in adults. The relatively large number of benign tumors in children and adolescents is attributable to the non-ossifying bone fibroma, which affects this site almost as frequently as the proximal part of the lower leg or the distal femur. Osteochondromas (cartilaginous exostoses) are also commonly encountered (■ Fig. 3.449), as are osteoid osteomas and aneurysmal bone cysts (■ Fig. 3.450). Of the malignant tumors, the Ewing sarcoma dominates in young people, while the chondrosarcoma is the most frequent malignancy in adults.

As regards the *location within the foot skeleton*, the talus is particularly predisposed to the development of bone tumors. Osteoblastomas, osteoid osteomas, chondroblastomas and other tumors are particularly found in the talar neck. Tumor-like bone cysts are commonly diagnosed in the calcaneus (■ Fig. 3.451).



■ Fig. 3.447. Oblique view of the forefoot of a 17-year old boy with an osteoid osteoma at the base of the 2nd metatarsal with a typical nidus and surrounding sclerosis.



■ Fig. 3.448a–e. Unusual progression of a Ewing sarcoma. a AP x-ray of the forefoot in a 9-year old boy with a lesion of the 1st metatarsal. A diagnosis of primarily chronic osteomyelitis was made at that time. b AP x-ray of the forefoot and MRI scan (c) of the foot and lower leg 1.5 years later. Only now was a Ewing sarcoma diagnosed. d, e X-rays of the lower leg and foot 9 years after diagnosis, chemotherapy, fore-foot amputation according to Boyd and bridging with allogeneic and autologous bone



Fig. 3.449. X-rays of the distal lower leg of a 13-year old girl with multiple osteochondromas of the distal tibia. The ankle mortise is widened and the fibular is eroded in places



Fig. 3.450. X-rays of the distal lower leg in a 13-year old girl with an aneurysmal bone cyst on the distal tibia



Fig. 3.451. Lateral x-ray in a 18-year old male with a pseudocyst on the calcaneus. Treatment is not required for this type of cyst

! But the diagnosis of »bone cyst« in the calcaneus is almost always incorrect. The trabecular structure of the calcaneus is arranged in such a way that the bone trabeculae are rarefied in a central area. Occasionally, the margins of this central section can also become slightly sclerosed and thus be mistaken for a bone cyst. Histological investigation of this hollow area reveals the absence of any epithelial lining on the walls, which simply constitutes a normal variant (Fig. 3.451).

Tumors are extremely rare in the small tarsal bones, occur slightly more frequently in the metatarsals and phalanges, but only in children over 10 years of age. Osteoid osteomas in these sites can cause unpleasant symptoms over a prolonged period.

### Soft tissue tumors

Soft tissue tumors (► Chapter 4.5.4) of the foot and distal lower leg are even rarer than bone tumors [4]. Out of 83 soft tissue tumors affecting the foot, only the ganglion was frequently diagnosed (24 times), while epidermal cysts, lipomas, synovial sarcomas and hemangiomas were observed with moderate frequency (Fig. 3.452) (approx. 5 times in each case). All other tumors only occurred in isolated cases [4]. Soft tissue tumors can occur at any age and be located anywhere in the foot, although the back of the foot tends to be slightly more frequently affected. Typical features of the synovial sarcoma are fine calcifications and occasional extension into the bone, although the displacement of the adjacent bone is also observed. A similar picture can also be produced by fibrosarcomas, but these are extremely rare in children.

### Diagnosis

Tumors of the distal lower leg and foot are generally diagnosed at an early stage since the soft tissue covering

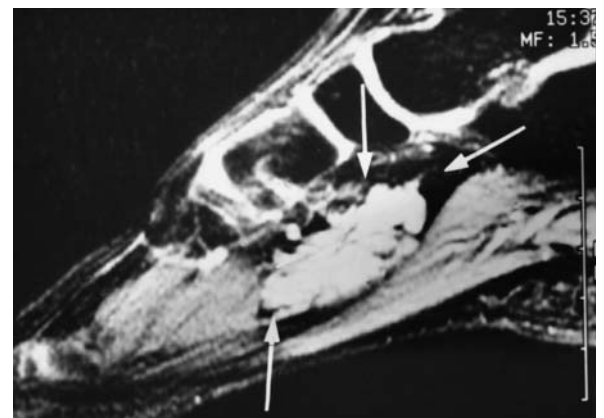


Fig. 3.452. MRI scan of the midfoot (sagittal section) in a 9-year old girl with cavernous hemangioma in the area of the interosseous plantar muscles

of the bone is minimal and any bulging is soon palpable. Pain is often the reason for visiting the doctor. Tumors in this area primarily produce load-related pain before the onset of nocturnal pain, since the shoe can produce pressure-related symptoms even if small bulges are present. The primary diagnosis is always based on an *x-ray* (► Chapter 4.5.1).

Tumors of the foot skeleton also have their own distinctive appearance. Only in the talus can it occasionally prove difficult to diagnose a small tumor clearly among the superposed bone structures. The plain *x-ray* usually allows a clear conclusion to be drawn as to whether a bone tumor is involved. Even the type of tumor can be diagnosed at this stage in some cases, e.g. osteochondromas or non-ossifying bone fibromas. Osteoid osteomas and aneurysmal bone cyst can also be established with a high degree of probability. If doubt exists about the presence of a tumor, a *bone scan* can clarify the situation. An *MRI scan* should be arranged if a biopsy or treatment is planned or if a soft tissue tumor is involved.

### Differential diagnosis

As already mentioned, a common source of error is the diagnosis of a solitary bone cyst in the calcaneus. This diagnosis is almost invariably incorrect. Nevertheless, such »cysts« are repeatedly surgically removed. Another tumor that can sometimes prove difficult to distinguish from a non-neoplastic lesion is the osteoid osteoma in the metatarsals. This site can also be affected by *fatigue fractures*, which can induce the formation of callus. On the *x-ray* this callus can resemble the cortical thickening of an osteoid osteoma (■ Fig. 3.447; ► chapter 3.4.9.5). Irregular sclerosed areas occur in a *melorheostosis*, while *pseudotumors* are observed in hemophilia. Occasionally, *foreign bodies* can induce tumor-like bone changes. Cases of *aseptic bone necrosis* can also be confused with tumors. As always with unclear tumors, the possibility of *osteomyelitis* should be considered in the differential diagnosis.

### Prognosis and treatment

The prognosis for tumors of the foot is better than that for similar tumors in other parts of the body, probably mainly because the foot has only a thin soft tissue covering and tumors are therefore diagnosed at an earlier stage than in other regions of the body. It is also perfectly possible that the tumors are also different in biological respects, as illustrated by the case shown in Fig. 3.448. A primarily chronic osteomyelitis of the 1st metatarsal was diagnosed in this 9-year old boy. The biopsy showed exclusively necrotic tissue, which appeared to confirm the diagnosis. A repeat biopsy was performed after one and a half years of unsuccessful antibiotic treatment and a gradual exacerbation of symptoms. This time a Ewing sarcoma was diagnosed on the basis of the histological findings. Further imaging investigations have also since

revealed a metastasis in the tibia on the same side. The patient was then treated with neoadjuvant chemotherapy, followed by a forefoot amputation after 3 months, resection of the metastasis in the tibia and bridging with an allograft combined with a vascularized iliac crest graft. Ten years after the diagnosis was made, the patient still survives with no sign of any tumor. A similar course would be inconceivable for a Ewing sarcoma as a primary tumor in the femur.

Around 25 years ago we once had a very similar experience with a Ewing sarcoma of the calcaneus. Here, too, the lesion had been misdiagnosed as osteomyelitis for over a year. This patient is also still alive today.

The following basic options are available as *surgical treatment* for tumors:

- curettage (intralesional resection),
- marginal excision,
- wide resection.

A *marginal excision* should always be attempted in *benign lesions*. If the tumors are near a joint, *curettage* may be sufficient, although, as ever, the quality of the curettage will determine the rate of recurrence (► Chapter 4.5.5).

If a malignant tumor is suspected it must always be biopsied beforehand (► Chapter 4.5.1). Depending on the extent of the malignant tumor in each case, a ray resection may be required, or else parts of the foot, or even the whole foot, may require amputation. Good function can be maintained with a Pirogoff or Boyd amputation, where the calcaneum is fused with the tibia and the heel pad is preserved (see ► chapter 3.4.5.11, also ■ Fig. 3.402). For tumors on the distal lower leg amputation should be avoided if possible, since even fairly large defects can be bridged. The vascularized fibula of the contralateral side or substantial allogeneic bone grafts (allografts) are particularly suitable for this purpose, while the use of tumor prostheses is less common. Since prostheses of the upper ankle have been very slow in gaining widespread acceptance, the use of tumor prostheses in this area is also fairly unusual.

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### 3.5 Upper extremities

#### 3.5.1 Examination of the upper extremities

##### 3.5.1.1 Shoulder girdle and upper arm

###### History

- Has *trauma* occurred? If so:
  - When did the trauma happen?
  - During what type of activity (sport, play, daily routine)?
  - Direct or indirect trauma?
  - What movement was involved (external rotation, abduction)?
- *Pain history*: Where is the pain located? When does it occur? Is the pain load-related, movement-related, or does it also occur at rest or even at night? If so, does the pain only occur when the patient changes position or does the patient awake at night because of the pain? For movement-related pain: What specific movements elicit the pain (external rotation, varus or valgus stress)?
- *Dislocations*: Traumatic dislocations are very rare in children and adolescents, whereas habitual and voluntary dislocations of the shoulder girdle frequently occur. During what movement do the dislocations occur, how frequent are they and can they be produced at will?

###### Inspection

- Are *swelling, redness, protrusions* or *muscle atrophy* present?
- *Anomalies*: While anomalies rarely manifest themselves in the shoulder area, hypoplasia or aplasia of the pectoral muscles is occasionally observed (Poland syndrome).

###### Palpation

The patient is examined while seated, ideally on a stool without a back or arm-rests, and the examiner palpates the shoulder region from behind. Children and adolescents very rarely experience pain in the shoulder area. Typical painful sites are the acromion and the coracoid, possibly also the greater tubercle and the long biceps tendon. The proximal humerus is a common site for osteochondromas. A fracture, fracture callus or pseudarthrosis involving the clavicles are all palpable.

###### Examination of the range of motion

The examination can be performed *actively* or *passively*.

- ❗ **The procedure for examining active mobility of the shoulder is very simple: The examiner stands in front of the patient, performs the movements in turn and asks the patient to copy everything.**



Today's child has his hands full



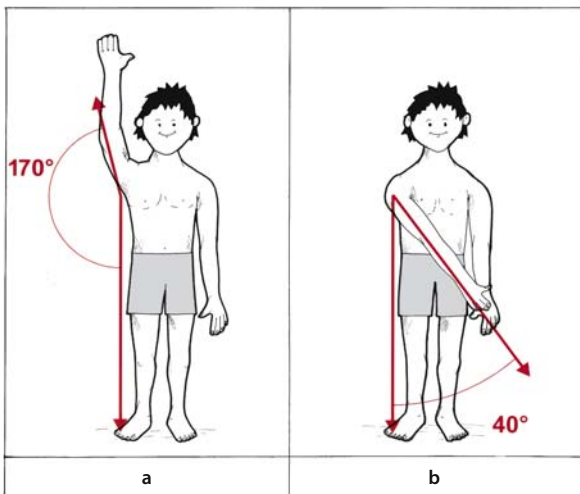
Perhaps future generations will be able to dispense with hands completely...



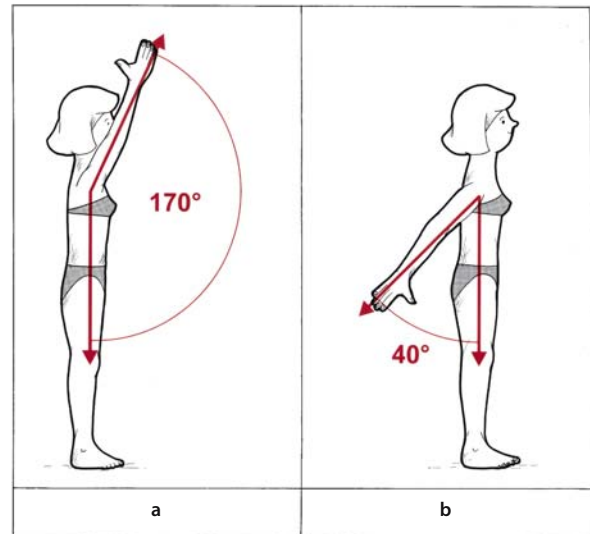
## 3.5.1 · Examination of the upper extremities

We test for passive range of motion only if active mobility is restricted.

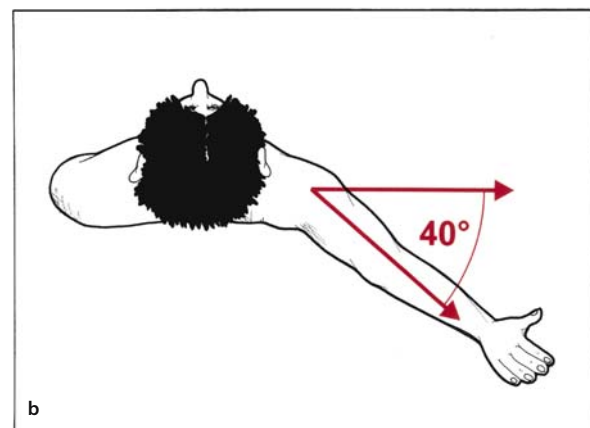
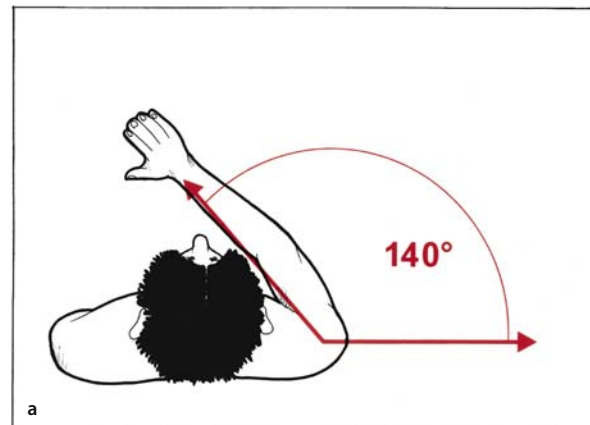
- **Neutral-0 position:** The patient stands with the arms hanging by the side and the thumbs pointing forward (▶ Chapter 2.1.1.3).
- **Abduction/adduction:** The arm is moved from the neutral position out to the side in the frontal plane and then inward (■ Fig. 3.453). Slight external rotation of the shoulder joint and rotation of the shoulder blade must occur to produce abduction beyond 90°. The shoulder blade usually moves synchronously with the arm from the start and must be held in place manually to prevent this movement. For the adduction, the arm is moved inward in front of the body. The typical range of motion for abduction/adduction: 170–0–40.
- **Forward and backward elevation (flexion/extension or forward flexion and backward extension):** From the neutral position, the arm is raised in the sagittal plane out to the front and then backward (■ Fig. 3.454). The typical range of motion for elevation to the front/back: 170–0–40.
- **Horizontal movement of the arm:** The arm is moved forward and backward in the transverse plane from a position of 90° abduction (■ Fig. 3.455). The typical range of motion for horizontal flexion/extension: 140–0–40.
- **Rotation:** With the arm hanging down and flexed 90° at the elbow, the forearm is rotated inward and outward (■ Fig. 3.456). The typical range of motion for external/internal rotation: 60–0–90.
- **Combined movements are tested with the neck grip** (■ Fig. 3.457 a) and **apron grip** (i.e. adduction, retroversion and internal rotation, as if tying an apron at the back) (■ Fig. 3.457 b).



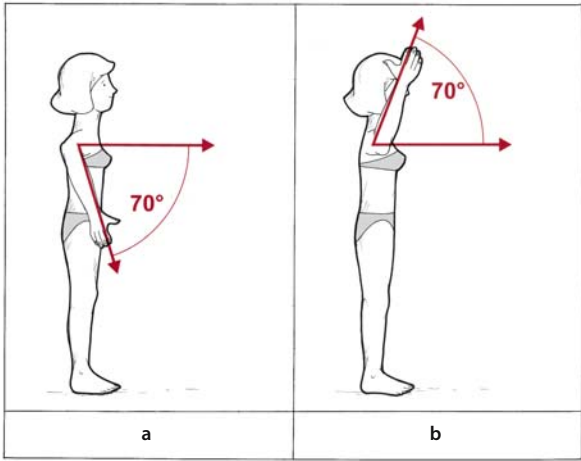
■ Fig. 3.453a, b. **Abduction/adduction in the shoulder:** The arm is raised sideways in the frontal plane as high as it will go (abduction; a) and then moved across the chest to its most medial position (adduction; b)



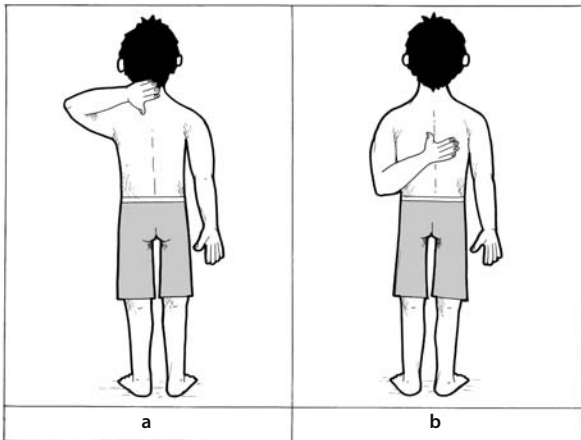
■ Fig. 3.454a, b. **Forward (flexion) and backward (Extension) elevation in the shoulder.** From the neutral-0 position and with the elbow extended, the arm is raised in the sagittal plane out to the front (a) and then back (b) as far as it will go



■ Fig. 3.455a, b. **Transverse movement in the shoulder:** The examination is performed in 90° abduction: The arm is moved as far as possible to the front (horizontal flexion, a) and back (horizontal extension b)



■ Fig. 3.456a, b. Rotation in the shoulder with arm abducted by 90°: From the baseline position of the shoulder with the forearm pointing forward and flexed 90° at the elbow, the forearm is rotated (a) upward (external rotation) and (b) downward (internal rotation). The maximum deviation from the baseline position is measured



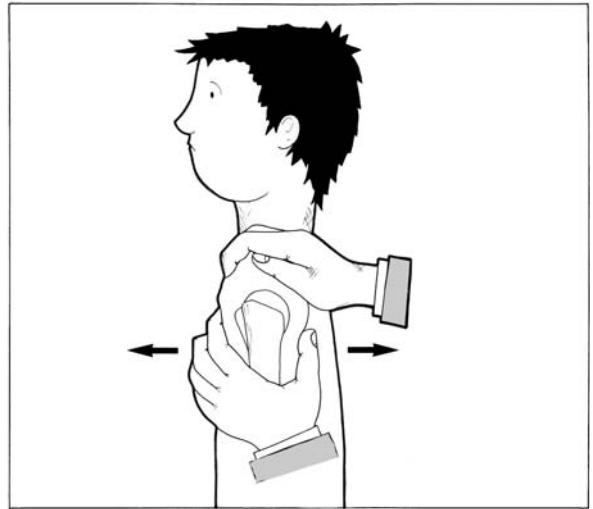
■ Fig. 3.457a, b. The neck grip is a combination movement required in everyday use (the hand is placed on the back of the neck; a). In the apron grip, the hand is moved to the lumbar spine (b)



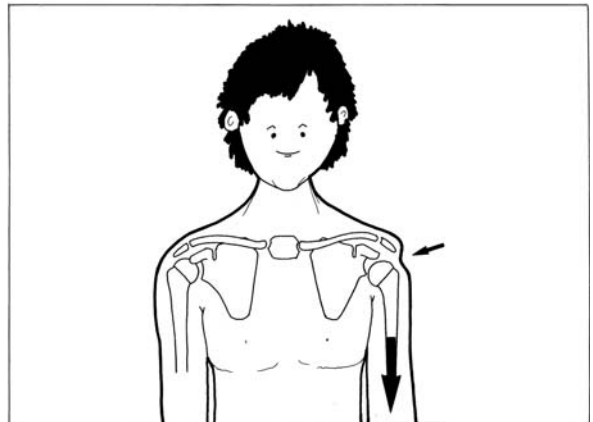
Some children have a different idea of the »apron grip«...

**Stability**

Testing for *glenohumeral translation (load and shift test)*: With the patient seated, the examiner stabilizes the shoulder with one hand from above and behind, while the other grasps the head of the humerus from the side (■ Fig. 3.458). The examiner must first check whether the humeral head is actually centered (it can be displaced in ventral, dorsal or caudal directions). After the initial centering of the humeral head, displacement to the front and back is checked and estimated in centimeters. The examiner then pulls the elbow downward and observes whether an indentation forms under the acromion (*sulcus sign*) (■ Fig. 3.459). Any displacement



■ Fig. 3.458. Testing for glenohumeral translation: One hand stabilizes the scapula while the other grasps the head of the humerus and pushes it forward and backward to check the subluxability of the humeral head

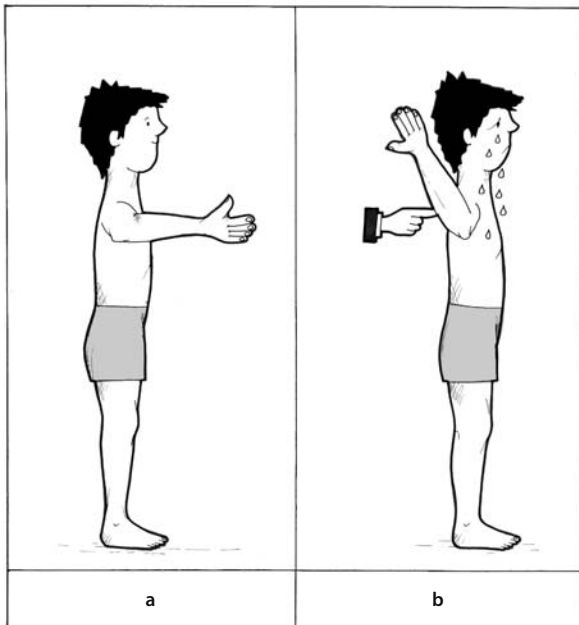


■ Fig. 3.459. To test for the *sulcus sign*, the examiner pulls the elbow downward and observes whether an indentation forms under the acromion. The displacement can be estimated in centimeters

### 3.5.1 · Examination of the upper extremities

in the caudal direction can also be estimated in centimeters [2].

**Apprehension test:** The apprehension test involves the partial reproduction of the dislocation event and the provocation of the patient's feeling before the dislocation. Most dislocations occur in a forward direction in abduction and external rotation. To test for an anterior dislocation, the examiner abducts the arm by  $90^\circ$  and starts to rotate it outward. With the increasing external rotation, the patient perceives the impending dislocation (*apprehension sign*; ■ Fig. 3.460). This test should be used with caution, avoiding the actual reproduction of the dislocation.



■ Fig. 3.460a, b. *Apprehension test:* To test for an anterior dislocation, the examiner abducts the arm by  $90^\circ$  (a) and starts to rotate it outward (b). With increasing external rotation, the patient perceives the impending dislocation (*apprehension sign*)

#### 3.5.1.2 Elbow and forearm

##### History

The patient is asked about trauma, pain and signs of locking. If *trauma* has occurred, the forearm should also always be checked for injuries (Monteggia fracture!). If *pain* is present, the examiner must establish whether it is movement- or load-related or whether it occurs at night. The precise circumstances of any *locking* must also be investigated.

##### Inspection

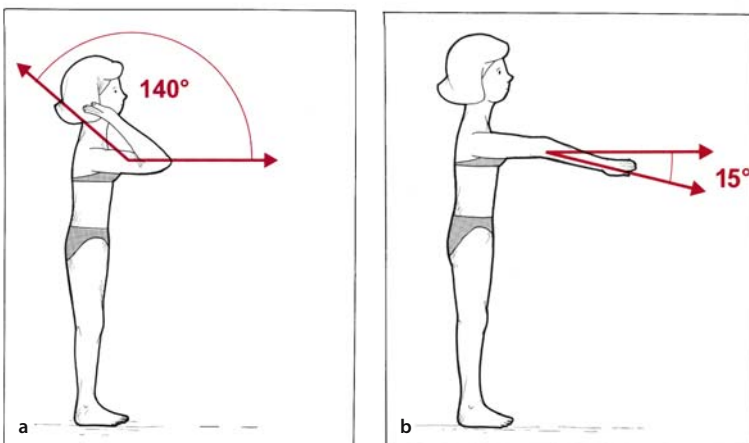
- We note any *swelling*, *redness* or *protrusions*.
- *Anomalies* in the elbow area are rare. Malformations such as longitudinal or transverse deformities or ring constriction are immediately apparent.
- *Axial deviations* must be checked. A valgus position of  $5^\circ$ – $10^\circ$  with the elbow extended is normal (and usually more pronounced in girls than in boys). A valgus position in excess of  $15^\circ$  corresponds to a cubitus valgus, while a cubitus varus is considered to be present if there is a varus position of  $5^\circ$  or more.

##### Palpation

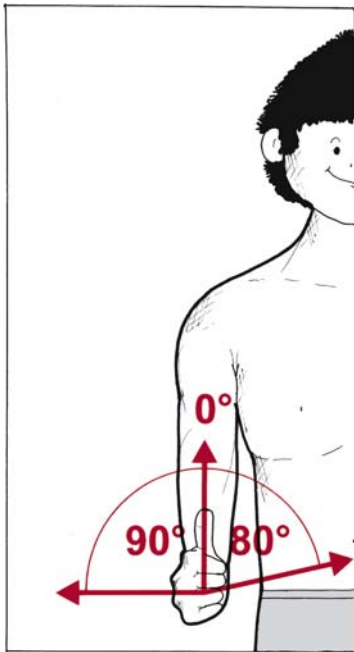
We palpate the joint capsule and look for an effusion, which is readily palpable particularly at the back at the level of the joint space. We also examine for tenderness of the radial head. This is usually readily palpable on the lateral side slightly distal to the joint space. Pronation and supination movements are also readily perceived. It may also be possible to elicit pain on applying pressure to the epicondyles.

##### Examination of the range of motion

- Neutral-0 position: Extended elbow (▶ Chapter 2.1.1.3).
- Flexion/extension: We actively and passively measure maximum flexion and extension (■ Fig. 3.461). The typical range of motion for flexion/extension;  $140$ – $0$ – $5$  in boys and  $140$ – $0$ – $10$  in girls. Hyperextension up to  $15^\circ$  is still normal.



■ Fig. 3.461a, b. *Flexion/extension in the elbow:* The arm is stretched out to the front and flexed (a) and extended (b) as far as possible at the elbow. In children, the elbow can usually be hyperextended by  $5$ – $10^\circ$



**Fig. 3.462. Pronation and supination in the elbow:** This test is performed with the elbow in 90° flexion. In the neutral position, the thumb points upwards. The upper arm axis serves as a proximal reference line and the transverse wrist axis as the distal pointer. The degree of supination (thumb in the lateral direction) is usually somewhat greater than the extent of pronation (thumb turned toward the body)

- Pronation/supination : These are measured from the following base position: The elbow is positioned next to the body and flexed by 90°, the forearm points straight forward, with the wrist extended and the thumb raised (■ Fig. 3.462). The forearm is rotated internally and externally about the longitudinal axis. The typical range of motion for pronation/supination: 80–0–90.

### Stability

After *ligament lesions*, a lateral instability can be established as follows: The examiner grasps the upper arm with one hand and the forearm with the other and performs a forced valgus and varus rotation. Comparison with the other arm is particularly important during this examination.

#### 3.5.1.3 Hand

##### History

###### — Trauma:

In addition to acute trauma, chronic trauma will also need to be investigated in the presence of certain symptoms. Chronic wrist pain is very common and attributable to a repetitive trauma (e.g. if the patient

on rollerblades repeatedly comes up against a wall and supports himself with his hands).

###### — Family history:

Certain deformities can be inherited (Apert syndrome, including polydactyly, syndactyly, symbrachydactyly, split hands, camptodactyly, see ► chapter 3.5.3), although these are usually the result of damage during pregnancy or spontaneously-occurring new mutations.

### Inspection

- We note any *swelling*, *redness* or *protrusions*. These occur in association with trauma, infection or tumors. The distal ulna is a not infrequent site for osteochondromas. Effusions and swellings of the hand caused by rheumatoid arthritis are rare in children and adolescents in contrast with adults. The major joints tend to be more affected in children.
- *Anomalies* are usually clearly apparent on visual inspection (► Chapter 3.5.3). Certain malformations are characteristic of certain *hereditary disorders*, for example the abducted thumb with a short metacarpal in diastrophic dwarfism, dystrophy of the nails toward the thumb side in nail-patella syndrome (► Chapter 4.6.4.17) and the typical hand crease seen in Down syndrome (► Chapter 4.6.5.1).

### Palpation

We palpate *tender sites* (e.g. the lunate bone in lunate malacia) and *protrusions* if a tumor or inflammation are suspected.

### Examination of the range of motion

#### — Neutral-0 position:

Hand with extended wrist and fingers, and thumb in line with the fingers.

#### — Flexion/extension (palmar flexion/dorsal extension) of the wrist:

We actively and passively measure the maximum flexion and extension (■ Fig. 3.463). The typical range of motion for flexion/extension: 70–0–80. Hyperextension up to 90° is still normal.

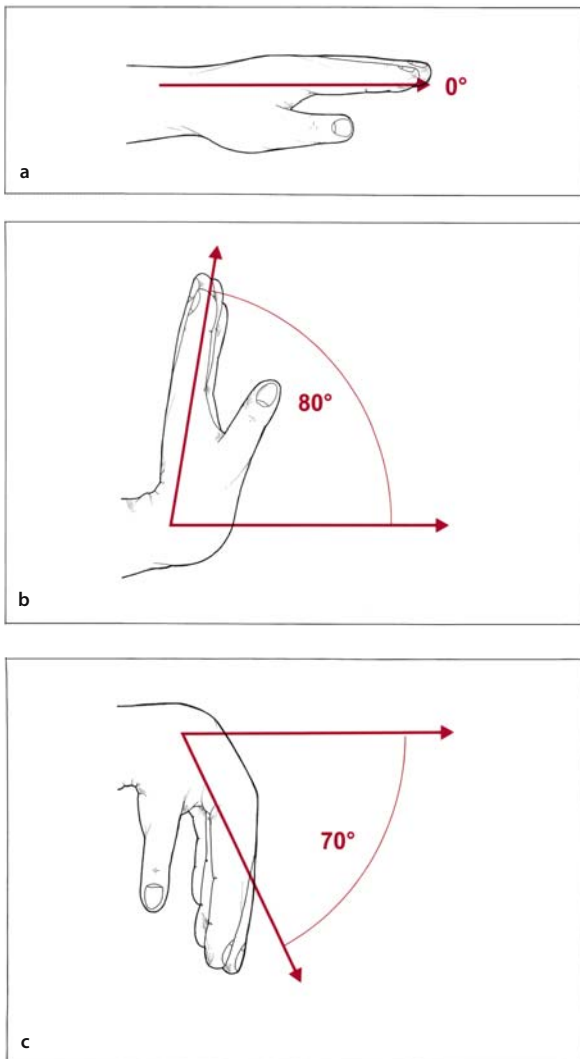
#### — Radial abduction/ulnar abduction:

From the neutral position, we actively and passively flex and extend the wrist to measure the abductability in the direction of the radius and ulna. The typical range of motion for radial abduction/ulnar abduction: 30–0–40 (■ Fig. 3.464).

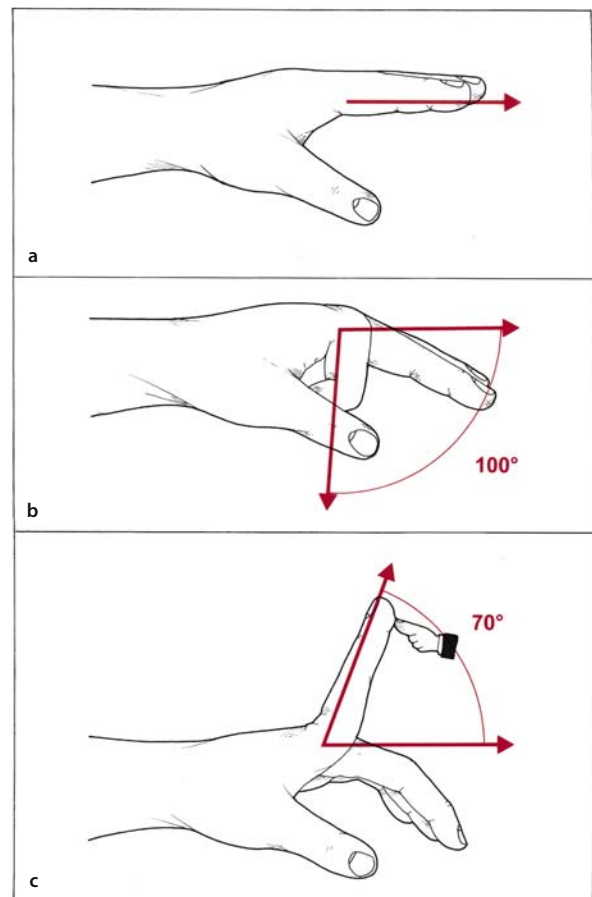
#### — Flexion/extension (palmar flexion/dorsal extension) of the finger joints:

We actively and passively measure the maximum flexion and extension of each individual finger joint (■ Fig. 3.465). The typical range of motion in the metacarpophalangeal (MP) joints for flexion/extension: 70–0–80. Hyperextension beyond 90° is a sign of general ligament laxity.

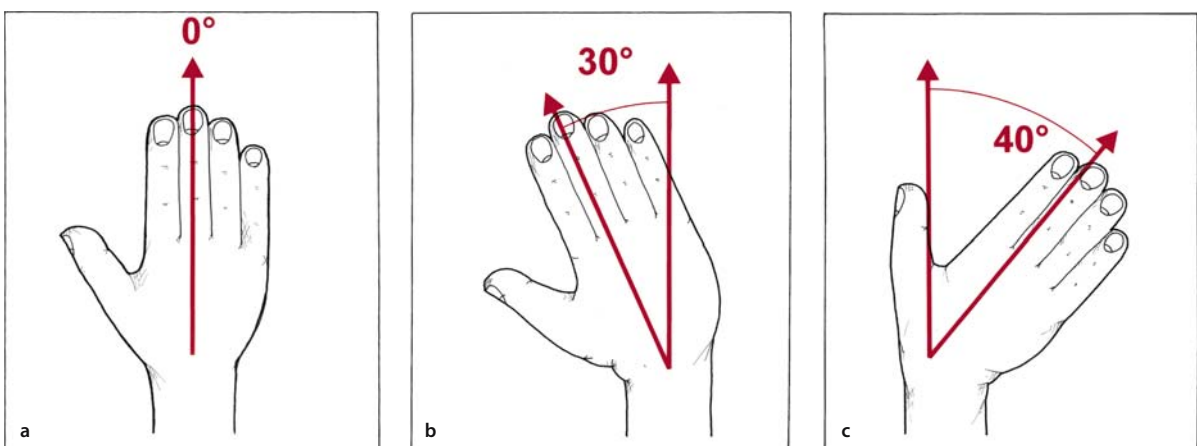
## 3.5.1 · Examination of the upper extremities



■ Fig. 3.463a–c. *Palmar flexion/dorsal extension in the wrist*: From the neutral position (a) the maximum deviation in the dorsal (b) and palmar (c) directions is measured



■ Fig. 3.465a–c. *Flexion/extension of the finger joints*: The flexion and extension of each individual joint can be measured actively and passively. From the neutral position (a) the joint is actively flexed to the maximum extent (b). While hyperextension cannot be achieved actively, the metacarpophalangeal joint can be hyperextended passively beyond 90° in children with ligament laxity. Figure c shows the testing of the metacarpophalangeal (MP) joint. The proximal interphalangeal (PIP) and distal interphalangeal (DIP) joints are tested in a similar manner

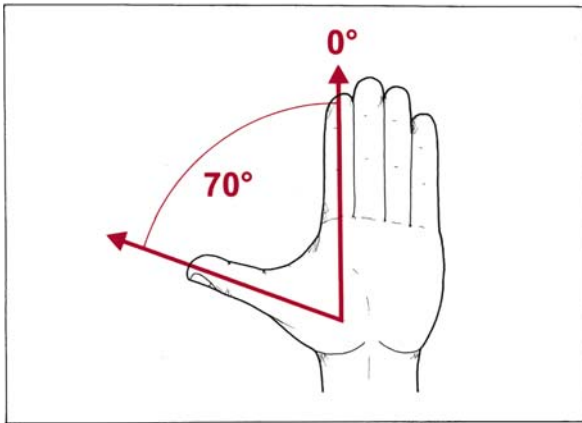


■ Fig. 3.464a–c. *Radial abduction/ulnar abduction in the wrist*: This test is generally performed with the hand pronated. From the neutral position (a) the maximum deviation of the hand in the radial (b) and ulnar (c) directions is measured

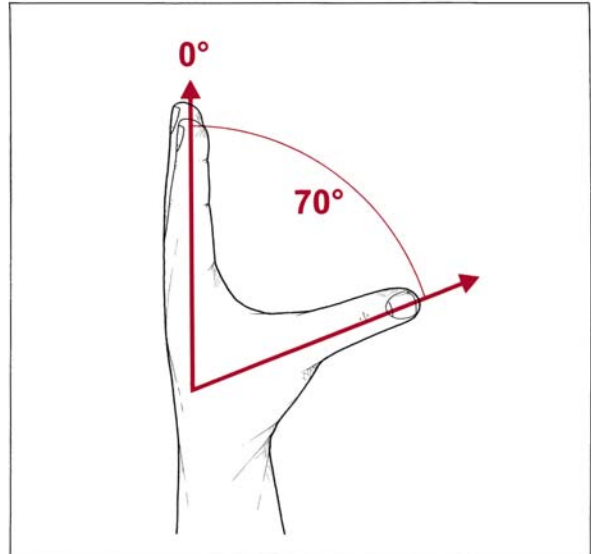


— *Thumb movements*

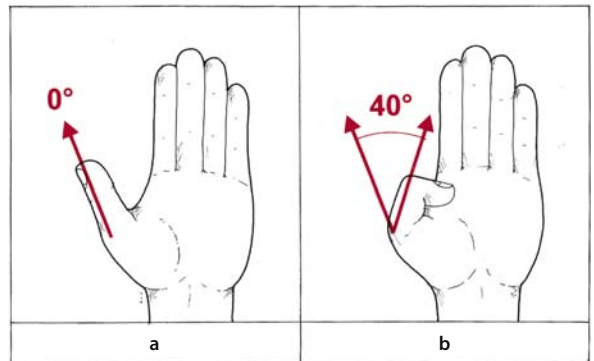
We actively and passively measure the ability to abduct and adduct (■ Fig. 3.466, normal range: 70–0–30), antepulsion/retropulsion (■ Fig. 3.467, normal range: 70–0–30) and flexion/extension at the MP and IP joints (■ Fig. 3.468). Examination of the *pinch grip* is extremely important from the functional standpoint (■ Fig. 3.469). The examination of the *thumb-forearm gap* provides an indication of the general condition of the ligamentous apparatus. General ligament laxity is present if the thumb can be pushed back against the forearm or if the gap is 1 cm or less (■ Fig. 3.470).



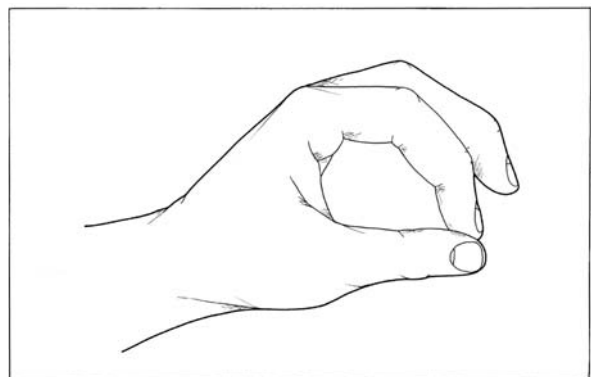
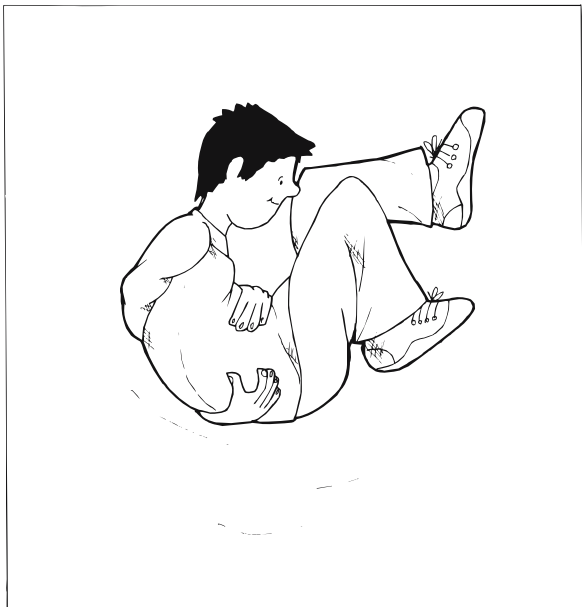
■ Fig. 3.466. *Abduction of the thumb*: This test is performed in the palmar plane from the neutral-0 position (the zero line corresponds to the axis of the index finger)



■ Fig. 3.467. *Antepulsion/retropulsion of the thumb*: This test is performed at 90° to the palmar plane from the neutral-0 position

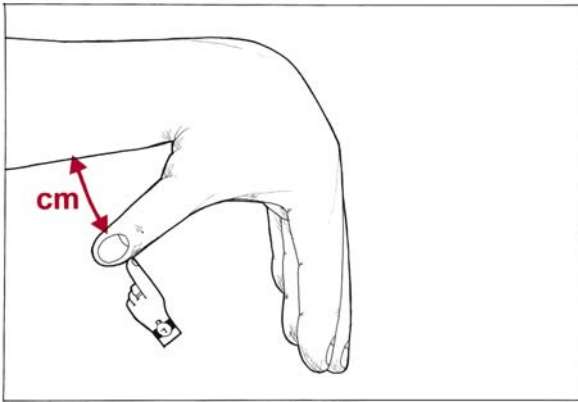


■ Fig. 3.468a, b. *Flexion/extension of the thumb*: This test measures the basic flexion and extension movements in the thumb: a maximum extension; b maximum flexion



■ Fig. 3.469. The *pinch grip* is the most important combination movement of the hand in functional respects. This test checks whether the thumb tip and the tip of the 2nd (possibly also the 3rd and 4th) finger can be approximated

## 3.5.2 · Radiographic technique for the upper extremities



■ Fig. 3.470. *Thumb-forearm gap*: The thumb is passively approximated to the forearm as far as possible. Normally a gap of 2–3 fingerwidths remains. In children with very lax ligaments the distal phalanx of the thumb can touch the forearm

## References

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### 3.5.2 Radiographic technique for the upper extremities

#### Clavicles, AP and oblique

In contrast with the positional technique applicable to adults and adolescents, for children, toddlers and infants we prefer an AP beam path with the patient standing or supine. If the findings on this view are not clear, the x-ray tube is then angled upwards at 30° to produce an oblique view. With both views, the central beam is aimed at the center of the clavicles.

#### Shoulder, AP with upper arm in 45° internal rotation

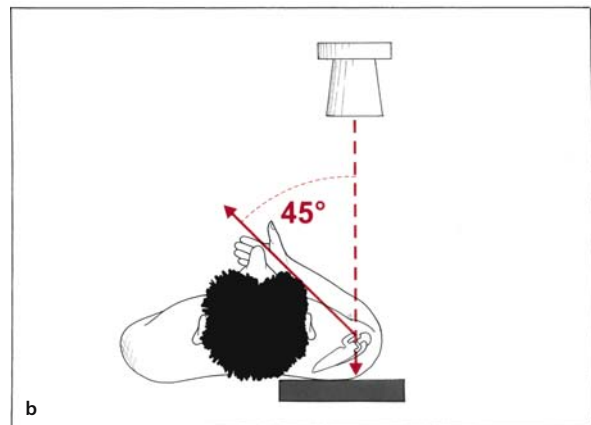
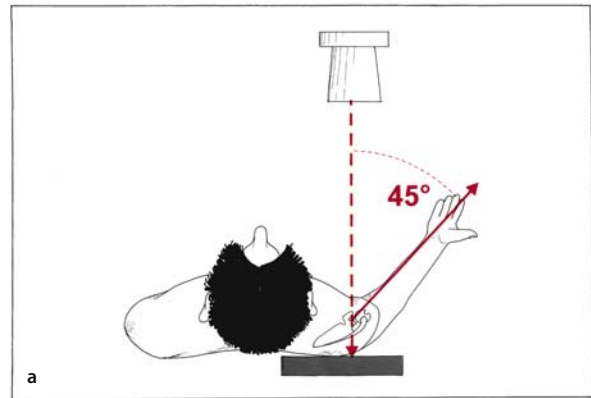
The patient sits with the shoulder blade flush against the cassette. The central beam points to the coracoid and is angled upwards at 15–20° (■ Fig. 3.471 a).

#### Shoulder, AP with upper arm in 45° external rotation

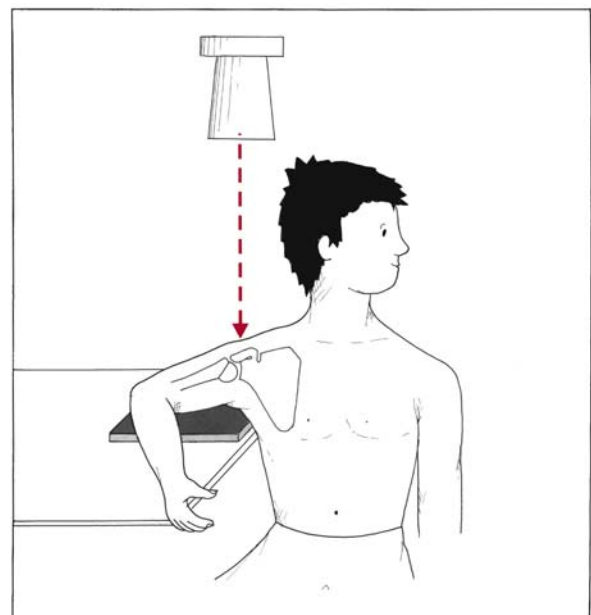
The central beam points to the coracoid and is angled upwards at 15–20° (■ Fig. 3.471 b).

#### Shoulder in 90° abduction, external rotation and 90° flexion at the elbow

The patient sits with the upper arm on the cassette in 90° abduction, external rotation and flexed at the elbow. The central beam is aimed at the humeral head (■ Fig. 3.472).



■ Fig. 3.471a, b. Radiographic technique for the AP views of the shoulder in 45° external (a) and 45° internal rotation (b)



■ Fig. 3.472. Radiographic technique for the axial x-ray of the shoulder

The three views described above provide an overview of the status of the shoulder.

### Shoulder in the event of a suspected dislocation (»Y-view«)

The patient stands with the front of the injured shoulder pointing to the cassette and at an angle of 40° to the cassette in the frontal plane. The elbow is flexed by 90°, the palm of the hand is placed on the patient's head. The central beam is aimed at the center of the scapula (■ Fig. 3.473). Any deviation of the humeral head from the central point of the Y-shape is indicative of a dislocation.

### Upper arm, AP with shoulder and elbow

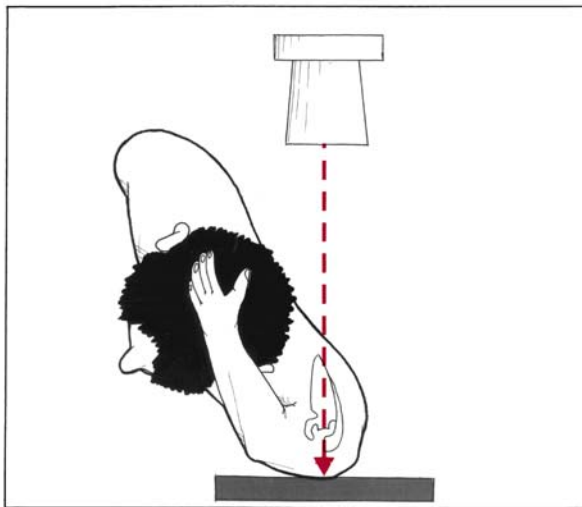
The patient stands or lies on his back with the elbow extended and the hand supinated. The central beam is aimed at the center of the humerus.

### Upper arm, lateral

The patient sits laterally at the edge of the table with the upper arm abducted by 90°, the elbow flexed by 90° and the hand supinated. The central beam is aimed vertically at the center of the upper arm.

### Elbow, lateral with 90° flexion of the joint

The patient sits at the table with the arm abducted, the ulnar side of the elbow resting on the cassette and the hand supinated. The central beam is aimed at the radial head (■ Fig. 3.474 a).



■ Fig. 3.473. Y-view of the shoulder. The patient stands with the injured shoulder at an angle of 40° to the cassette plane. The central beam is aimed at the center of the scapula. This view is particularly effective for showing any forward or backward displacement of the humeral head

### Elbow, volodorsal

The patient sits at the table with the elbow extended and the hand supinated. The central beam points to the center of the elbow joint (■ Fig. 3.474 b).

### Whole forearm AP, with wrist and elbow

Toddlers and infants lie on their back on the examination table. The elbow is extended and the hand supinated; the central beam is directed at the center of the forearm.

### Whole forearm, lateral

The patient sits laterally at the examination table with the elbow flexed by 90° and the wrist extended and supinated. The beam path travels in a radioulnar direction.

### Wrist, lateral

The wrist and fingers are extended, with the ulnar resting on the cassette. The central beam is aimed at the wrist, radioulnar beam path (■ Fig. 3.475 a).

### Wrist, dorsovolar

The hand is pronated on the cassette. The central beam is directed at the center of the wrist (■ Fig. 3.475 b).

### Scaphoid (navicular), dorsovolar

Position: The hand is pronated and abducted towards the ulnar side, the wrist rests on the cassette with the fingers flexed and the thumb extended (■ Fig. 3.476 a).

### Scaphoid (navicular), AP and lateral

For the lateral view, the ulnar side rests on the cassette with the hand slightly dorsiflexed. The fist is loosely closed and the beam path is aimed at the scaphoid in a radioulnar direction (■ Fig. 3.476 b).

### Thumb, volodorsal

Position: The extensor side of the thumb rests on the cassette with the hand in maximum pronation. The central beam is aimed at the thumb metacarpophalangeal joint at right angles to the cassette.

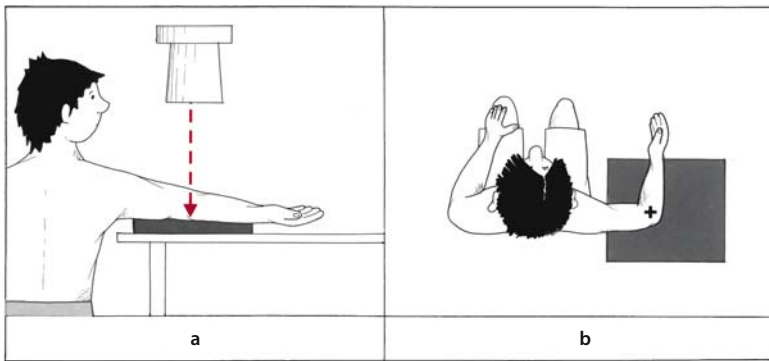
### Thumb, lateral

Position: The radial side of the thumb rests on the cassette. The hand is raised on the ulnar side by approx. 20°. The central beam is aimed at the thumb metacarpophalangeal joint (■ Fig. 3.477).

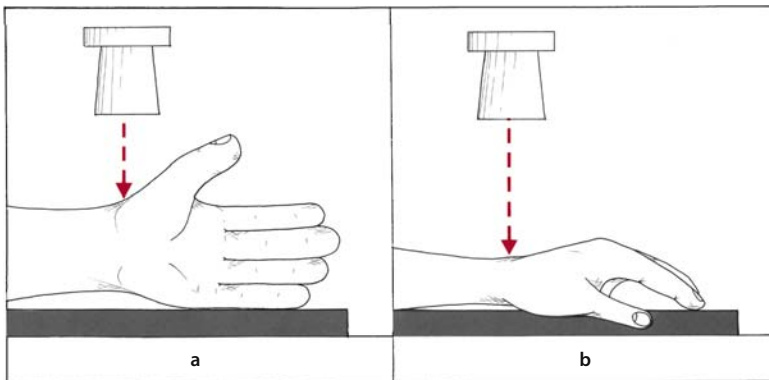
### Whole hand, dorsovolar

Position: The hand rests on the cassette with the fingers extended and slightly apart. The central beam is aimed at the head of the 3rd metacarpal. The epiphyses of the radius and ulna must be included in the x-ray if the bone age needs to be established. With uncooperative toddlers, it is sometimes better to x-ray the hand in supination with

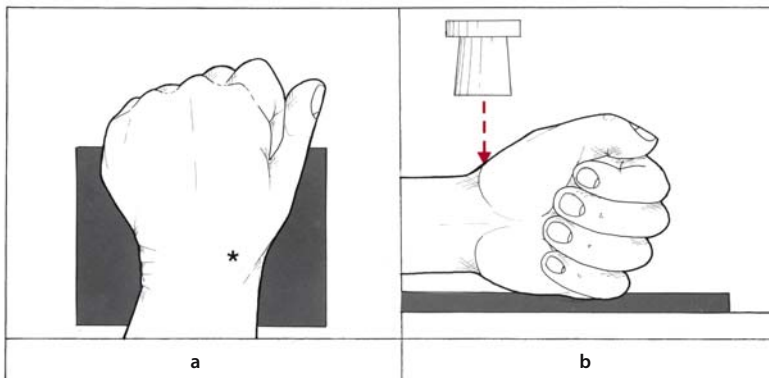
## 3.5.2 · Radiographic technique for the upper extremities



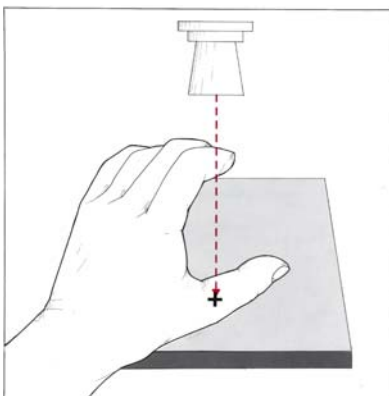
■ Fig. 3.474a, b. Recording technique for x-rays of the elbow: (a) AP, (b) lateral (see text)



■ Fig. 3.475a, b. Recording technique for x-rays of the wrist: (a) lateral and (b) AP (see text)

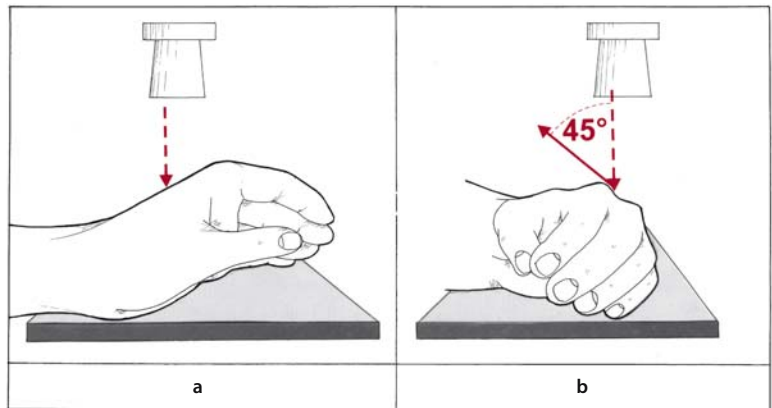


■ Fig. 3.476a, b. Specific view for the scaphoid bone: AP (a) and lateral (b, see text)



■ Fig. 3.477. Lateral x-ray of the thumb (see text)

**Fig. 3.478a, b.** Oblique x-ray of the hand. View from the side (a) and from the front (b). The hand is pronated and the radial side is raised by approx. 45°. The central beam is aimed at the head of the 3rd metacarpal



the aid of a 10 cm wide Plexiglas strip secured on both sides with two sandbags.

### Whole hand, oblique

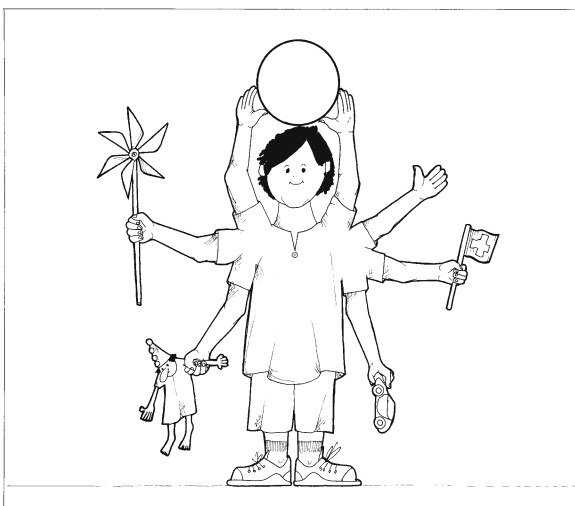
**Position:** The hand is placed in pronation with the radial side raised by approx. 45°. The ulnar side rests on the cassette. The central beam is aimed at the head of the 3rd metacarpal (■ Fig. 3.478).

## 3.5.3 Congenital deformities of the upper extremities

### 3.5.3.1 General aspects – classification

#### ► Definition

Congenital anomalies of the arms and/or hands that have formed before birth. Most cases result from damage that occurs during early pregnancy, although certain malformations are also inherited.



### Occurrence

Figures on the occurrence of congenital deformities are difficult to obtain. In a study of 50,000 births in Edinburgh, the authors calculated that just 3.3% of all deformities were located in an upper limb [38]. The incidence of all malformations and hereditary disorders is estimated at 2–3%, which roughly means that 1 anomaly of an upper limb is observed per 1000 births [18, 26], although more recent investigations have found only approx. 0.4 anomalies /1000 births [40]. This reduction in the incidence is possibly attributable to the improved prenatal diagnosis with ultrasound, which allows the fingers to be recognized as early as the ninth week of pregnancy and most malformations to be diagnosed by an experienced examiner in the 15th week.

A sudden increase in the frequency of severe limb deformities occurred at the end of the 1950's. This rise was attributable to the drug thalidomide, which caused serious damage when taken during pregnancy (between days 26 and 50). After the connection was finally confirmed in 1961, the incidence returned to its previous level.

### Classification

In the middle of the 19th century Saint-Hilaire [39] introduced Greek terms to describe various malformations. Thus an »amelia« referred to the absence of an extremity (Greek: melos = limb). Other terms included »hemimelia« (Greek: half limb), »phocomelia« (Greek: seal limb) and »ectromelia« (Greek: ectros = absence). Since this term ectromelia has been used to describe a wide variety of malformations it has proved unsuitable as a precise description.

The first useful systematic classification was proposed by Frantz and O'Rahilly in 1961 [17]. This classification formed the basis for the current classification, which has been modified and adopted by various international associations, including the »National Academy of Sci-



■ Table 3.76. International classification of congenital deformities of the upper extremity

Type	Description			Example
I	Formation defects	Transverse	Terminal	Phalangeal, carpal, metacarpal, forearm, upper arm
			Intercalary	Symbrachydactyly, phocomelia
		Longitudinal		Radial (preaxial) or ulnar (postaxial) clubhand, split hand
II	Differentiation (separation) defects			
III	Duplication			Polydactyly, triphalangeal thumb
IV	Overgrowth			Macroductyly
V	Hypoplasia			Thumb hypoplasia, Madelung deformity
VI	Ring constriction syndrome			–
VII	Generalized skeletal anomalies			Apert syndrome, Poland syndrome, arthrogryposis

ence», the »American Society for Surgery of the Hand«, the »International Federation of Hand Societies« and the »International Society for Prosthetics and Orthotics«. This classification has now gained general acceptance [31] (■ Table 3.76), even though the old Greek terms still surface occasionally in the literature.

*Failure of formation* refer to all deformities in which a part of the body has not formed correctly. *Transverse arrest* run across the axis of the extremity. If these are terminal, they constitute a congenital amputation, which is described according to the level of the amputation. If distal parts of the extremity are still present, the deformity is intercalary (i.e. interposed). Rudimentary fingertips at the end of a stump provide a typical example. This form is termed symbrachydactyly, a term that is not commonly used in the English literature but one that has gained widespread acceptance in German-speaking countries. Amputations with preserved fingertips are much more common than terminal deformities. An extreme form is the lack of an upper arm and forearm with (partially) preserved hand, a condition previously termed phocomelia.

*Longitudinal arrest* can be located preaxially (on the radial side), centrally or postaxially (on the ulnar side). The thumb and index finger are often missing together with the radius, while the absence of the ulna is usually associated with the absence of the 4th and 5th fingers. Since the absence of one of the two forearm bones leads to a characteristic bowing of the remaining bone and to a deviation of the wrist because of the lack of any counter support, the deformity is also known as a radial or ulnar clubhand. In a central defect, the 2nd, 3rd and possibly 4th rays, with the corresponding metacarpals, are missing. This is known as a split hand or »lobster-claw hand«.

A failure of *differentiation* involves a problem with the separation of tissues. The commonest and most typical deformity of this kind is syndactyly. Radioulnar synostosis is also not infrequently seen.

*Duplication* arises as a result of the splitting of the embryonic tissue, which leads to the formation of the supernumerary phalanges. After syndactyly, polydactyly is the commonest deformity affecting the hand. In *overgrowth* a single part of the body (usually a finger) is enlarged. Macroductyly are more common in the lower limbs than the upper limbs.

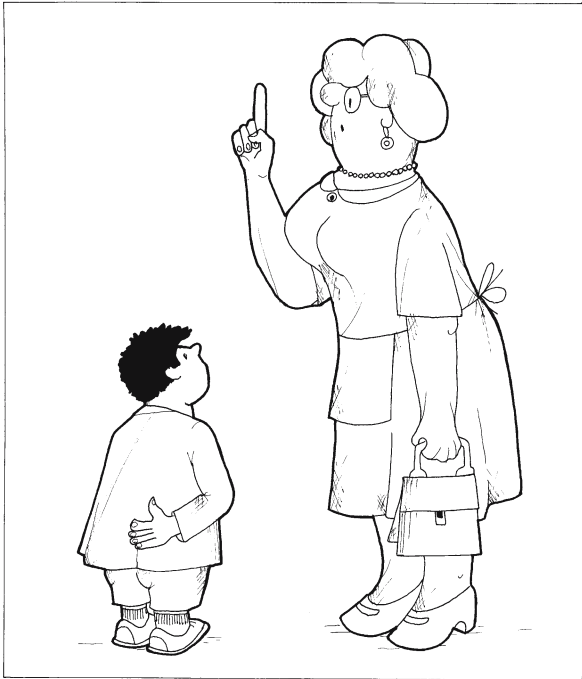
In *hypoplasia* the skeleton and soft tissues of a part of the body are formed smaller than normal, while a *ring constriction syndrome (congenital band syndrome)* involves a complex picture of intercalary and terminal soft tissue, and also in some cases bony, deformities.

The term *teratological series* is used to characterize the severity of the damage. Thus, a slight impairment may lead to hypoplasia of the thumb, while a serious impairment results in aplasia.

Although this classification does not include *congenital deformities in the area of the shoulder girdle* (Sprengel deformity, clavicular pseudarthrosis), these are discussed at the end of this chapter since the girdle forms part of the upper extremity.

### Management of children with congenital hand deformities

The hand is far more than just a part of our locomotor apparatus. It also represents a hugely important sense organ and a means of expression. We »speak« with our hands, and use them to repel, embrace, threaten, caress and warn. We extend the hand of friendship, live from hand to mouth. When things are out of control they get out of hand. We help someone by giving them a helping hand.



Parents often force their children to shake hands, although the latter are anxious about large adult hands.



Children try to »grasp« the world

We use our hands to explore our environment by touch. Without this differentiated sense of touch we would be unable, for example, to fish out the car keys from a pants pocket full of individual items. Adults greet each other by extending the hand. Parents often also force their children to do the same, although the latter are often anxious about large adult hands.

It is particularly striking how often our speech uses contact metaphors to describe emotions. The very word »feeling« is derived from the sense of touch, but we are also »moved«, »touched« or »gripped«, we »suffer under pressure«, are »nervous« when stressed, we can be »injured« and »sensitive« or »stirred« by poems and stories. If we understand something, we »grasp« it.

Since it usually comes as a shock to the parents when they realize that their newborn child has a missing or deformed body part, it is very important to provide competent advice at an early stage and given them a realistic picture of what they can expect. Some mothers and fathers have completely unrealistic ideas about the possibilities of modern medicine. Others will suppress the problem and may miss out on genuine possibilities for improvement and the help that is available for the child.

Most parents of children with deformities have strong feelings of guilt, which are invariably under the surface in any case, often with a religious origin. The deformity is felt as a »punishment from God« or a sign of »original sin«. Such feelings can be strongly reinforced by an excessively

detailed history-taking, since hardly any mother can be completely certain about what harmful substances she may or may not have come into contact with during early pregnancy (particularly during the period when she had been unaware of her pregnancy). It is cruel to connect the child's deformity with a glass of wine or cough preparation that she may have taken at that time. Asking about the family history can also arouse feelings of guilt. Not infrequently the father or mother will blame each other. Consequently, taking a detailed pregnancy and family history is useful only if it has implications for the diagnosis and/or treatment or if *both* parents express a wish to discuss the history. With most deformities, the precise identification of the harmful substance is irrelevant and should therefore be disregarded.

The parents should be advised by a team so that all aspects of the underlying problem (including additional deformities) and treatment can be taken into account.

The team should include the following members:

- an orthopaedic surgeon,
- a hand surgeon trained in microsurgery,
- occupational therapist,
- orthotist.

If possible, these specialists should assess the patient and advise the parents jointly. In certain instances, *genetic*

## 3.5.3 · Congenital deformities of the upper extremities

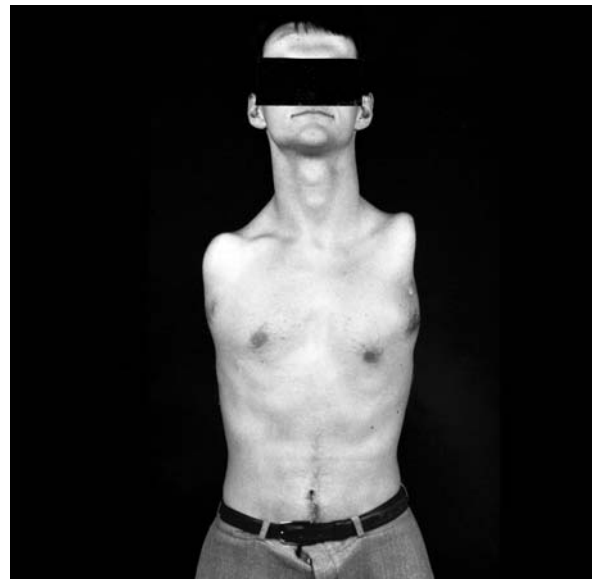
■ **Table 3.77.** Most favorable ages for surgery

Deformity	Condition/Operation	Age
Syndactyly	Simple	12 months
	Bony	8 months
	Acrosyndactyly	4 months
Clubhand	Centralization	12 months
	Pollicization	2 years
	Lengthening	12 years
Polydactyly	5th finger	4 months
	Thumb	1 year
Finger aplasia	Pollicization	1–2 years
	Finger transfer	1–2 years
	Lengthening	12 years
Ring constriction syndrome	With vascular impairment	Emergency
Symbrachydactyly	Finger stabilization	1–2 years
Delta phalanx	Osteotomy	3–4 years
Radioulnar synostosis	Osteotomy	7–8 years

*counseling* may be required, in which case the corresponding specialists should be on hand.

*Surgical measures* are required for various deformities, and choosing the right time for the operation requires considerable experience. The earlier the operation, the greater the potential for adaptation. On the other hand, the surgical procedure is technically more difficult, the smaller the extremity. For certain procedures the risk of recurrence is also higher. Moreover, it is not possible to obtain the cooperation of very small children in the postoperative phase. While ■ Table 3.77 gives an indication of the most favorable ages for surgery [18], the exact timing must be based on the individual situation of the patient and family and the surgeon's own experience.

For congenital amputations, the possibility of *prosthetic provision* must also be considered, and will require close cooperation with an orthotist. Experience has shown, however, that children with unilateral amputations below the elbow almost never require a prosthesis, or else use an existing prosthesis rarely if at all. The artificial replacement does not provide any functional benefit for such children. If the affected child has one dexterous hand, he or she can largely compensate for the absence of the other provided an upper arm, and possibly forearm, stump is present. With writing, for example, the primary task of the non-writing hand is to hold the paper steady, but this task can also be achieved with the elbow. The only drawback



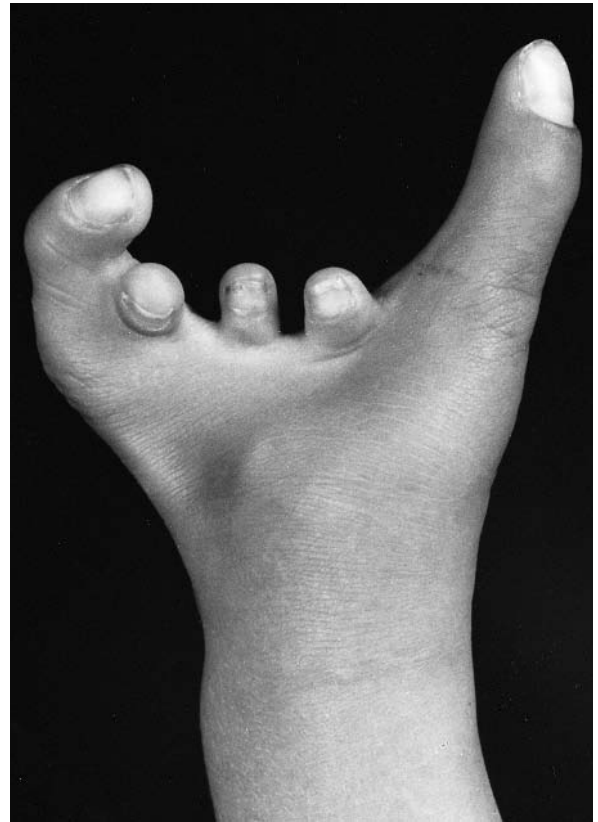
■ **Fig. 3.479.** *Bilateral amelia* in a 16-year old boy. Despite repeated attempts with bilateral prostheses, the patient no longer uses them but performs all tasks using his feet and legs, which he has learned to manipulate with an extremely high degree of dexterity. The lack of any sensation in the prosthetic hands means that they are not suitable for everyday tasks

is that the child has to lean forward more and thus hold the back in an asymmetrical posture. However, the risk of the development of scoliosis as a result of this posture is low. With a rudimentary forearm stump the child can also hold objects in the crux of the elbow, making a prosthesis seem less important. Nevertheless, children should at least be offered the option of an artificial prosthesis so that they themselves can decide whether to wear one or not. If the amputation is located at upper arm level, however, a prosthesis is useful since the reach of such a malformed extremity is too small. Even a lightweight cosmetic prosthesis can be used as a counter support for the other hand to enable the child to pick up objects or stabilize a piece of writing paper. Another option is a mechanical prosthesis with cable traction for the actuation of an elbow or a hook, in which objects can be wedged. Devices are also available for holding a spoon or fork so that the child can eat with both hands. The provision of a myoelectric prosthesis should be reserved for children with a bilateral amputation.

! **It should always be borne in mind that even the best and most sophisticated arm prosthesis cannot provide the most important element, i.e. sensation. Those of us with normal hands tend to think of the hand primarily as a tool. Only when a hand is lacking do we realize that the non-dominant hand is principally an organ of sense (■ Fig. 3.479).**



■ Fig. 3.480. Myoelectric prosthesis in a 6-year old girl with bilateral dysmelia



■ Fig. 3.481. 4-year old boy with *symbrachydactyly of the split hand type with camptodactyly of the little finger*

The control of myoelectric prostheses is now much faster thanks to the use of new electronic components. Some of the latest prostheses can move almost as fast as a normal hand and incorporate a tactile grasp function, i.e. they can adjust the degree of pressure when picking something up so as to avoid crushing the object. The following basic movement directions can be incorporated in a prosthesis: elbow flexion and extension, wrist pronation and supination and a grasp function. However, the incorporation of all three functions makes the prosthesis extremely complex and thus very difficult for the child to manage. The child has to learn how to activate a completely different set of muscles in the upper arm in order to produce a specific hand function (■ Fig. 3.480). Such prostheses also tend to be rather heavy.

### 3.5.3.2 Terminal and intercalary transverse deficiencies (I) (including phocomelia, symbrachydactyly)

#### *Clinical features, diagnosis*

Transverse deficiencies can be terminal or intercalary. Buds of several fingertips are very frequently present at the end of an amputation stump. This intercalary mal-

formation is known as *symbrachydactyly* and is the commonest form of congenital amputation. The amputation is very often at the level of the proximal third of the forearm, usually resulting in a forearm stump length at birth of only 5–7 cm. The elbow is generally normal. The fingertip buds project from the skin at the end of the stump and are highly sensitive. More proximal or distal transverse deformities are less common. Finger joints may be missing completely at phalangeal level. A more common finding, however, is shortening of the finger joints (*brachyphalangia*) or metacarpals (*brachymetacarpia*). Here too, intercalary deficiencies are much more common than terminal deficiencies.

The absence of the carpus is rarely observed. More usual at this level is an absence of the whole hand apart from the fingertips. If central fingers are missing completely the condition is known as a *split hand*. If the central fingers are reduced to a few distal sections (fingertips), then we talk of *symbrachydactyly of the split hand type* (■ Fig. 3.481).

If the metacarpals end in rudimentary phalanges with normal fingertips, the *symbrachydactyly* is said to be of the *short-finger type*. Other forms of *symbrachydactyly* are described as *mono-, bi- and triphalangeal*

### 3.5.3 · Congenital deformities of the upper extremities

[34]. The teratological series in symbrachydactyly is as follows (with increasing severity of the damage): short-finger type, monodactylous type, split hand type and peromelia type.

Symbrachydactyly is not inherited, but based on damage occurring during pregnancy. The clinical examination includes palpation, testing of mobility and, in particular, a functional examination. A crucially important factor is whether the thumb is opposable and whether a pinch grip is possible. Knowledge of the age at which the ossification centers appear is useful for the radiological evaluation (■ Table 3.78).

■ Table 3.78. Age at which the ossification centers appear on the x-ray of the hand

Ossification center	Age
Capitate and hamate bones	2 months
Triquetrum bone	1.7 years
Lunate bone	2.5 years
Scaphoid, trapezium and trapezoid bones	4 years
Metacarpals and proximal phalanges	1.5 years
Middle phalanges	2 years
Distal phalanges	2.5 years

### Treatment

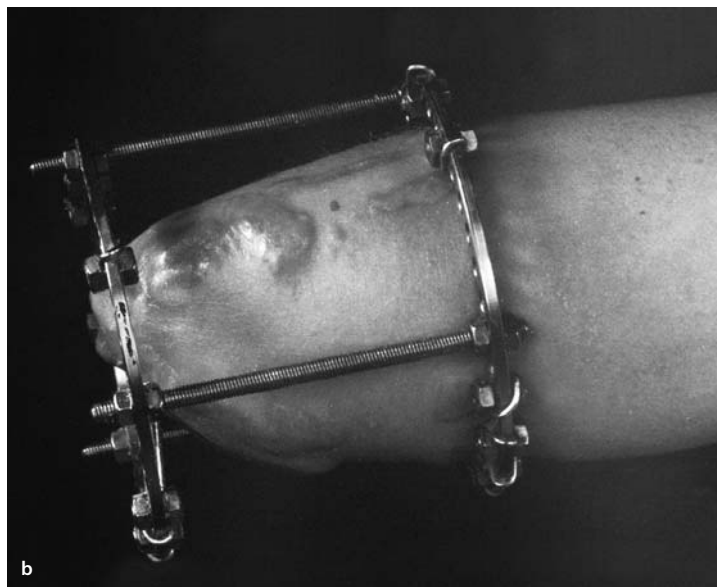
The following surgical options are available:

- stabilization of the finger buds with free bone grafts,
- lengthening osteotomies,
- pollicization,
- microvascular transfer of a toe to the hand.

Lengthening osteotomies are an effective option for achieving functional improvement and, in some cases, a cosmetic benefit as well. It should always be borne in mind, however, that muscle function is invariably made worse and not improved by the extension. The functional gain must therefore be achieved by other means. Thus, for example, the lengthening of an extremely short forearm stump will allow objects to be wedged in the elbow or produce a better fit for a forearm prosthesis (■ Fig. 3.482).

Even the surgical lengthening of the phalanges can produce a functional improvement if it facilitates opposition and thus a pinch grip. As a rule, however, the functional gain is limited since the lengthening of the muscles results in a loss of power, and the surgery tends rather to produce a cosmetic improvement. The functional benefit is particularly significant, on the other hand, if a radial ray incapable of opposition is rotated into a position of opposition and thus allow the patient to perform a pinch grip (pollicization; ■ Fig. 3.483).

Naturally the muscles must be rearranged and modified in order to produce a genuine pinch grip. This is a technically complex procedure that requires considerable



■ Fig. 3.482a, b. Congenital forearm amputation with short forearm stump. Since this patient was unable to wedge objects in her elbow, a *stump lengthening procedure with the Ilizarov apparatus* was performed at 15 years of age. **a** X-rays pre- (top) and postoperatively after extension (bottom). **b** Fitted ring fixator. The mobility in the elbow was preserved, producing a significant functional gain





**Fig. 3.483a, b.** 1-year old boy with thumb aplasia. **a** Preoperatively with 4 long fingers and missing thumb. One year after *pollicization* of the index finger. A powerful pinch grip is now possible

experience. If the outcome is successful, however, the operation is of great benefit to the patient. Much more problematic is the transfer of toes to the hand [5]. The nerve supply is uncertain and, since the toe can always be identified as such, the aesthetic gain is very limited. In the peromelia type of symbrachydactyly, however, a transferred toe can at least provide a grasp function. In individual cases, rotational or wedge osteotomies may be appropriate for correcting abnormal positions of rudimentary fingers.

### Conservative treatment

Conservative treatment consists, on the one hand, of occupational therapy and, on the other, of prosthetic provision of some kind. The aim of the *occupational therapy* is to enable the child to use the malformed hand as much as possible in the most effective way. The child should be taught to use the arm stump as a counter support, wedge objects in the elbow or use any rudimentary fingers as

effectively as possible. Occupational therapy is particularly important in the postoperative phase, for example, if the pinch grip has to be taught after a pollicization procedure.

*Prosthetic provision* may be considered if significant parts of the hand are lacking. Only very simple braces have proved effective, for example those that allow something to be wedged or that can be used as a counter support. Complex prosthesis offering numerous functions are almost never used in practice for unilateral deformities since the unimpaired hand can take over all the relevant tasks with the exception of counter support which, in any case, only requires a simple appliance. As already mentioned in ► chapter 3.5.3.1, myoelectric prostheses are only appropriate for bilateral deformities. Even mechanical prostheses with shoulder traction, elbow movement or a grasp function with a hook are rarely used by patients with a unilateral deformity. For certain occupations, however, a hook may be needed to perform the work, although this factor only comes into play after completion of growth.

### 3.5.3.3 Longitudinal defects (I)

#### **Radial clubhand**

##### **Clinical features, diagnosis**

Radial clubhand is the commonest longitudinal defect, and involves the complete or partial absence of the radius. The clubhand can occur unilaterally or bilaterally. The incidence of radial clubhand is approx. 1:100,000 and is bilateral in around 50 percent of cases. Boys are more frequently affected than girls (1.5:1). The etiology is unknown, and clubhand usually occurs sporadically.

The clubhand may be associated with the following other congenital anomalies:

- thrombocytopenia (TAR syndrome = thrombocytopenia absent radius syndrome): A normal thumb may be present in this syndrome, despite the absence of the radius
- Fanconi syndrome
- Holt-Oram syndrome (hereditary form, associated with atrial septal defect, see ► chapter 4.6.6.4)
- VACTERL syndrome (anomalies of the vertebral column, anal atresia, cardiac anomalies, tracheoesophageal fistula, renal malformation and limb deficiencies).

At birth, the forearm is shortened and the hand deviates in the radial direction. The forearm is around 25–50% shorter than normal. At birth, the flexibility of the elbow may be restricted. However, since this usually improves spontaneously, treatments should not start at too early a stage. Apart from contracture of the elbow, stiffness of the metacarpophalangeal and proximal interphalangeal joints may also be present. Usually the thumb is missing, or else

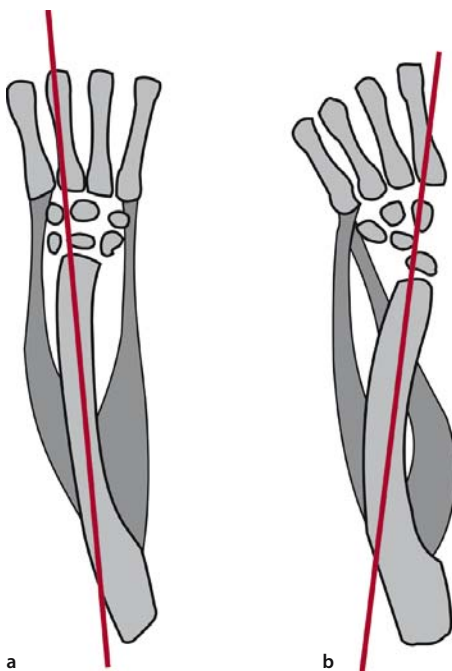
### 3.5.3 · Congenital deformities of the upper extremities

present in only rudimentary form. The 1st metacarpal is always hypoplastic. But the deformity affects not only the bone structures, but also the soft tissues. The flexor pollicis longus muscle is usually missing, the flexor carpi radialis is usually present, but the radial carpal extensors are weak or completely missing. The extensors of the index finger and the middle finger are hypoplastic and frequently have an abnormal insertion. In the event of any reconstruction, it is very important for the surgeon to be aware of the fact that the vessels and nerves also show an abnormal configuration.

#### Treatment

At birth, the hand can usually be corrected from its abnormal ulnar position. Corrective exercises should be initiated by the physiotherapist and the mother at an early stage. While the fitting of a splint can prove very difficult in some cases, nocturnal splint treatment is important and should definitely be administered.

The decision to operate should be made at the age of 6–12 months, particularly if the surgeon intends to perform a pollicization of the index finger at a later date. A very wide variety of operations has already been proposed for stabilization of the wrist. Since the wrist with one bone (ulna) is very unstable, the risk of a recurrence is relatively high for all operations [25]. The centralization of the ulna has proved to be the best solution. An alternative procedure is the radialization of the carpus as proposed by Buck-Gramcko (■ Fig. 3.484) [6].



■ Fig. 3.484a, b. Operations for radial clubhand: a Centralization, b Radialization of the carpus. The radial muscles are transferred to the ulnar side

In the centralization procedure, the ulna is transferred to the middle of the carpus beneath the lunate bone and stabilized with an osteoperiosteal flap, leaving the muscles in place. In the radialization procedure, on the other hand, the ulna is transferred right over to the radial side beneath the navicular bone, while the radial muscles are transferred to the dorsoulnar side of the wrist. Both operations are capable of producing a stable configuration. In two-thirds of cases, pollicization of the index finger is also required, although the configuration of this finger is also not completely normal as a rule. Nevertheless, it is usually possible to produce an opposable thumb with this procedure, which is, of course, particularly important for a bilateral deformity.

#### Ulnar clubhand

##### Clinical features, diagnosis

The ulnar clubhand is roughly ten times rarer than the radial form. Here, too, the forearm bone is completely or partially missing. The defect usually affects the distal two-thirds of the ulna. A fibrous structure is present in most cases and can produce a deforming force by pulling the wrist in the ulnar direction. As the forearm grows, the bowing of the radius increases until it subluxates at the elbow. If the ulna is completely missing, a severe flexion contracture results at the elbow. The ulnar muscles (flexor carpi ulnaris and extensor carpi ulnaris) and the carpal bones on the ulnar side are generally missing, as are the ring and little fingers. In two-thirds of cases, however, additional anomalies of the thumb are present, and these are crucial in functional terms [11].

##### Treatment

If the ulna is merely hypoplastic, treatment is not usually required. Occasionally, the fibrous band must be excised to prevent progression of the ulnar deviation of the wrist. In this operation it is important to spare the malformed ulnar vessels and nerves. Other surgical treatments are rarely indicated. The ulnar deviation can generally be prevented by splints, and the resection of the fibrocartilaginous structure is rarely required. Measures on the fingers are required if additional anomalies exist on the side of the 1st ray and the grasp function is impaired [11].

#### Split hand

Split hand occurs in two forms. The typical form is hereditary, and usually bilateral. The central metacarpals are missing, as are normally the middle finger, occasionally the index finger and rarely the ring finger. The defect is V-shaped. The feet are also often affected. The second atypical form is a symbrachydactyly (of the split hand type), which is unilateral and not genetic in origin. The metacarpus is present, although several central fingers are missing. The defect is U-shaped and the feet are not affected. The typical form is often associated with

other malformations, though not on the extremities (43% [13]). In the typical form the radial side tends to be more affected in severe cases whereas, in symbrachydactyly of the split hand type, the ulnar side tends to be more severely affected. Syndactyly is also frequently present. The treatment is based on the functional impairment. If syndactyly is present, deepening of the web space is indicated. Since the thumb and little finger are present, opposition is usually possible. Rotational osteotomies are also occasionally required. While it may be possible to lengthen rudimentary finger structures in the central part of the hand, few therapeutic procedures are required in most cases, since relatively little functional impairment is present. The cosmetic disfigurement, on the other hand, is relatively pronounced. Since the »lobster-claw hand« is very unsightly, the request for reconstructive procedures is understandable. A narrowing and amelioration of the cleft formation can be helpful in this respect.

### Proximal humeral focal deficiency

By analogy with a proximal femoral deficiency (► Chapter 3.2.7.2), a similar condition can affect the proximal humerus although, to our knowledge, this has not been described in the literature. This involves a defective zone in the proximal humeral metaphysis where it is supplied by the circumflex humeral arteries. As with the femur, this produces a varus deformity of the epiphysis and shortening. The extent of abduction at the shoulder is severely impaired as a result of the varus deformity. ■ Fig. 3.485 shows the restoration of function produced by an osteotomy in a female patient with this lesion.

### 3.5.3.4 Differentiation defects (II)

#### Syndactyly

##### Clinical features, diagnosis

Syndactyly is the commonest congenital deformity of the hand. It is usually inherited, and the family history is positive in up to 40% of cases [29]. In around half of the sufferers the condition is bilateral. 50% of syndactylies are located between the middle and ring finger, 30% between the ring finger and little finger, while other locations are rare [29].

Syndactyly occurs in the following forms [14]:

- Complete: The syndactyly affects the whole web space down to the distal tip of the phalanx.
- Incomplete: A web space between the two fingers exists in the area of the distal phalanges.
- Complex: In addition to webbing of the skin, there is also a bony union.
- Simple: No bony union.
- Acrosyndactyly: syndactyly in the area of the distal, and possibly the middle, phalanx, but with a window at the level of the proximal phalanx.

The more complex the syndactyly, the more likely it is that tendons, nerves and blood vessels will also be involved. Even with simple syndactylies, the nerves divide at a more distal level than normal, and the surgeon should be aware of this fact at operation. Acrosyndactyly often occurs in association with a ring constriction syndrome. Syndactylies can also form part of the Poland syndrome (► Chapter 4.6.6.5), and are also a highly typical feature of Apert syndrome (► Chapter 4.6.6.1). During the clinical examination, in addition to the skin union, we note the mobility of the joints and the length of the bones. Bone shortening is not infrequently seen in cases of brachysyndactyly, in which case the x-ray will also reveal any bony unions.

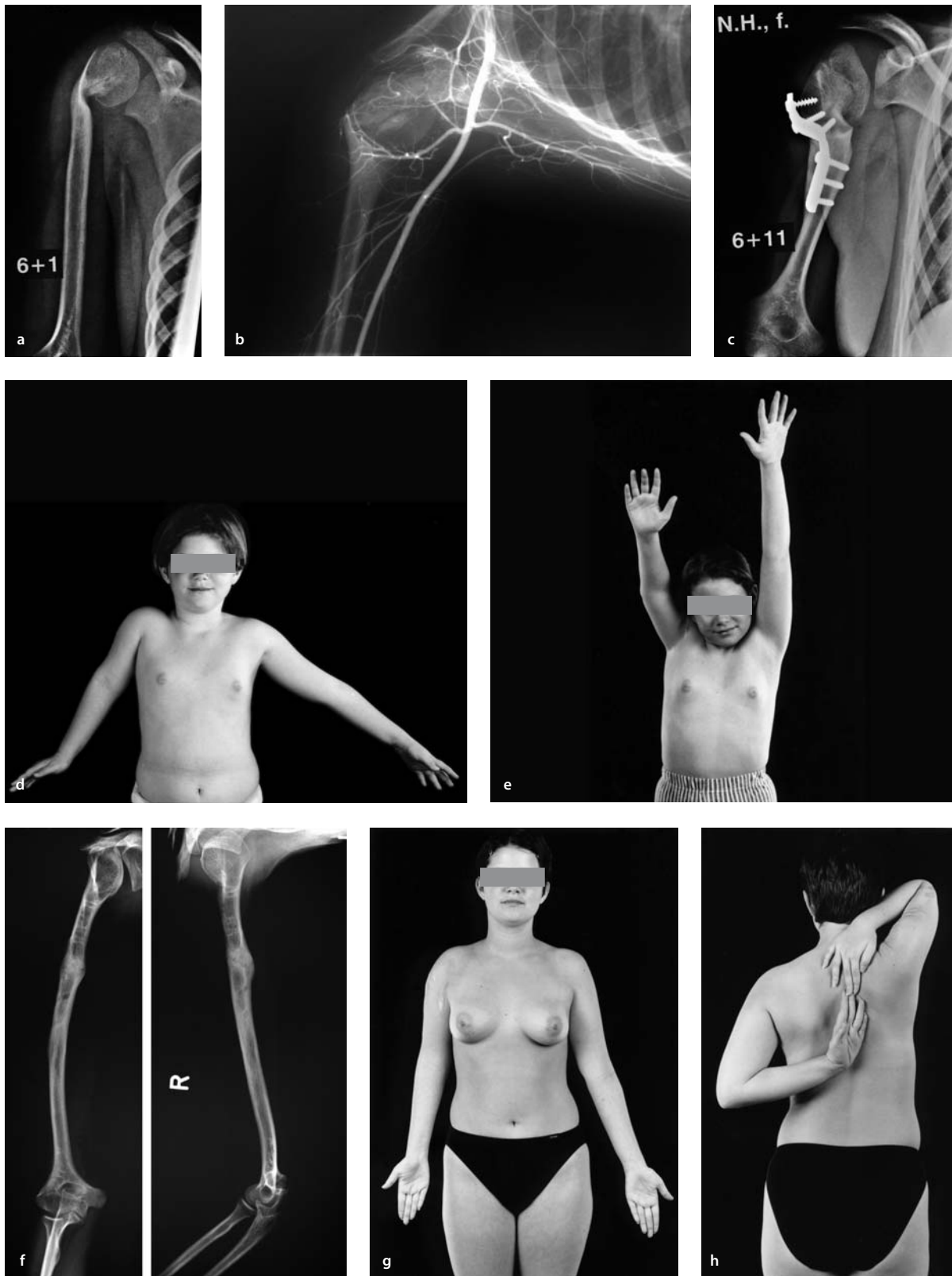
#### Treatment

Very early separation of the fusion is required in cases of acrosyndactyly if the soft tissue connection produces compression or additional deformation of the affected fingers. Early separation is also indicated if the connected fingers are unequal in length. This also applies if a bony union is present. A simple syndactyly can be divided at a slightly later stage, i.e. at the age of 1–1.5 years. Bilateral syndactylies can be operated on during the same session. Note that skin grafting is always required in cases of complete syndactyly. The web space should be formed from local skin. If a joint nail is present, the finger tip on both sides will also require corresponding reconstruction. Under no circumstances should two adjacent syndactylies be separated at the same time, as this would seriously jeopardize the circulation (■ Fig. 3.486).

#### Radioulnar synostosis

In this condition a bony union exists proximally between the radius and ulna (■ Fig. 3.487). The radial head is usually missing. 60% of cases are bilateral [10]. Around 25% of cases are inherited [46]. The forearm is usually fixed in pronation of varying degrees. In around half of the cases, the pronation exceeds 50°. In the past, attempts to separate the radius and ulna and restore mobility have invariably failed [10], although a positive recent report from Japan describes a procedure involving the interposition of a vascularized fat graft [22]. Whether this method will ultimately gain acceptance cannot be determined at this stage, although it seems rather unlikely. On the other hand, derotation osteotomies can be useful, subject to certain conditions. It is much more difficult to compensate for a fixed pronation of the forearm at the shoulder than for a constant supination. But pronation is important for writing, and compensating for the lack of pronation at the shoulder is very tiring as the elbow must constantly be raised. The writing hand should therefore preferably be in a position of slight pronation, while the other hand should ideally be in a neutral position or slight supination [10, 37]. If there is a substantial

## 3.5.3 · Congenital deformities of the upper extremities



**Fig. 3.485a–h.** Proximal humeral focal deficiency with varus deformity of the humeral head. **a** 6-year old girl. **b** The angiogram shows that the defect is in the area of the circumflex humeral artery. **c** 8 months after valgization by 60°. **d** deficient abduction in the right shoulder preoperatively. **e** Abduction almost equal on both sides

2 years postoperatively after valgization osteotomy, but still with clear shortening. **f** AP and lateral x-rays 1 year after lengthening osteotomy. **g** Postoperatively, only slight shortening of the right arm. **h** Good functioning of the extended arm in the neck grip





**Fig. 3.486a, b.** 5-year old boy with bilateral *syndactylies* of the 3rd/4th and 4th/5th digits. **a** Preoperatively before syndactyly separation of the 3rd/4th digits. The 4th/5th digits on the right have already been separated. **b** Postoperatively after separation of 3rd/4th digits on both sides. Syndactylies on adjacent fingers should never be separated during the same operation as this would jeopardize the circulation



**Fig. 3.487.** Radioulnar synostosis in severe pronation in a 6-year old boy

deviation from this position, a derotation osteotomy at the level of the synostosis is worthwhile. The result is ideally stabilized with an external fixator, since the proximal fragment is usually too short for stable anchorage with a plate. The procedure is best performed between the ages of 8 and 10.

### **Camptodactyly**

Camptodactyly involves a congenital flexion deformity of a finger, usually at the proximal interphalangeal joint. It occurs most commonly in the little finger (■ Fig. 3.481). Normally the deformity occurs on both sides and is inherited as an autosomal-dominant condition. Clinical examination reveals a fixed flexion contracture of the affected joint: A passive and active extension deficit is present. This can be observed when the metacarpophalangeal joint is in both the extended and flexed positions. On the x-ray there is recurvation of the distal end of the proximal phalanx, while the joint head is deformed and angulated towards the palm. Splint treatment is indicated for small children. Although the flexion contracture can be surgically corrected by an extending osteotomy or an arthrolysis procedure, it has been clearly shown that the range of motion is not thereby improved. What is gained in extension is lost in flexion. The joint should not be fully extended during the corrective procedure as the extended little finger can also prove troublesome.

### **Clinodactyly**

Clinodactyly involves a deviation of the finger in the frontal, i.e. radioulnar plane. The deformity frequently occurs in connection with syndromes (e.g. in arthrogryposis, ► chapter 4.6.5.1). While the little finger is usually affected, a triphalangeal thumb is also often present. If pronounced angulation is present the condition is described as a delta phalanx. This is the result of abnormal epiphyses, which are rotated around the metaphysis in a C-shape, and is clearly visible on an x-ray. Less pronounced forms of clinodactyly do not require treatment. In the event of marked deviation, an osteotomy can restore the normal anatomical configuration.

### **Tendovaginitis stenisans («trigger finger«)**

Tendovaginitis stenisans almost always affects the thumb and involves a narrowing of the tendon sheath (or pulley) of the flexor pollicis muscle. This produces thickening of the tendon, which can only be drawn through the pulley after overcoming a certain resistance. Weakness or hypoplasia of the extensor pollicis muscle is also frequently present however. A flexion contracture of the metacarpophalangeal joint is also occasionally observed. The condition can be left untreated during the 1st year of life since 30% of the contractures resolve spontaneously. In the other cases, simple surgical opening of the pulley (annular ligament release) will suffice. If a flexion con-



### 3.5.3 · Congenital deformities of the upper extremities

tracture is present at the metacarpophalangeal joint, however, physiotherapy may be needed to stretch the finger. Long-term studies have shown that recurrences almost never occur after an annular ligament release, although a reduction in interphalangeal mobility remains in approx. 20% of cases [30].

#### 3.5.3.5 Polydactyly (III)

##### Occurrence

After syndactyly, polydactyly is the second commonest deformity of the hand. A study in the USA calculated an incidence in the white population of approx. 70:100,000 [44], while a more recent study from Germany found a figure of 33:100,000 [40]. Girls are slightly more frequently affected than boys. The frequency in the black population (particularly of the postaxial form) is roughly ten times that for the white population [44]. A Brazilian study calculated a prevalence of 143:100,000 in a population with a relatively high proportion of black individuals [9]. The duplication of the little finger is usually inherited as an autosomal-recessive condition and is often part of a syndrome. The duplication of the thumb, on the other hand, is not usually hereditary [29], although familial occurrence has been described [12].

##### Classification

The *traditional classification* is as follows:

- *Preaxial*: Duplication on the side of the thumb
- *Central or axial*: Duplication in the area of the 2nd–4th fingers
- *Postaxial*: Duplication on the side of the little finger

The commonest forms of polydactyly are postaxial. Preaxial duplications are slightly less common, while the axial type is extremely rare.

##### Classification according to Blauth [4]

Classification in *two directions*: longitudinal and transverse:

- The transverse axis refers to the affected (1st, 2nd, 3rd, 4th, 5th).
- The longitudinal axis refers to the site of the duplication: distal phalanx, middle phalanx, proximal phalanx, metacarpus, carpus.

##### Classification of radial duplication according to Wassel [43]

■ Table 3.79 shows the classification of radial duplication according to Wassel [43].

##### Clinical features, diagnosis

During the clinical examination we note the size of the supernumerary finger, which is usually smaller than the other fingers (■ Fig. 3.488). It can show a varying de-

viation from the normal axis. Function must be carefully investigated, particularly if there are two phalanges of similar size on the thumb. The mobility may be worse in one of the two partners than the other. The radiographic examination will usually reveal which is the stronger and »more normal« phalanx.

##### Treatment

In types I and II according to Wassel [43], slightly more than half of each thumb can usually be combined to form a single thumb in the Bilhaut-Cloquet operation [2]. The risks of this procedure include the development of a nail deformity and the production of an unintentional epiphysiodesis. In the other types, a complete ray

■ Table 3.79. Classification of radial duplication. (According to Wassel [43])

Type	Characteristic features	Frequency
I	Split distal phalanx	2%
II	Bipartite distal phalanx	15%
III	Split proximal phalanx	6%
IV	Bifid proximal phalanx	43%
V	Split metacarpal	4%
VI	Bifid metacarpal	20%



■ Fig. 3.488. 1-year old girl with *preaxial polydactyly*

is usually resected. In the common type IV the ulnar thumb should normally be left in place and the radial thumb removed. A duplicated 5th ray is generally resected on the ulnar side. Central duplications are rare and frequently associated with a syndactyly. This can conceal the actual polydactyly, in which case an accurate diagnosis may occasionally be reached only after an x-ray has been recorded. The supernumerary phalanx is usually resected, taking care to preserve the tendons and nerves. Sometimes a deviation of the »normal« phalanges has to be corrected.

### 3.5.3.6 Macroductyly (IV)

Macroductyly is a non-inherited congenital enlargement of a finger. In 90% of cases, the condition is unilateral. If more than one finger is affected, the two affected fingers are always supplied by the same nerve (normally by the median nerve). The cause of the macroductyly is usually neurogenic. Occasionally a neurofibromatosis is also present (see ► chapter 4.6.6.2). Tendons and blood vessels are configured normally. Evidently, hyperplasias of the fingers and toes do not have the same origin [21]. Macroductyly occurs in a static form that is present at birth and in a progressive form that is usually only detected at a later stage during the first two years of life. In this form the difference in size increases gradually over this period. The diagnosis is based on clinical examination. The difference in length should be measured and the function carefully investigated. Radiological documentation is also important. Treatment either involves partial amputation of the finger or a reduction in its size, although the latter procedure is extremely difficult and plagued by complications, particularly if soft tissue anomalies are present (neurofibromatosis). An elegant solution is epiphysiodesis at the time the toe roughly reaches its adult size, combined with a soft tissue reduction with preservation of the neurovascular bundle.

### 3.5.3.7 Hypoplasia (V)

#### Thumb hypoplasia

The hypoplasia almost always affects the 1st ray. This deformity has been classified by *Blauth* as follows [3]:

- Grade I: Minimal hypoplasia, all elements present
- Grade II: Adduction contracture in the 1st web space, instability of the ulnar collateral ligament, hypoplasia of the thenar muscles, normal skeleton on the x-ray
- Grade III: Significant hypoplasia with aplasia of the intrinsic muscles, only rudimentary extrinsic tendons, hypoplasia of the bones
- Grade IV: »Floating thumb«: Uncontrollable stump attached at the metacarpophalangeal joint of the index finger
- Grade V: Complete absence of the thumb

The condition can be classified on the basis of the clinical examination. Active mobility in particular should be investigated. The x-ray will also reveal the absence or presence of any hypoplasia of the bony structures.

The treatment is based on the degree of the deformity:

- Grade I: No treatment required
- Grade II: Release and deepening of the 1st web space, opponensplasty, stabilization of the metacarpophalangeal joint, exploration of the flexor and extensor tendons and possible correction of anomalies. The opponensplasty uses the ring finger flexor digitorum superficialis or abductor digiti minimi muscles
- Grade III–V: Pollicization of the index finger

#### Madelung deformity

Madelung deformity is an autosomal-dominant inherited disorder involving the inadequate development of the epiphyseal plate of the distal radius. The anomaly is not yet visible at birth, and the diagnosis is usually not made until the ages of 8–12 years, i.e. when the impaired growth becomes apparent.

The abnormality involves the following elements:

- Radius: Shortening, increased inclination of the joint surface in the ulnar direction of more than 20°
- Ulna: Dorsal subluxation, enlargement of the ulnar head
- Carpus: Wedge-shaped deformity. A fibrocartilaginous ligament known as Vickers ligament extends between the radius and the ulnar section of the carpus (■ Fig. 3.489) [42].

The prominence of the ulnar head is a particularly striking clinical finding. The mobility of the wrist is reduced, with impairment of dorsal extension, radial abduction and supination in particular. Pain is also present occasionally. Surgical treatment may be indicated if symptoms exist. This involves resection of the Vickers ligament combined with a wedge osteotomy of the radius and possibly a correction on the ulnar side.

### 3.5.3.8 Ring constriction syndrome (congenital band syndrome) (VI)

This syndrome occurs sporadically and no hereditary factor has been shown to date. The cause has been the subject of considerable dispute and has still not been satisfactorily explained. The pregnancy history is often characterized by complications [16].

Patterson has proposed the following useful *classification* for use in everyday clinical practice [35]:

1. Simple ring constrictions
2. Ring constrictions accompanied by distal deformities (with or without lymphedema)

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3. Ring constrictions associated with distal fusion – Acrosyndactyly
4. Amputations

Ring constriction syndromes may require urgent treatment, particularly if pronounced edema is present. If the circulation and the nerve supply are jeopardized, immediate surgical release is required. Failure to implement this procedure in time may lead to necrosis of the distal part of the finger and subsequent amputation. If no significant edema is present, treatment can be carefully planned. For simple ring constrictions, Z-plasties in series are usually required to relieve the soft tissues compressed by the skin constriction. This particularly applies in the event of distal deformities. Acrosyndactylies are generally divided during the first six months of life. Measures are also occasionally required to improve the circulation. While skin problems often occur later on as a result of problems with sensation or circulation, these are more frequently observed in the foot than the hand. Partial aplasia of the thumb is sometimes encountered, and may require phalangization or possibly even pollicization.



■ Fig. 3.489. MRI scan of a 14-year old girl with *Madelung deformity*. The *Vicker's ligament* can be seen between the radius and lunate bone. Individual fibers also extend to the triquetrum bone. The ligament displaces the ulna in a dorsal direction

#### 3.5.3.9 Sprengel's deformity

##### ► Definition

Congenital deformity of the scapula with inadequate descent of this bone from the cervical to the thoracic area during the third month of pregnancy.

##### Historical background

This change was first mentioned by Eulenberg in 1863. [15]. Sprengel subsequently described four cases in 1891 [41].

##### Etiology, pathogenesis, occurrence

The condition usually occurs sporadically, although a familial occurrence has been observed. Girls are more frequently affected than boys. It is currently assumed that the Sprengel deformity is connected with an anomaly in the formation of the cerebral ventricles during early pregnancy, as a result of which cerebrospinal fluid leaks into the subcutaneous tissues. The deformity is associated with other malformations in 75 percent of cases, e.g. anomalies of the cervical spine (Klippel-Feil syndrome, see ► chapter 3.1.7) [20]. The scapula is initially formed at the level of the 5th cervical vertebra and normally migrates downward during the 3rd month of pregnancy. This migration fails to occur in a Sprengel's deformity. The scapula remains too high and too small. A bony connection also occasionally exists between the scapula and the spine (omovertebral bone).

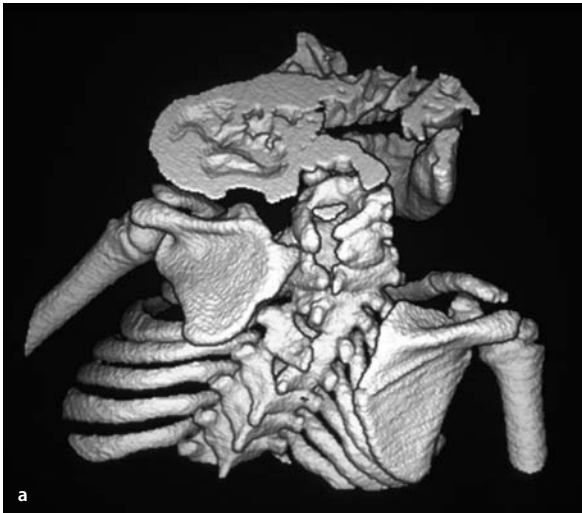
##### Clinical features, diagnosis

The scapula is too high on one side (possibly as much as 10 cm higher than the shoulder blade on the other side), smaller than normal and usually externally rotated. Clinical examination reveals a restricted abduction (frequently less than 90°) in particular. The neck muscles are shortened on the affected side, and an omovertebral bone is sometimes palpable. The *x-ray* shows the elevated and rotated scapula. The omovertebral bone can be identified on axial views of the scapula. A CT scan is useful preoperatively for showing the bone relationships (■ Fig. 3.490).

##### Treatment

Surgical correction is indicated, if possible between the ages of 4 and 6 years, if abduction is significantly restricted. Whereas, in the past, we have used the procedure described by Woodward [45], nowadays we prefer a modified form of the operation specified by Green [19, 28]. In this procedure the patient is placed in the lateral position and the clavicles are first osteotomied anteriorly. Then, via a posterior approach, the lateral trapezius is detached, the deep muscles (rhomboid and levator scapulae muscles) are detached directly at the scapula and any omovertebral bone is resected. The scapula is then transferred distally and anchored to the ribs in a pocket under the latissimus

dorsi muscle. The detached muscles are then refixed to the scapula at a more cranial point. This operation can improve the abductability of the arm by 50° on average, and the cosmetic results are also very satisfactory. Positive results are also reported with a vertical scapular osteotomy [33].



■ **Fig. 3.490a, b.** Sprengel's deformity in a 4-year old girl. **a** CT scan of the shoulder region. The left scapula stands 6 cm higher than the right one, is smaller and is in contact with the posterior part of the skull. Multiple anomalies of the cervical spine also exist. **b** Clinical picture

### 3.5.3.10 Congenital clavicular pseudarthrosis

An awareness of this rare anomaly is important for the differentiation from the clavicular fracture that occurs as a result of birth trauma. In contrast with a fresh fracture, the edges of the pseudarthrosis are rounded (■ Fig. 3.491). The etiology of the congenital pseudarthrosis is not known. Isolated familial cases have been described [36]. In contrast with congenital tibial pseudarthrosis, there is no association with neurofibromatosis. The lesion is almost always on the right side [8].

The differential diagnosis must consider the possibility of a cleidocranial dysostosis (▶ Chapter 4.6.2.19). A striking clinical finding at the age of 2–3 years is a painless swelling in the area of the clavicles, combined with asymmetry of the shoulders. In order to prevent worsening of the deformity, surgical correction with a wide resection of the pseudarthrotic tissue, including the periosteum, cancellous bone graft and stable fixation, should be performed around the age of 5 years [8, 24].

### 3.5.3.11 Congenital radial head dislocation

Congenital forms of radial head dislocation are rarer than overlooked forms after Monteggia fractures (▶ Chapter 3.5.7.12). The lack of a trauma history does not necessarily rule out a traumatic cause of the dislocation. Factors suggestive of a congenital form include bilateral occurrence, excessively long radius, convex instead of concave shape of the proximal surface of the radial joint and the lack of any deformation of the ulnar shaft. Under no circumstances should an attempt be made to reduce the radial head in the congenital form. The symptoms and functional restriction are usually minimal in this form, although the excessively long radius may cause problems and can be treated by resection of the head on completion of growth.



■ **Fig. 3.491.** X-ray of the clavicle in a 2-year old boy with congenital pseudarthrosis in the center of the shaft



### 3.5.3.12 Generalized skeletal abnormalities (VII) (Apert and Poland syndromes, Arthrogryposis etc.)

Hand deformities occur in certain syndromes, typically in Apert, Poland or Klippel-Trenaunay-Weber syndromes and in arthrogryposis. The *Apert syndrome* is discussed in detail in ► chapters 4.6.3.1 and 3.4.5.12. This is a rare inherited disorder that involves, in particular, syndactylies on the hands, often with bony involvement (► Fig. 3.492).

The treatment consists of separation of the syndactylies and bony connections, although one should not be deluded into thinking that this will significantly improve mobility. In the Apert syndrome, the hand usually forms a rigid plate that can only be used as a whole, rather like a shovel. Operation of the individual fingers is therefore not possible. Apart from the hand deformities, children with Apert syndrome are additionally handicapped in the upper extremities by movement restrictions at the elbow and shoulder joints [23].

Patients with *Poland syndrome* suffer much less impairment. This non-inherited condition is described in ► chapter 4.6.6.10 and involves aplasia of the pectoral muscles and syndactylies, occasionally with missing middle phalanges (symbrachydactyly) [9]. These deformities are treated as described above. Function is not usually significantly impaired.



► Fig. 3.492. AP x-ray of the right hand in a 1-year old girl with *Apert syndrome*. Bony connections exist between all the rays

Similarly syndactylies, in some cases with bony connections, also occur in *arthrogryposis* (► Chapter 4.6.5.1). As these are additionally associated with flexion contractures, the prognosis is less favorable than for Poland syndrome. A wide variety of hand deformities are also frequently observed in *Klippel-Trenaunay-Weber syndrome* [32].

*I should like to thank Dr. Beat Simmen and Dr. Renato Fricker, specialists in hand surgery at our hospital, for the critical perusal of this chapter and their many suggestions.*

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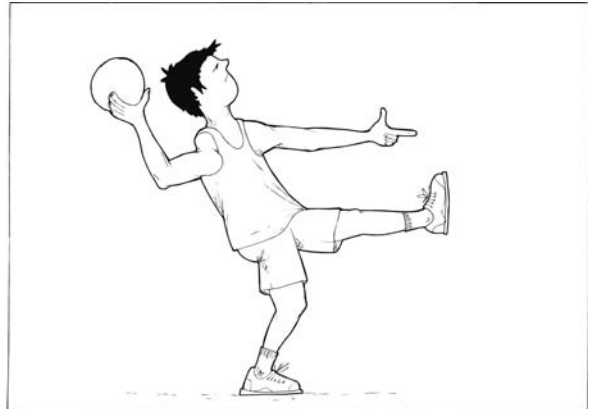


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### 3.5.4 Dislocations of the shoulder

#### Definition

Dislocation of the humeral head from the glenoid in an anterior, posterior or inferior direction.  
Synonym: Dislocatio glenohumeralis



#### Classification

We distinguish between the following types:

- Acute traumatic shoulder dislocation: Dislocation of the humeral head, usually in a ventral or ventral/caudal direction, caused by adequate trauma.
- Acute constitutional shoulder dislocation: First dislocation of the shoulder in the presence of predisposing factors.
- Recurrent shoulder dislocation: Shoulder dislocations that occur repeatedly and with increasing frequency. These can be anterior, inferior or posterior and also frequently occur in different directions in succession (multidirectional instability).
- Habitual or voluntary shoulder dislocation: The humeral head can be dislocated in an anterior or posterior direction at will by muscle activity.
- Congenital shoulder dislocation: The humeral head is dislocated at birth as a result of a formation defect and cannot be reduced.
- Shoulder dislocation due to birth trauma: This occurs as a result of injury caused at birth from a breech presentation and is often associated with plexus palsy.
- Neuromuscular shoulder dislocation: Shoulder dislocation produced by abnormal muscle forces, particularly of the latissimus dorsi muscle.
- Iatrogenic shoulder dislocation: Dislocation occurring after an operation in the opposite direction to the direction corrected during surgery.

#### Etiology

While the classification indicates that the etiology is not uniform, predisposing factors play a significant role in

### 3.5.4 · Dislocations of the shoulder

most of the most common forms (traumatic, recurrent, habitual / voluntary dislocations). This also applies to iatrogenic dislocation. The etiology for these forms will be discussed together, but first the special forms not included in this category will be addressed briefly.

#### Congenital shoulder dislocation

This form is extremely rare. The humeral head is permanently dislocated anteriorly at birth. The cause is agenesis of the anterior joint capsule.

#### Shoulder dislocation due to birth trauma

This form has also become extremely rare, since a natural delivery from a breech presentation is now performed only in isolated cases. The dislocation is usually in a posterior direction and a plexus palsy is often present at the same time.

#### Neuromuscular shoulder dislocation

This form is also very rare. Dislocation (usually posterior) can occur in a patient with a hemiparesis or spastic tetraparesis as a result of abnormal muscle activity. A caudal dislocation is generally observed in flaccid paralyses, for example in a patient with a lesion of the axillary nerve (e.g. after a tumor resection).

#### Traumatic and constitutional shoulder dislocation

This is the most important group of shoulder dislocations. The etiology of traumatic and constitutional shoulder dislocations will be addressed jointly since constitutional factors usually play a role in adolescents even in the presence of adequate trauma. This presupposes that recurrences occur much more frequently in adolescents than in adults [12]. Often the opposite side will also dislocate at a later stage following a traumatic dislocation [11]. The distinction between a traumatic dislocation with and without predisposing factors is very difficult. Recurrences can occur at a later date even after a genuine traumatic dislocation.

The cause can be found in two lesions that arise at the first dislocation:

- Lesions of the anterior glenoid rim: Small shell-shaped tears (*Bankart lesion* [1]) or large shear fragments of the socket.
- Indentations on the edge of the humeral head: These are the result of pressure exerted by the glenoid rim in the dislocated state. In a case of anterior instability the indentation is usually located on the posterior superior side of the humeral head (*Hill-Sachs lesion*). Moreover, an anterior dislocation can often be accompanied by tearing of the ligaments with the glenoid labrum, even without bone fragments.

The following *constitutional predisposing factors* also apply [4]:

- Abnormal head-socket relationship: Relatively small socket with large humeral head.
- Incorrect ratio of the bone curvature radii for the head and socket (primarily an excessively large curvature radius for the socket).
- Abnormal inclination of the socket in relation to the shoulder blade (no physiological retrotorsion of approx. 5°).
- Torsional defects of the humeral head: deviation from physiological retrotorsion of 30°–40°.
- Abnormal laxity of the joint capsule: General ligament laxity or systemic connective tissue disorder, e.g. as in Ehlers-Danlos or Marfan syndromes (see ► chapter 4.6.3).

Apart from shoulder dislocations, there is also the problem of unintentional positional instability of the shoulder in sporting adolescents with lax ligaments, which can result in symptoms [16].

#### Occurrence

An epidemiological study in Minnesota/USA calculated an incidence for an initial traumatic shoulder dislocation of 8.2:100,000 persons/year, while that for all shoulder dislocations (including recurrences) was 11.2:100,000/year. This rate was significantly higher in adolescents than in adults [3].

#### Clinical features, diagnosis

##### Acute shoulder dislocation

With an initial shoulder dislocation it is usually difficult to establish whether predisposing factors are present or not. Careful history-taking is therefore required to determine whether an abnormal trauma producing substantial deflection has actually occurred or whether the dislocation has arisen in connection a simple abduction-external rotation movement. An anterior shoulder dislocation is predominantly caused by this movement direction, whereas posterior dislocation is provoked by abduction, forward elevation and internal rotation. If the dislocation can be reduced spontaneously, it must be assumed that predisposing factors play a significant role. On the other hand, if the dislocation cannot be reduced without medical assistance, the trauma is probably the predominant factor.

The clinical diagnosis of an acute dislocation is not difficult. Pain and abnormal shoulder contours indicate the diagnosis. The AP x-ray of the shoulder, and also the Y-view (► Chapter 3.5.2), show the dislocation and its direction. The humeral head is always in a caudal position, regardless of whether the dislocation is in an anterior or posterior direction. As a result, the dislocation is always clearly seen on the AP view. The situation is even clearer on the Y-view, which shows the glenoid from above. Additional imaging procedures are not indicated for the acute form.

### Recurrent and habitual shoulder dislocation

During the history-taking, the patient should be asked about the frequency of the dislocations and the intervals between the dislocations. The direction of movement at the time of the dislocation should be established. The pain history must also be recorded. The shoulder should then be examined meticulously (► Chapter 3.5.1).

The examination for recurrent or voluntary shoulder dislocations must include the following:

- palpation,
- investigation of the range of motion,
- glenohumeral translation,
- »apprehension test«.

During the *palpation* we look for painful sites in the area of the anterior or posterior joint capsule. The *range of motion* must be investigated carefully so as to avoid provoking any further dislocation. In particular, concurrent abduction, external rotation and extension must be avoided if an anterior dislocation is present. If *glenohumeral translation* is present, the subluxability of the humeral head is tested. The aim of the *apprehension test* is to provoke the patient's sensation *before* dislocation by reproducing the dislocation event (both tests are described in ► chapter 3.5.1). The clinical examination can be supplemented by a *CT scan*. This investigation is ideal for evaluating the head-socket relationship, the ratio of the bone curvature radii for the head and socket, the shape of the cartilaginous socket and its inclination in relation to the shoulder blade, as well as any torsional defects of the humeral head. The Bankart lesion and the Hill-Sachs groove can also readily be assessed on the CT scan. Since MRI scans do not provide much additional information they are not usually required.

*Diagnostic arthroscopy* is by far the best method for identifying capsuloligamentous lesions. However, since it is an invasive investigation it should be performed only if surgery is already indicated, which is very rarely the case in children and adolescents.

### Course, prognosis

We must assume that the majority of both »traumatic« and non-traumatic shoulder dislocations that occur during adolescence involve predisposing factors. This is based partly on the bone configuration and partly on a constitutional ligament laxity. Since the collagenous tissue steadily shrinks in individuals with lax ligaments, this phenomenon has a positive influence on the course of the condition. It is important, however, that the ligaments should not be repeatedly overstretched. Voluntary dislocation, in particular, must be avoided. One long-term study that followed up 18 untreated children with voluntary shoulder dislocations over an observation period of 12 years revealed a good, problem-free status in 16 cases, while surgery was required in only 2 cases. By contrast, in 7 patients with a similar initial situation who underwent

surgery at the outset, the status was good in only 3 cases, the situation was still unstable in 2 cases and painful in 2 cases, and the shoulder was stiff in one patient [8].

### Treatment

#### Conservative treatment

As may be concluded from the above statements, the decision to operate should be made with extreme caution. It should, in fact, be considered only for a recurrent dislocation that was originally clearly traumatic and whose corresponding lesions (Bankart lesion, Hill-Sachs groove) are identifiable on the CT or MRI scan. If such lesions are not present and if the initial trauma is doubtful, an operation should be avoided. Much more important is conservative treatment with consistent muscle training. The »San Antonio training program« is particularly suitable for this purpose (■ Fig. 3.493) [2]. Various reports have confirmed the positive effect of this kind of exercise program, even in anatomical hypoplasia of the glenoid [2, 13], in contrast with the situation produced by immobilization [9].

The most important measure is to persuade the young patient to stop practicing sports that involve arm movements above head height (tennis, baseball, basketball, volleyball, gymnastics etc.). One study has shown that the chances of recovery are almost nine times higher in those who avoid such sports than in those who continue training in them [6]. The particular problem of unintentional positional instability of the shoulder in sporting adolescents with lax ligaments (see above [16]) can be countered by avoiding certain positions and performing muscle-strengthening exercises.

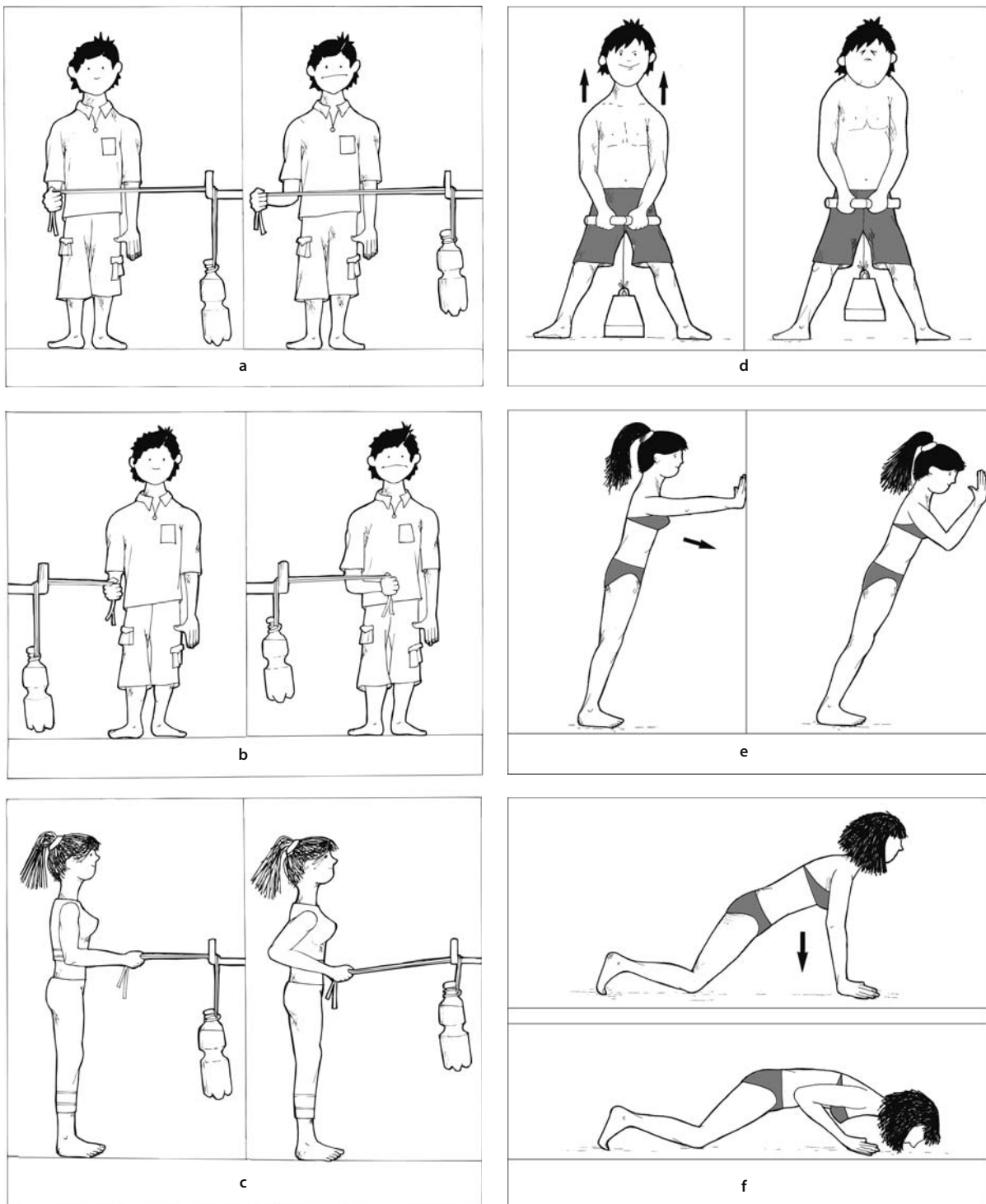
#### Surgical treatment

Possible surgical procedures include the following:

- refixation of the Bankart lesion (open or arthroscopically)
- capsular shrinkage (by conventional means or with the laser device)
- glenoid reconstruction in dysplasia
- overlapping of the subscapularis muscle (Putti-Platt operation)
- anterior or posterior strengthening of the glenoid with a bone graft
- rotational osteotomy of the coracoid (Trillat operation)
- rotational osteotomy of the humerus

One deciding factor for treatment is the presence or absence of a Bankart lesion. The lesion is present in 80% of traumatic dislocations [14] and nowadays tends to be refixed arthroscopically with »anchors« (e.g. the Mitek anchor). The repair of the Bankart lesion produces good results in children and adolescents in a high percentage of cases [7, 14, 15]. The combination of refixation of the Bankart lesion with capsuloplasty can produce satisfactory results even if multidirectional instability is present.

## 3.5.4 · Dislocations of the shoulder



**Fig. 3.493a–f.** Exercises from the San Antonio muscle training program for shoulder instability [2]. **a** The child stands sideways next to the wall. A weighted object (e.g. a full plastic bottle) is fastened to a cord which is then suspended from a door handle. The child holds the cord with the upper arm hanging down and the elbow flexed at 90° (left) and then pulls on the cord by rotating the arm outwardly at the shoulder (right). **b** In a position similar to **a** (left) the cord is pulled with inward rotation at the shoulder (right). **c** The child stands in front of the wall with the fastened cord and holds it with the upper arm hanging down and the elbow flexed at 90° (left). The cord is pulled back by elevation

at the shoulder (right). **d** The child stands with legs slightly apart and holds a cord to which a weight of approx. 2 kg has been fastened (left). By raising the shoulders (and not by flexing the arms), the child pulls the weight off the floor (right). **e** The child leans against the wall with arms extended at a slight angle of 20–30° (left). She moves her upper body closer to the wall by flexing the arm at the elbow (right) and then pushes it back by extension. **f** The child kneels on the floor, supports herself with both arms and raises and lowers her upper body by flexion (left) and extension (right) at the elbow



Reconstruction of the glenoid can prove useful in certain cases of habitual shoulder dislocation with dysplasia of the glenoid [17], although this procedure should only be performed by a very experienced shoulder surgeon.

Most of the other operations are associated with, in some cases serious, drawbacks. The overlapping of the subscapularis muscle and anterior capsular shrinkage result in a restriction of external rotation [8]. A posterior dislocation can occur after bone grafts or a rotational osteotomy of the coracoid according to Trillat. Furthermore, rotational osteotomy of the humerus is now an obsolete procedure. Basically, one should attempt to reconstruct the disrupted anatomy rather than create a new pathology by performing procedures outside the actual lesion.

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## 3.5.5 Growth disturbances of the upper extremities

### Definition

Conditions involving disorders of the bones and joints of the upper extremities that occur in growing children and adolescents, generally in connection with overexertion.

#### 3.5.5.1 Panner's disease

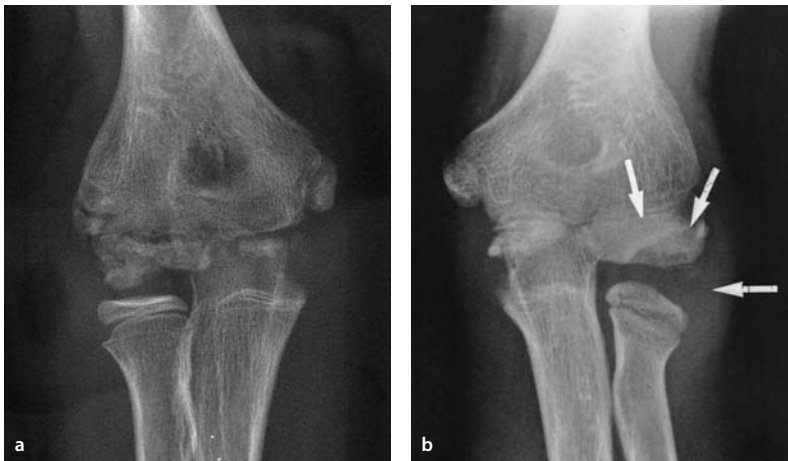
In 1927, Panner described a lesion of the capitellum resembling that of Legg-Calvé-Perthes disease [5]. The term »Panner's disease« was then coined by Smith in 1964 [6]. This condition affects children under 10 years of age with pain and swelling in the elbow area. There is no history of trauma or overexertion. The x-ray shows irregular areas in the vicinity of the capitellum with central brightening, possibly surrounded by sclerosis (■ Fig. 3.494a). The joint cartilage is not affected by the condition. The pathogenic mechanism is probably similar to that of other forms of aseptic bone necrosis (Legg-Calvé-Perthes disease, Freiberg's disease). Provided no fragment has dissected, the prognosis of the disease is good. Analgesics and temporary splinting are the most useful measures. If a fragment threatens to break off (which is very rare in this age group), it should be refixed (if possible with a resorbable polylactate screw) or (if this is not possible) resected.

#### 3.5.5.2 Osteochondritis dissecans of the capitellum

Osteochondritis dissecans of the capitellum occurs in older children and adolescents and is associated with chronic trauma. A lateral compression mechanism [4] is usually involved, and this arises predominantly in athletic events, gymnastics or throwing disciplines. Signs and symptoms include pain, swelling and movement restriction. The x-ray shows changes similar to those of Panner's disease, although the lesion is usually better demarcated (■ Fig. 3.494b) and dissection is common. The dissected fragment itself is not usually visible on the x-ray, although secondary changes of the radial head can occur as a result of the resulting pressure. An MRI or CT scan will usually reveal the fragment.

Provided no fragment has broken off, the treatment can remain conservative (with analgesics and immobilizing splint). However, a ban on the triggering sport should be imposed as a rule, since the consequences of the dissection can be very serious. Sporting youngsters often find this difficult to accept. If dissection threatens, or has already occurred, refixation should be attempted (e.g. with a resorbable polylactate screw). If this is not possible the fragment must be resected, although this generally has adverse consequences for elbow function.





**Fig. 3.494** a) AP x-ray of the elbow of a 9-year old boy with Panter's disease. Clinically, there is pain and swelling, while the x-ray shows irregular ossification of the capitulum and, in places, of the ulnar condyle. b) AP x-ray of the elbow of a 13-year old athletically active boy (gymnastics) with osteochondritis dissecans in the area of the capitulum. The longstanding dissected fragment has led to deformation of the radial head.

### 3.5.5.3 Lunatomalacia

As with necrosis of the radial condyle, the rare condition of lunatomalacia can occur in children under 10 years of age spontaneously with no history of overexertion [2]. In adolescence, however, this disease, which is not all that rare in adults, is typically the result of chronic overexertion [1, 3, 7]. This may be observed in gymnasts, but also, for example, in children on rollerblades who regularly crash into a wall, or in those who often chop wood. The x-ray often reveals minor changes only, while the MRI scan shows distinct edema of the lunate bone, and often definite necrosis with collapsing of the bone structures. The first therapeutic measure is the imposition of a ban on the triggering activity, followed by the administration of analgesics and immobilization of the wrist with a splint. If no improvement has occurred after months of suffering, a graduated radial shortening osteotomy is occasionally indicated [1, 3, 7].

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### 3.5.6 Neuromuscular disorders of the upper extremity

*R. Brunner*

Whereas the functions of the lower extremity can be clearly and simply defined in terms of standing, walking and sitting, the use of the upper extremity – particularly the hand – is far more complex. During development, the upper extremity changes from a support organ, like the lower limb, into a functional organ. In addition to the numerous discriminating motor tasks, it also fulfils the function of a sensory organ. Depending on the particular clinical picture, children with neurological disorders not infrequently experience an impairment of both functions. But without an intact sensory system the hand is not really usable. Since the condition is already present at birth in many cases, the patients have never been able to familiarize themselves with a normal situation. Despite their functional impairment, they attempt to perform the same activities with their two hands as unimpaired children of the same age and soon get into the habit of employing what can be highly efficient compensatory mechanisms.

Consequently, before any treatment the orthopaedist must accurately establish to what extent the children are handicapped by their neurological disorder and the associated functional problems and deformities, identify the compensation mechanisms that are already being employed and the extent to which their situation can be improved by treatment. However, deformities of the upper extremity are also invariably of cosmetic and social significance, with the corresponding psychological impact. Since the treatment of the upper extremity must take all these factors into consideration, a careful evaluation of the disorder must incorporate the cosmetic expectations of the patients and their mental attitude.

The motor aspects of the *functional investigation* include mobility in the shoulder, elbow and wrist, contractures of the fingers, the grasp function, the pinch grip and opposition of the thumb in relation to the fingers. The sensory aspects must also be tested. Apart from the various types of sensory perception (superficial and deep sensation, pain and temperature sensation), two-point discrimination and the hyperaesthesia must be examined. The expectations of the patient in respect of the treatment must be explained and discussed before any therapeutic measures are implemented in order to avoid subsequent disappointment and dissatisfaction. A tendon transfer that produces an ideal functional result can be disappointing for the patient if cosmetic aspects were the most important factor for him or her and these had received insufficient consideration.

### 3.5.6.1 Primarily spastic paralyzes

#### ➤ Definition

Functional disorders and deformities of the upper extremity caused by spastic muscle activity.

#### Etiology and pathogenesis

Spastic cerebral palsies most often result in disorders of the upper extremities with spasticity. Both arms are affected in tetraparesis, while the arm on the weakened side is affected in hemiparesis. In severe cases of diparesis and hemiparesis, even the supposedly unaffected extremities usually show slight functional problems. Rare spastic functional disorders with a spinal origin also occur, for example as a result of tetraplegia after accidents, malformations of the spinal cord and column or tumors.

#### Clinical features and diagnosis

The patients may show impairment of motor, sensory or autonomic function and global perception. From the motor standpoint, there is adduction spasticity at the shoulder with concurrent slight flexion. Other patients may show the opposite: abduction and extension. At the elbow and the wrist the spasticity affects the flexors, pulling the elbow into flexion and the wrist into a position of palmar flexion-pronation and ulnar deviation (■ Fig. 3.495).

The spastic muscles tend to be very susceptible to contractures, which restrict the usability of the extremity and, in severe cases, nursing care as well. Function is also impaired by co-contractions, which usually manifest themselves as concurrent palmar flexion when the fingers are closed. As a result of the unfavorable position at the wrist, the strength of the finger flexors is reduced, thus hampering the coordinated use of the fingers. The spastic activity of the flexor carpi ulnaris muscles draws the hand and fingers out of the field of view, thereby preventing visual control and compensation for any existing sensory impairment.



■ Fig. 3.495. Typical hand position in *spastic hemiparesis*

Another typical deformity that can affect the fingers involves flexion at the interphalangeal joints with hyperextension at the metacarpophalangeal joint (swan-neck deformity). The deviating thumb can also prove troublesome and involves the added risk of dislocation at the metacarpophalangeal joint if it is placed in abduction without corresponding support (■ Fig. 3.496).

Impaired sensory perception of varying severity is almost always present at the same time, manifesting itself as hypoesthesia, paresthesia or hyperesthesia. Exaggerated sensation, in particular, is subjectively unpleasant for the patient, who may stop using the affected arm solely for this reason. But in all cases the sensory impairment interferes with the touch function of the hand, which is obviously needed for normal everyday use. The deficit may, at least in part, be compensated by visual control, although the spastic muscle activity may prevent this as it draws the hand out of the visual field by pronating and flexing the wrist. Autonomic signs and symptoms are not infrequently present in addition to the motor and sensory impairments. The hands are moist and cool, and tend to sweat more than normal. These problems are unpleasant for the patient, particularly if the right hand, i.e. the one used for shaking hands with others, is affected.

As a result of the constant underuse of the hand, the patients develop compensatory mechanisms and tend to use the unaffected hand to a much greater extent as this enables them to perform numerous tasks faster and more securely and efficiently. Explanations and exhortations to use the affected extremity as well do little to change this fact. Quality and speed are required in everyday life. Only by creating the need for bimanual operation can function be improved through training.

❗ **If sensation and central nervous representation of the arm are not present, even a functional gain for the patient will be of no help in everyday tasks.**

Since the functions of the upper extremity are complex, and motor and sensory function are interlinked to a much greater extent than with the lower extremity, all functions



■ Fig. 3.496a, b. *Swan-neck deformity* (hyperextension at the thumb metacarpophalangeal joint) with the risk of dislocation of the metacarpophalangeal joint. **a** Abduction with dislocated position in the

metacarpophalangeal joint. **b** Supported abduction without dislocation at the metacarpophalangeal joint

must be investigated. Only a comprehensive analysis can help avoid therapeutic failures. On the other hand, even minor gains in function can improve a patient's quality of life. Muscle activity can be investigated by electromyography. Since the activation pattern usually persists even after a muscle has been transferred, EMG can also provide clues to the future functioning of the muscles and the functional effects.

### Treatment and prognosis

#### Conservative treatment

The aim of *occupational therapy* is to improve the overall function of the extremity. The treatment includes motor training, contracture prophylaxis and the promotion of the coordination of muscle activity. In addition to the motor functions, sensory perception is also crucial, particularly for the upper extremity. Accordingly, hyperesthesias or hypesthesias must be corrected as far as possible by corresponding sensory training. The occupational therapist is also responsible for adapting and testing various braces, including functional orthoses and braces.

Given the complex functions of the upper limb, it is difficult to achieve any improvement with a *functional orthosis*. While an orthosis for the foot can control an instability and improve walking by providing added security, an instability or deformity of the hand is not the sole crucial factor for function. For example, while the strength of the finger flexors and thus the grasp function may be poor without splinting, hand braces restrict sensory function and thus interfere with its role as an organ of touch. Orthoses therefore have to be regularly tested and their use adapted to everyday requirements, even function will still be restricted. Compromises are required in the use of braces and, in many cases, the only appropriate solution is a brace for preventing any exacerbation of contractures (■ Fig. 3.497).



■ Fig. 3.497. *Brace for the hand*

*Cast treatments* can also be used successfully for managing contractures of the upper limb. However, since such casts tend to be less well tolerated, in our experience, than lower-limb casts, they are often worn for only a few days [21].

The plastering of the unaffected hand in hemiparetics in order to encourage the use of the other hand has recently been rediscovered. This treatment forces the patient to use his »poor« hand, with consequent positive effects on motor and sensory function [21]. Care should be taken, however, to ensure that the patient is not psychologically overstressed by this forced use treatment.

#### Surgical treatment

*Botulinum toxin A* is used for the treatment of troublesome muscle hyperfunction. This is an alternative way of inactivating the adductor pollicis muscle in a deviating thumb or the flexor carpi ulnaris muscle in a spastic ulnar deviation-flexion deformity of the wrist. The botulinum toxin temporarily disables the locally injected muscles (▶ Chapter 4.7). The injection on its own can lead to func-

tional improvements similar to those obtained by surgical corrections [12, 6]. The functional can be checked and the injection repeated. The additional use of functional or positional braces is also possible. The administration of botulinum toxin A also enables a surgical treatment plan involving muscle lengthening and weakening procedures to be tested in advance [1]. If the plan does not prove to be favorable and the treatment leads to a deterioration, the temporary effect wears off after approx. 3 months, and the patient suffers no residual damage [4].

Spastic muscle activity in patients with contractures can interfere with stretching treatments, and thereby often frustrate the goals of physiotherapy, orthoses and casts. The efficiency of these treatments can be enhanced, and a surgical procedure possibly postponed or even avoided, by injecting botulinum toxin A into the affected muscle groups.

*Surgical measures* are more difficult to plan for the upper extremities than for the lower limbs. The aim of an operation on the upper extremities is to restore the muscle equilibrium and thus improve the use of the hand, promote coordination and enhance its function as an organ of touch [23]. The ideal preconditions for an operation are: patient's willingness to cooperate, predominantly pyramidal signs and symptoms, minimal emotional effect on spasticity, good voluntary control of the spastic muscles and the necessary willpower to concentrate and cooperate [23]. In all cases, the patient must at least understand the goals of the surgical treatment and cooperate postoperatively. These extensive conditions are rarely fulfilled in patients with spastic tetraparesis since they are almost invariably retarded to a greater or lesser extent, difficult to motivate and unable to provide sufficient cooperation. Additional sensory changes further diminish the prospects of a good results. An intervention is ideally implemented after the nervous system has fully matured (after the age of 5 or 6) [23]. For all the above reasons, surgical corrections of the upper extremity are relatively rarely indicated.

A disruptive adduction is usually present at shoulder level, which can pose a problem in respect of nursing care particularly in severely disabled patients. Injections of botulinum toxin A or muscle lengthening procedures may resolve this problem. Osteotomies and arthrodeses have been described for patients with fixed deformities.

Flexion contractures at the *elbow* are relatively common. However, since these are usually slight or moderate, and as long as they do not hinder the patient, surgical treatment is not required. Nocturnal splints can be used for patients with significant progression of the contractures. We have only encountered very troublesome flexion contractures in severely tetraspastic patients. Elbow extension orthoses are difficult to use, particularly if spastic counter-tension is present. In such cases, the injection of botulinum toxin A can slacken the countertension. It can also be used to distinguish between a contracture that is merely func-

tional and a fixed contracture. Severe cases of the latter will require surgery, and procedures to lengthen the muscles, particularly the biceps brachii and brachioradialis, can be therapeutically beneficial.

The *hand* is frequently in a position of palmar flexion and ulnar deviation with pronation at the wrist, flexion of the fingers and an adduction-pronation deformity of the thumb. The palmar flexion at the wrist is inauspicious since the strength of the finger flexors is reduced in this position. There is also the risk of contractures of the finger flexors if these are never used over their full length. The ulnar deviation and pronation rotates the hand outside the field of vision, making it difficult for the patient to have any visual control over the hand function. This rules out the option of visual compensation for any impaired sensory function and considerably aggravates the use of the disabled hand. The position of the wrist can be improved functionally by a splint. This, usually palmar, orthosis extends from the forearm to the metacarpus, but should not extend beyond the distal flexion crease otherwise it will interfere with free finger function.

Good support beneath the thumb metacarpophalangeal joint counteracts the dislocation of the 1st metacarpal in this joint. This appliance can also be used to test how a patient would react to a corrective, stabilizing procedure in which the flexor carpi ulnaris muscle is transferred, in one of various ways, to the extensor carpi radialis brevis or longus muscles (*Operation according to Green* [10]).

A preoperative botulinum toxin A injection will preclude any functional deterioration resulting from a loss of power of the transferred muscle. A wrist arthrodesis can also produce a functional improvement by providing extra stability [19]. This corrects the position at the wrist and the grasp function of the hand. The results after this operation are also good in the long term [2].

Additional procedures on the pronator quadratus muscle may be indicated at the same time, particularly if the improvement in the rotation of the forearm and hand is of prime concern. If active supination up to the neutral position only is possible, the pronator quadratus muscle or the pronator teres muscle should be lengthened. If active supination is absent, but free movement is possible passively, transfer of the pronating muscles is indicated. If movement restriction without pronatory activity is present, the pronator quadratus is lengthened and can be transferred at a later date (■ Table 3.80) [11]. The results are better after transfer than after lengthening. A possible alternative to muscle weakening by surgical lengthening is the injection of botulinum toxin A.

For fixed flexion deformities of the wrist or a concurrent troublesome instability, an *arthrodesis of the wrist* can produce good results. When correctly performed this procedure can also be employed for young patients without growth disturbances. In addition to the pronation-flexion position of the wrist, the whole hand is often



## 3.5.6 · Neuromuscular disorders of the upper extremity

deformed, with spastic contractures, and the fingers are flexed. Braces can be used to prevent and improve flexion contractures. If the patient has undergone previous surgery, however, and severe finger deformities persist, operations for correcting the finger function and position must be considered as a supplement to the transfer of the flexor carpi ulnaris muscle (■ Table 3.81).

The options for correcting the adduction-pronation deformity of the thumb are listed in ■ Table 3.82 and essentially involve muscle lengthening procedures and transfers of the extensor pollicis longus [23].

For cosmetic reasons we have occasionally performed the simple Green operation combined with procedures for lengthening the finger flexors. No negative results have been noted to date. In the swan-neck deformity of the fingers (see above) it is usually sufficient to correct the wrist contracture. In severe cases, a release of the pronator teres muscle may be required. Muscle surgery is generally inadvisable in patients with athetotic atactic-dystonic syndromes in view of the risk of overshooting deformities in the opposite direction, which then usually become more troublesome than the originally treated primary deformities (for example, a hyperextension develops after a flexion contracture at the elbow). Consequently, conservative measures tend to be more appropriate than surgical interventions, although stabilizing operations (usually arthrodeses) may be required in some cases. It is technically difficult, however, to provide sufficient stability by internal fixation until the arthrodesis has consolidated. In one patient, for example, we have had to stabilize a wrist arthrodesis with two plates instead of just one. Despite a plaster dressing postoperatively, a further operation was required to correct a pseudarthrosis. Since then the arthrodesis has consolidated to produce a good end result (■ Fig. 3.498).

Recent studies have shown that deteriorations are unlikely to occur after operations on the upper extremity. Patients undergoing surgery for purely cosmetic reasons have usually benefited from a functional improvement as well [18, 7]. The sensory problem is of secondary impor-

■ **Table 3.80.** Protocol for the treatment of pronation contracture. (According to Gschwind and Tomkin [11])

Functional deficiency	Surgical treatment
Active supination beyond the neutral position	No operation
Active supination up to the neutral position	Release of the pronator quadratus muscle, poss. with aponeurotic lengthening of the flexors
No active supination, but passively free, loose supination	Transfer of the pronator teres muscle
No active supination, passive supination restricted or stiff	Release of the pronator quadratus muscle with aponeurotic lengthening of the flexors, poss. secondary transfer of the pronator teres muscle

■ **Table 3.81.** Protocol for the surgical treatment of the hand in spastic cerebral palsy. (According to Zancolli et al. [23])

Group	Functional deficiency	Surgical treatment
1	Slight flexion contracture in the wrist (flexor spasticity)	Distal tenotomy of the flexor carpi ulnaris muscle
2	Pronounced flexion contracture in the wrist	Distal tenotomy of the flexor carpi ulnaris muscle, aponeurotic release of the muscles at the medial epicondyle, flexor detachment from the medial epicondyle ( <b>N.B.:</b> avoid overcorrection)
	<p><b>a</b></p> <p>Wrist extensors active</p> <p>Finger flexors active during wrist flexion &lt;70°</p> <p>Finger flexors active during wrist flexion &gt;70°</p>	
	<p><b>b</b></p> <p>Wrist extensors inactive</p>	Transfer of the flexor carpi ulnaris to the extensor carpi radialis brevis or longus muscles
3	Flexion contracture in the wrist	Flexor detachment from the medial epicondyle or tendon lengthening of the finger and wrist flexors, tenotomy of the pronator teres muscle

■ **Table 3.82.** Correction of thumb deformities. (According to Zancolli et al. [23])

Deformity	Functional deficiency	Operation
Thumb abducted	Adductor pollicis muscle spastic, metacarpophalangeal joints extended In hyperextension >20°	Detachment of the adductor pollicis longus from its origin and tenodesis in abduction Volar capsulodesis
Thumb adducted and flexed (»thumb in palm«)	Adductor pollicis muscle spastic, metacarpophalangeal and interphalangeal joints flexed due to spasticity of the flexor pollicis longus muscle	Detachment of the adductor pollicis longus from its origin and tenodesis in abduction and release of the flexor pollicis longus and transfer radially of the extensor pollicis longus





Fig. 3.498. Wrist arthrodesis in severe athetosis

tance in this context [9]. In view of these considerations, surgical procedures on the upper extremities should be indicated more liberally, subject to the requirement that, as with all functional procedures, reliable and adequate follow-up treatment is provided.

### 3.5.6.2 Primarily flaccid paralyses *Plexus palsy*

#### ► Definition

A plexus palsy refers to a nerve lesion between the point at which the spinal roots leave the cord and the point where they divide to form the peripheral nerves. Various mechanisms can produce the lesion, from simple stretching to tearing of nerve fibers or the avulsion of the roots from the spinal cord.

#### Etiology and pathogenesis

Plexus palsies of the upper extremity can occur at birth as a result of manipulations. Labor-induced lesions occur in 0.04%–0.25% of all live births (with a clearly declining trend). Infants in a breech presentation are particularly at risk, although these are almost always delivered by cesarean section nowadays. Older children and adults can suffer plexus lesions of the upper, or more rarely the lower, extremity in traffic accidents (particularly motorcycle accidents) [15]. No basic distinction is made between plexus lesions in the neonate and the adult [17]. If the nerve is not completely severed or torn, the axon will regrow from

the center. However, the more proximal the lesion, the more likely abnormal sprouting will occur.

### Clinical features and diagnosis

#### Initial signs and symptoms

Sensation and motor activity can be tested in older children and adults. The striking finding in neonates is a failure to move the arm, which hangs limply with the elbow extended and does not even move when the typical infant reflexes are elicited (e.g. the Moro reflex). If the lower cervical roots are also involved the grip movement of the hand is absent. A distinction is made between an upper and a lower plexus palsy. Most children present with an upper (Erb) or complete plexus palsy. The lower plexus palsy (Klumpke) is rare (0.6% of all plexus palsies). In the more common *upper plexus lesion (Erb-Duchenne)* the roots of C 5 and C 6 are affected. Examination of motor function reveals the absence of abduction and external rotation at the shoulder, elbow flexion and, to some extent, elbow extension, supination and wrist extension. The shoulder hangs down low with the elbow extended. While sensory impairment is not necessarily present, any impairment that does occur is located on the outer aspect of the upper arm and the radial side of the forearm.

The *lower plexus palsy (Klumpke-Déjérine)* primarily affects the T1 segment and, in some cases, C 8. The small hand muscles show no activity and the ulnar fingers, in particular, adopt a claw-like posture. But the long finger flexors and wrist flexors may also be weak or inactive. Sensation is impaired on the ulnar side of the hand and forearm. A Horner syndrome (ptosis, myosis, enophthalmos) may also be present, indicating the involvement of the sympathetic nerves on the same side. Besides these two typical clinical pictures, a total arm plexus palsy or other forms of plexus palsies (involvement of C 7 or fascicular palsies) may also be present.

With the aid of an electromyogram, a distinction can be made between a complete and incomplete paralysis and the progression of reinnervation monitored. The roots and root pouches can be visualized on an MRI scan or myelogram. More peripherally located lesions can be detected by the appearance of hemorrhaging on the MRI scan.

If deficient movement is present in the upper limb, the *differential diagnosis* must first rule out other injuries (fractures of the humerus or clavicle or infections in the shoulder area). In such cases a pain-induced pseudoparalysis is involved, and the reflex activity of the muscles is normal.

#### Late sequelae

Pareses due to birth trauma are typically followed by the development of a muscular dyskinesia at shoulder level, where abduction occurs rather between the scapula and thorax than in the glenohumeral joint. At the same time, flexor activity is triggered in the arm, leading to flexion

at the shoulder and elbow. The deltoid muscle is inadequate as an abductor. The dyskinesia can be explained by altered motor neuron activity, abnormal sprouting or by the abnormal development of the maturing nervous system. If abnormal sprouting is present, the defective muscle activity can be corrected – at least temporarily – by the intramuscular administration of botulinum toxin A [8, 20].

The shoulder muscles may remain flaccid or be contracted, usually in a position of internal rotation/adduction. This type of contracture may also lead to the development of bone deformities as a result of the modified habitual use of the shoulder. The glenoid flattens out and becomes broader, while the physiological retrotorsion at the humerus is increased. This leads to posterior subluxation. A flexion contracture often occurs at the *elbow* as a result of hyperactivity of the biceps and brachialis muscles and the use of the elbow in a flexion position. The olecranon and coronoid process become enlarged and additionally block extension. Radial head dislocations also occur. A pronation contracture of the *forearm and hand* is present in upper plexus palsies. Lower palsies mainly affect the hand muscles only.

### Treatment and prognosis

The prognosis for an upper plexus palsy is better than that for a lower palsy. Overall, over 90% of cases of postpartal palsy recover completely [14], this figure dropping to just 50% in cases of posttraumatic palsy. Unfavorable parameters include the effect of substantial force, complete palsies, additional injuries and pain. The latter is indicative of a root avulsion. The sensory functions recover much better than the motor functions.

### Conservative treatment

In view of the inherently good prognosis, the primary goal of treatment must be to prevent the onset of secondary deformities as these will restrict the function of the extremity following its reinnervation. Training of the existing and newly innervated muscles is also helpful. In order to promote the best possible healing, the arm is placed on the rib cage after birth so as to relax the plexus – an ideal precondition for recovery. From this position, the muscles are stretched and the existing muscles activated by physiotherapy. The method according to Vojta is often used for these palsies, but this should probably aim at strengthening the existing muscles rather than promoting reinnervation. The extent to which such stimulation treatments actually produce a positive effect on axonal sprouting and the healing of the plexus palsy has not been objectively demonstrated. In any case, this would be difficult to prove in a condition with such a strong tendency towards spontaneous healing.

Orthoses and braces can be used, at best, as braces if no recovery occurs in the long term and the contracture

tendency persists. In most cases, however, they are unnecessary. They also involve the risk of shoulder subluxations, particularly if there are deficits in the muscles that stabilize the shoulder.

### Surgical treatment

A *revision of the brachial plexus* will be required in approx. 15% of patients, for both the posttraumatic and postpartal forms. This procedure is indicated if there are no signs of a recovery of motor or sensory function by the end of 3 months. The surgical options include neurolysis, direct suturing or a nerve interposition, and neurotization. Neurolysis is indicated if pain is present or, to a lesser extent, in order to improve function. If the plexus is completely severed, nerve suturing, possibly with the interposition of a graft, can prove successful. The prognosis is better for the upper roots than the lower roots, and this also applies to secondary procedures. Neurotization may be indicated in the event of root avulsions. In this procedure less important nerves (e.g. intercostal nerves) are connected to the stumps of the plexus. The prognosis for improvement of elbow function with this method is good (> 50% of patients); the prognosis for shoulder and finger function is limited and poor respectively [16].

Any subsequent *corrective operation* must be preceded by a careful investigation of the functional disorders and deformities and the resulting impairment to the patient. Not only the motor aspects, but also the cosmetic and psychological components, must be identified. Troublesome functional deficits can be improved by muscle transfer procedures, and existing contractures must be eliminated beforehand or at the same time with a muscle transfer performed by means of muscle lengthening procedures rather than tenotomies.

*External rotation and abduction* of the arm can be improved by transfer of the levator scapulae to the supraspinatus and relocation of the teres major, with or without the latissimus dorsi, to the infraspinatus muscle. The trapezoid can also be transferred to improve the shoulder function. In addition, there may be posterior or anterior *subluxation* or *dislocation* of the humeral head. A suitable procedure for correcting this problem is a rotating osteotomy of the proximal humerus in the direction required to center the joint [23]. The latissimus dorsi muscle can also be used as a replacement for the deltoid [17]. If severe and troublesome instabilities are present, an arthrodesis of the shoulder may be indicated. Usable *elbow* function is important for any functional use of the arm. The latissimus dorsi can also be used to improve elbow flexion. Alternatively, the triceps brachii or pectoralis muscle can be transferred to the biceps brachii [3].

A supination deformity can be rectified by transferring the biceps brachii so that it can be used as a pronator. A precondition for this procedure is a free pronation/supination movement. At the same time, a corrective oste-

otomy at forearm level may be required if the radius, or more rarely the ulna, is deformed. A posterior radial head dislocation can also occur, and this can be corrected by radial shortening (in younger children) or a radial head resection (in older children). *Thumb* opposition can be restored by transferring the flexor digitorum superficialis of the ring finger or the abductor digiti minimi to the adductor pollicis brevis muscle.

### Peripheral nerve palsy

#### ➤ Definition

A peripheral nerve palsy involves a partial or complete failure of its motor and/or sensory functions.

#### Etiology and pathogenesis

Peripheral nerves are injured in accidents, as the result of deliberate harm or iatrogenically following their accidental severing at operation or traction exerted, for example, during the course of corrective axial and lengthening procedures on the extremities.

#### Clinical features and diagnosis

A lesion of a peripheral nerve leads to disorders of sensation and motor function in the area supplied by that nerve. The signs and symptoms in typical areas enable the damage to be assigned anatomically to the individual nerves (► Chapter 2.1.2). The sensory impairment can be present in the form of anesthesia, hypoesthesia, paresthesia or hyperesthesia. In terms of motor function there is weakened muscle activity, possibly extending to complete failure. The palsies are invariably flaccid. A distinction must be made between prognostically favorable incomplete paralyzes and complete failures with anesthesia and complete paresis. A spontaneous remission can generally be expected for incomplete lesions. In cases of complete paralysis, however, only over time will it become apparent whether function will recover or whether the deficit will persist unchanged. An electromyogram can be recorded to reveal any muscle innervation that is not clinically detectable and to rule out any severed nerves. The same technique can be used to monitor regeneration.

#### Treatment and prognosis

##### Conservative treatment

The orthopaedic treatment is primarily aimed at preserving mobility and avoiding contractures so that the muscles can resume their function under the optimal conditions. The main focus of conservative measures is on exercise-based treatments in the context of occupational therapy and/or physiotherapy. In addition to stretching and strengthening exercises, training is provided in the use of the limb for everyday tasks. Braces can be helpful for avoiding contractures. Moreover, functional orthoses can make up for lost muscle functions.

#### Surgical treatment

A curative procedure is *suturing of the damaged nerve*, possibly with an interposed graft. The prognosis is better, the more distal the lesion, the smaller the damaged section, the younger the patient and the shorter the period that elapses until treatment. Primary reconstructions of peripheral nerves are successful in 80–90% of cases, even though a complete recovery cannot be expected. Secondary procedures produce slightly poorer results, with success rates of 60–70%. Some nerves, e.g. the peroneal nerve, tend to show poorer recovery from the outset. Functionally usable muscle activity can be expected in approx. 50% of patients.

Palliative surgical procedures include *muscle transfers* and *stabilizing measures* such as arthrodeses or tenodeses. Before any operation the functional deficits and the degree of handicap must be clarified and the expectations discussed with the patient (► Chapter 3.5.6). If *radial nerve palsy* is present, wrist extension can be improved by transferring the pronator teres muscle to the long and short radial extensor muscles, while finger extension can be enhanced by relocating the flexor carpi ulnaris or a part of the flexor digitorum superficialis muscle to the extensor digitorum communis. In a *median nerve palsy* the deficit interferes with finger flexion and thumb opposition. Abduction and pronation of the thumb can be restored by transferring the extensor indicis muscle, and adduction by relocation of the extensor carpi radialis brevis muscle in combination with a thumb metacarpophalangeal joint arthrodesis.

The claw fingers in *ulnar nerve palsy* can be improved with a »lasso transfer« of the flexor digitorum superficialis, although this will result in a loss of power in respect of fist closure. The pinch grip must be restored with additional measures such as a metacarpophalangeal joint arthrodesis of the index finger and transfer of the extensor carpi radialis brevis as the thumb adductor. In any muscle transfer procedure, however, the surgeon must weigh the loss resulting from the removal of the muscle against the benefit obtained by the new function.

#### Arthrogryposis

Further details are provided in ► chapter 4.6.7.1.

The goal of treatment is to give the patient as much independence as possible. Although the range of motion in the joints can often not be increased, it can be relocated so that the use of the extremity can be improved. At the same time it may prove necessary to treat both upper limbs asymmetrically in order to bring one side more in extension and the other more in flexion. This can help restore the functions needed for eating and personal hygiene. Contractures and muscle weakness at the shoulder are often present concurrently. Functional restrictions, however, tend to be present only in cases of fixed internal rotation, because the arm can no longer be controlled in a sideways direction (for the purposes of personal hygiene

or walking exercises with walking aids). Rotational osteotomies are indicated in such cases.

Contractures at the *elbow* can progress in a variety of ways. Whereas some patients respond well to conservative measures, the deformity becomes progressively worse in others, ultimately resulting, for example, in a flexion contracture at the elbow with movement around the right angle, but with sufficient power remaining in the biceps brachii muscle. Treatment is not usually indicated in these patients. On the other hand, the elbow may stiffen in an extended position, which can significantly interfere with everyday functioning. In these cases, lengthening or transfer of the triceps brachii muscle, possibly combined with a flexor reconstruction, may be indicated.

Often a severe flexion contracture at the *wrist* will already be present at birth, and sometimes fingers and thumb are also affected. Measures to correct this deformity must be initiated as soon as possible, with stretching exercises and splints. At a later stage, the wrist instability, and particularly the lack of dorsal flexion, will present a major problem. Stabilizing orthoses for the wrist can significantly improve the functioning of the hands (■ Fig. 3.499). Alternatively, a tendon transfer (transfer of the flexor carpi ulnaris posteriorly to the base of the 3rd metacarpal) can be offered. A wrist arthrodesis can produce positive effects and provide stability in the corrected position. A pronation contracture can be eliminated by a tenotomy of the pronator teres muscle. Often the thumb is adducted and the interdigital space between the 1st and 2nd rays is too narrow. If conservative stretching exercises prove unsuccessful, a reconstructive procedure to widen the interdigital space may be necessary to improve thumb abduction. At finger level, a distinction must be made between joint contractures and shortening of the finger flexors. If the range of motion of the fingers is improved with palmar flexion at the wrist, a contracture of the flexor muscles is present and tendon lengthening is indicated. A soft tissue release can be useful for joint contractures. In the windswept deformity the fingers deviate in the ulnar direction, are flexed at the metacarpophalangeal joint, while the other finger joints are stiff in flexion or extension. The deformity can be corrected by recentralizing the ulnar-deviating extensor

tendons and transferring the ulnar interosseous muscles. In older children or in cases of more pronounced deformity, reconstructive procedures on the skin and corrective osteotomies on the metacarpals may also be required. For pronounced contractures, the necessary lengthening of the muscles can be achieved by bone shortening, either by a shortening osteotomy of the forearm or by resection of the proximal row of carpal bones.

### Post-polio syndrome

On the upper extremity, the deltoid is the muscle most commonly affected in this disorder, although the muscles of the rotator cuff may also be paretic and possibly lead to an inferior shoulder dislocation. Troublesome dislocations require an arthrodesis to stabilize the joint. At the elbow, both flexors and extensors can show weaknesses while, at hand level, thumb opposition in particular is impaired.

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■ Fig. 3.499. Stabilizing orthosis for the wrist in *arthrogryposis*



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### 3.5.7 Fractures of the upper extremities

C. Hasler

#### 3.5.7.1 Scapular fractures

##### Occurrence

Apart from the spina scapulae, the acromion and the coracoid, the scapula is deeply embedded on all sides in the protecting musculature.

**!** **Scapular fractures are very rare and evidence of considerable force, which can produce serious additional injuries. In terms of prognosis, the latter are more decisive than the scapular fracture.**

In the absence of adequate trauma in the history, the possibility of child abuse should be considered, particularly in small children [40].

##### Diagnosis

##### Clinical features

The clinical picture is dominated by the additional injuries to the skull, thorax and abdomen. The striking finding locally is a painful restriction of movement of the shoulder, particularly from 70–90° of glenohumeral abduction, when the scapula starts to rotate as well.

##### Imaging investigations

The conventional radiological presentation of a scapular fracture on AP and Y views is occasionally inconclusive. A CT scan is effective in doubtful cases, particularly in

respect of the possible indication for surgery. The two ossification centers of the coracoid and the 2–5 centers in the acromion can occasionally lead to confusion and misdiagnoses, particularly if they persist as a bipartite or tripartite acromion [40].

##### Fracture types

Scapular body, scapular neck, coracoid, acromion, glenoid.

##### Treatment

Most scapular fractures heal without complications with temporary immobilization in an arm sling or a Gilchrist bandage and early functional follow-up. *Surgical revision* may be required, in rare cases, for glenoid fractures with glenohumeral instability, scapular neck fractures in combination with a clavicular fracture and displaced coracoid fractures [49].

##### Prognosis

The prognosis depends primarily on the *additional injuries*.

#### 3.5.7.2 Clavicular fractures

##### Occurrence

10% of all fractures during growth. Viewed from the front, the clavicle is straight, while from above it appears S-shaped with a forward-facing convexity in the middle third. Given the absence of muscles on the anterior and superior sections, the shape and length of the clavicles substantially determine the appearance of the shoulder girdle. As a spacer between the acromion and sternum, it is in a vulnerable position, particularly in the event of falls on the lateral shoulder. A clavicular fracture is the commonest injury caused by *birth trauma*.

**!** **The clavicle plays a key role in the functional connection between thorax and arm and provides bony protection for the subclavian arteries, subclavian veins and the neural network of the brachial plexus.**

##### Diagnosis

##### Clinical findings

In children and adolescents the local pain over the clavicle confirms the fracture. Although rarely associated with a clavicular fracture, a plexus palsy should be ruled out [63]. The latter can also be induced by an excessively tight figure-of-eight strap.

##### Imaging investigations

AP x-ray of the clavicle. In view of the superficial position of the clavicle, diagnosis by ultrasound is a valid alternative to radiographic investigation.



### Fracture types

*Closed, uncomplicated shaft fractures of the middle third* account for the highest proportion, by far, of all clavicular fractures. The younger the child, the more likely it is that the fracture will be non-displaced.

*Lateral fractures* frequently correspond to epiphyseal separations and, in clinical respects, resemble an acromioclavicular dislocation as seen in over 13-year olds or adults.

**!** In children with lateral clavicular fractures the inferior section of the periosteal sleeve and the adjacent coracoclavicular ligaments remain intact. The outstanding osteogenic potential of the periosteum leads to rapid consolidation and impressive remodeling of the clavicle in its elevated position.

*Medial fractures* are rare and represent epiphyseal separations with retrosternal dislocation of the lateral fragment [26].

### Treatment

#### Conservative

**!** Because of their short shoulder girdle, a figure-of-eight strap is ineffective and uncomfortable in children and would also need to be retightened at regular intervals.

All that is required for treating the pain, therefore, is immobilization in a simple arm sling for 2 weeks in combination with oral analgesics for 3–4 days. The results after a figure-of-eight strap and an arm sling are identical. Depending on the severity of the symptoms, arm-hanging exercises may be initiated independently after just 1–2 weeks. For initially displaced fractures, an x-ray



should be arranged after 4 weeks if local pain on palpation persists. Otherwise the absence of symptoms is evidence of consolidation.

### Surgical

Open reduction and internal fixation of shaft fractures is indicated only in exceptional cases:

- Shortening in excess of 2 cm after physeal closure. The risk of pseudarthrosis is higher during childhood and the cosmetic result is often experienced to be unsatisfactory [33]. In this case, the patient should be informed, preoperatively, particularly about the wide, keloid-like scars that can often result.
- Open fractures or fractures with threatened penetration.
- Concurrent neurovascular symptoms.
- Established symptomatic pseudarthrosis.
- Pathological fractures.
- Combination with scapular neck fracture.

We prefer internal fixation with a small-fragment plate fixed to the clavicle from the bottom.

*Medial epiphyseal separations* with retrosternal dislocation require emergency reduction, usually as an open procedure [26].

### Prognosis and complications

Displaced fractures with an ad latus deformity and shortening result in a distinct bony bulge, which is often even more accentuated at a later stage as a result of marked callus formation. Both the bulging and the shortening remodel themselves if the growth plates are still open, although this takes from 6–12 months. Informing the parents and the patient accordingly will prevent additional consultations and unnecessary corrective procedures.

- Shortening of the shoulder girdle of over 5 mm after initially displaced fractures are common, but is of no consequence for shoulder function [60].
- Since *pseudarthroses*, *incorrect healing* and *neurovascular complications* are rare after isolated, uncomplicated clavicular fractures, follow-up beyond the consolidation period is not normally indicated [8].
- *Pseudarthroses* must be differentiated from rare conditions with a *congenital* etiology and from *cleidocranial dysostosis*.
- *Growth disturbances* and *neurovascular complications* are rare. The latter can also be provoked secondarily by hypertrophic callus.

### 3.5.7.3 Proximal humeral fractures

#### Occurrence

Less than 5% of all pediatric fractures. Apart from the few cases resulting from birth trauma, these fractures occur mainly in over 10-year olds.

❗ **A conservative approach with early functional therapy is particularly suitable for fractures of the proximal humerus since the highly active growth plate, which accounts for over 80% of the length growth of the humerus, possesses outstanding remodeling potential and since minor residual deformities can be offset by the very mobile shoulder joint and concealed by the thick muscle covering [5].**

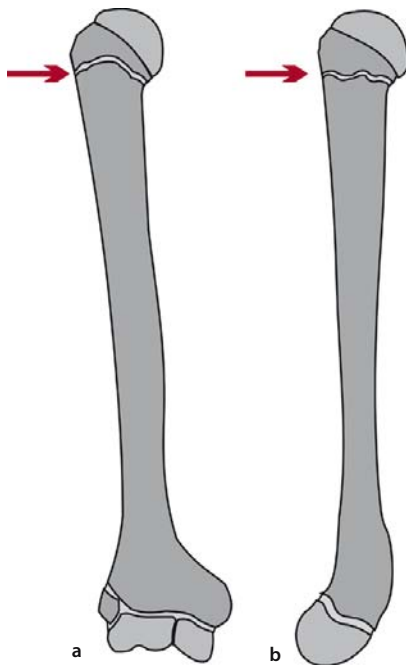
### Diagnosis

#### Clinical features

Pain in the area of the proximal humerus. Deformities are not usually visible.

#### Imaging investigations

AP and Y x-rays are indicated for children and adolescents (■ Fig. 3.500). Depending on the forced posture produced by the pain, the proximal humerus may not appear to be affected from the front on the AP view or from a strictly lateral position on the Y view. The diagnosis can be assisted by, on the one hand, including the elbow in the x-ray or, more reliably, by noting the shape of the proxi-



■ **Fig. 3.500.** Radiographic diagnosis of proximal humerus: AP and lateral: If the elbow is not x-rayed at the same time it can sometimes be difficult to differentiate between the AP and lateral planes on the x-rays of the proximal humerus. The course of the epiphyseal plate serves as a pointer: On the AP view it shows a tent-shaped elevation (a), on the lateral view it appears straight and runs at right angles to the shaft (b)

mal humeral epiphyseal plate, which appears roof-shaped from the front and flat from the side.

#### Fracture types

— *Epiphyseal separations due to birth trauma:* Children in the less frequently affected age group of 5- to 10-year olds suffer from *metaphyseal*, subcapital fractures. Beyond this age, *epiphyseal separations* in particular, occur either without (Salter type I; ► chapter 4.1), or in adolescents with, a posteromedial metaphyseal wedge (Salter type II). However, such differences are of no therapeutic importance, and very rarely of any prognostic significance, since relevant growth disturbances are extremely rare. A crucial requirement is to note the degree of ad latus displacement and the tilt angle. The hyperextension traumata lead to tilting in the dorsal direction, but rarely to instability.

Epiphyseal fractures (Salter types III and IV) and avulsion fractures of the lesser tubercle are rare, as are subcapital fractures in combination with glenohumeral dislocation [61].

#### Treatment

##### Spontaneous corrections

In view of the outstanding remodeling potential, the following deformities can be left untreated [43]:

- <10-year old girls, <12-year old boys: varus-, ante- and recurvation deformities up to 50–60°, valgus deformities (much rarer) up to 20°. Ad latus deformities by the full shaft width and shortening of up to 2 cm.
- Above these ages, approximately half the above values can be tolerated while the physéal plates are still open.

❗ **Comprehensive briefing of the parents and patient about the biological and chronological processes of spontaneous remodeling of untreated deformities is very important in order to avoid unnecessary »medical tourism« or even surgical interventions.**

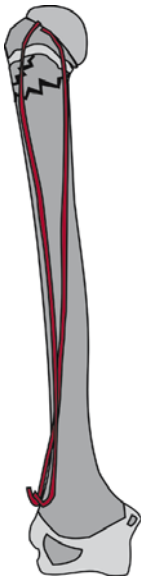
##### Conservative

After 1 or 2 weeks of immobilization in an arm sling or, if the condition is painful, in a Gilchrist bandage, the patient is given instruction on mobilizing the shoulder independently with active and passive arm-hanging exercises.

##### Operation

###### Closed reduction

Although rarely indicated, a reduction under anesthetic is usually appropriate if there is unacceptable angulation in a dorsal direction and a varus position. The reduction maneuver consists of traction, abduction, flexion and slight external rotation. If the fracture reduction appears stable under the image intensifier, the follow-up treatment is the application of a Gilchrist bandage for 2 weeks.



■ **Fig. 3.501.** Treatment of displaced fractures of the proximal humerus at the age of >12 years: If the situation is unstable after closed reduction in patients older than 12 years of age and an unacceptable degree of displacement is present (>20°), it is advisable to stabilize the reduced position by intramedullary nails (inserted via the radial epicondyle)

#### Closed reduction and stabilization

In cases of persistent instability or for patients aged over 12, the fracture should be stabilized after reduction with two flexible medullary nails inserted from the distal end of the humerus on the lateral side (■ Fig. 3.501). We do not perform percutaneous Kirschner wire fixation since it interferes with early independent shoulder mobilization and often results in superficial infections.

#### Open reduction

In the rare cases of fractures that cannot be reduced satisfactorily by the closed method, the long biceps tendon is usually interposed and can be freed from the fracture gap via a short deltoideopectoral incision.

#### Follow-up controls

A consolidation x-ray after 4–5 weeks is indicated only for untreated deformities and after reductions with or without fixation. A significant correction is often apparent just a few weeks after the onset of untreated deformities, primarily as a result of the restoration of normal muscle tone.

#### Complications

- *Growth disturbances* in the context of premature *partial physeal closure* occur particularly after epiphyseal separations due to birth trauma that had been overlooked. These usually result in a varus deformity, but rarely involve any functional restriction [18]. Shortening of up to 2 cm can occur in association with fractures that are completely displaced initially and left to remodel spontaneously, but this is of no clinical significance.
- *Avascular necroses* do not occur during childhood and adolescence [43].

### 3.5.7.4 Humeral shaft fractures

#### Occurrence

Less than 5% of all pediatric fractures. The possibility of child abuse must be ruled out particularly in under 3-year olds. Humeral fractures account for almost two-thirds of all acute fractures discovered in cases of child abuse [69]. Most humeral shaft fractures however are seen in adolescents, particularly as a result of direct trauma in sports-related and traffic accidents [12].

#### Diagnosis

##### Clinical findings

The diagnosis usually readily confirmed by clinical examination (pain, swelling, deformity). Careful identification and documentation of the *neurovascular status* is essential. Radial nerve and, rarely, ulnar nerve palsies occur in approx. 4% of all childhood humeral shaft fractures, particularly those involving the distal third of the shaft. In a case of a nerve palsy, we simply monitor the spontaneous course over 6–8 weeks. Recovery can be expected in over 80% of cases as these usually only involve neurapraxia. If no improvement is observed clinically or on an EMG, the lesion should be explored and, depending on the findings, treated by neurolysis or a graft to bridge any defect. In cases of open fractures with suspected nerve laceration, the nerve revision procedure should be performed primarily in connection with the fracture treatment [6].

#### Imaging investigations

AP and lateral x-rays of the humerus, including the humeral head and elbow.

#### Treatment

##### Conservative

Most axial deviations in humeral shaft fractures can be managed with conservative measures:

- For *simple, stable* fractures (compression fractures, greenstick fractures), immobilization in an arm sling is sufficient.
- For *unstable* fractures with an existing axial deviation or with a risk of secondary dislocation, we immobilize the fracture until the swelling and pain subside in a plaster-of-Paris Desault bandage that incorporates both the shoulder and elbow. The plaster bandage is preferably applied to the seated patient while slight traction is exerted on the upper arm. After 5–7 days, a Sarmiento brace is individually fitted to the patient after a check x-ray [67]. This is a double-shell for the upper arm made from a semi-rigid thermoplastic material. The pressure can be adjusted by Velcro fasteners and is applied evenly to the soft tissues of the upper arm thanks to the optimal anatomical contouring. This produces hydraulic splinting of the fracture. The shoulder and elbow are

freely movable and should be actively and passively mobilized independently by the patient within the limits of pain. After one week with the brace, another check x-ray is recorded.

Management with a brace can be difficult in obese patients or in ventrally angulated fractures with substantial distal extension. We have dispensed completely with the use of the so-called »hanging cast«, since the weight of the plaster is very uncomfortable for the young patients and the fracture control is no better than with an upper arm brace.

### Surgical

The use of an *external fixator* and *flexible intramedullary splinting* are two minimally-invasive methods that respect the biology of the fracture zone and minimize the risk of an iatrogenic radial nerve palsy, cases of which have been reported in association with internal plate fixation and, in particular, implant removal (■ Fig. 3.502).

Nevertheless, the course of the radial nerve must be carefully noted, particularly during the insertion of fixator screws. We prefer nailing for short oblique fractures and transverse fractures, resorting to the unilateral external fixator for long oblique fractures, multifragmented fractures and open fractures.

Surgery is most often indicated for cases in which the axial deviations cannot be controlled by conservative treatment. Other indications for surgery, including

humeral shaft fractures in polytraumatized patients, bilateral fractures, open fractures, compartment syndromes, extensive soft tissue lesions and concomitant vascular injuries, are rare.

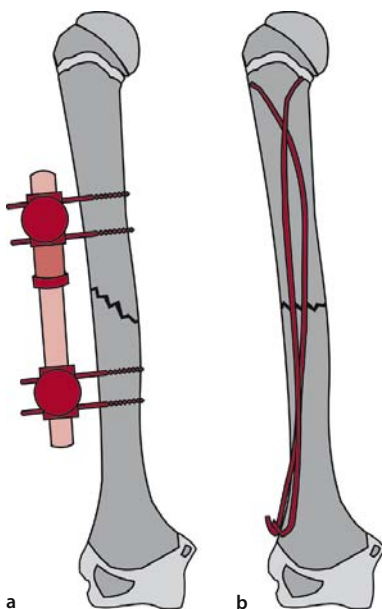
### Follow-up controls

The radiological positional check is indicated after 7–10 days and a consolidation x-ray after 6 weeks. Once an anatomical axial position has been achieved, clinical follow-up is continued until full elbow and shoulder function is restored. If residual axial defects are present, the patients should be monitored until completion of growth.

### Complications

#### Growth disturbances and posttraumatic deformities

- *Axial deformities*: The potential for spontaneous correction in the shaft of the upper arm is very limited, i.e. less than 10°. This applies particularly to valgus deformities. However, since axial kinks in this context are neither cosmetically conspicuous nor of any mechanical importance, they can also be tolerated in adolescents before the end of growth. In particular, varus angulations of up to approx. 20° are not very obvious, in contrast with anterior bowing, which is the most conspicuous type of deviation. Side-to-side displacement and shortening may be left untreated, provided the axes are acceptable.
- *Rotational defects*: It should be assumed that conservative treatment will lead to a certain level of internal rotational deformity of the distal fragment, since the arm is initially held in this position (arm sling, Desault bandage). However, we have never encountered a problem in clinical respects, since the most mobile joint of the body, the shoulder, can compensate for this defect. Posttraumatic derotational processes may also play a role. Protracted remodeling processes, e.g. as in ad latus deformities, can lead to *stimulation of length growth*, although the resulting length discrepancy is of no clinical significance.
- *Radial nerve palsy*: The spontaneous recovery rate for primary palsies is over 80% within the first 3 months after trauma.
- *Pseudarthroses* in children are rare, in contrast with the situation for adults.

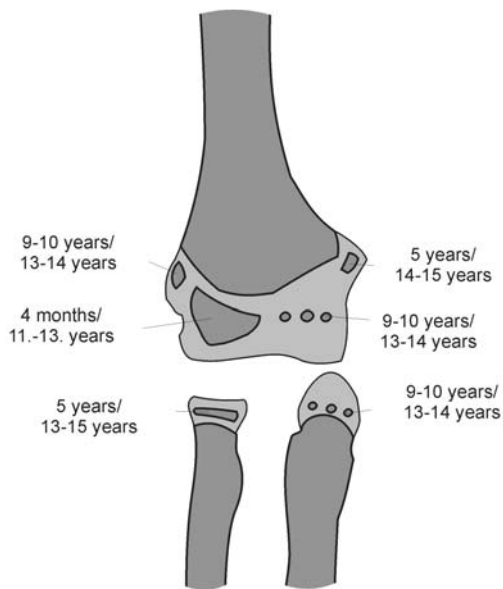


■ Fig. 3.502a, b. Treatment of displaced humeral shaft fractures: If (in polytraumatized patients, open fractures etc.) internal fixation is definitely indicated (rare), the external fixator is ideally suited for comminuted and long oblique fractures (a), while intramedullary nailing is more appropriate for short oblique and transverse fractures (b)

#### 3.5.7.5 Elbow fractures

- ❗ **After fractures of the hand and distal radius, fractures of the elbow are most frequently suffered by children and adolescents. However, since they often involve posttraumatic deformities and movement restrictions they are associated with the highest level of complications by far.**

## 3.5.7 · Fractures of the upper extremities



■ **Fig. 3.503.** *Ossification system of the elbow:* The most important epiphyseal ossification center is that of the capitulum humeri, which can be seen on an x-ray around the age of four months. The epiphyseal center of the radial head and the apophyseal center of the ulnar epicondyle appear around the age of five. The epiphyseal center of the trochlea, that of the ulna and the apophyseal center of the ulnar epicondyle appear – likewise together – between the ages of nine and 12. Between the ages of 11 and 13, the centers gradually fuse with the metaphysis, concluding with the apophyseal center of the ulnar epicondyle, the epiphyseal center of the radial head and the epiphyseal centers of the ulna

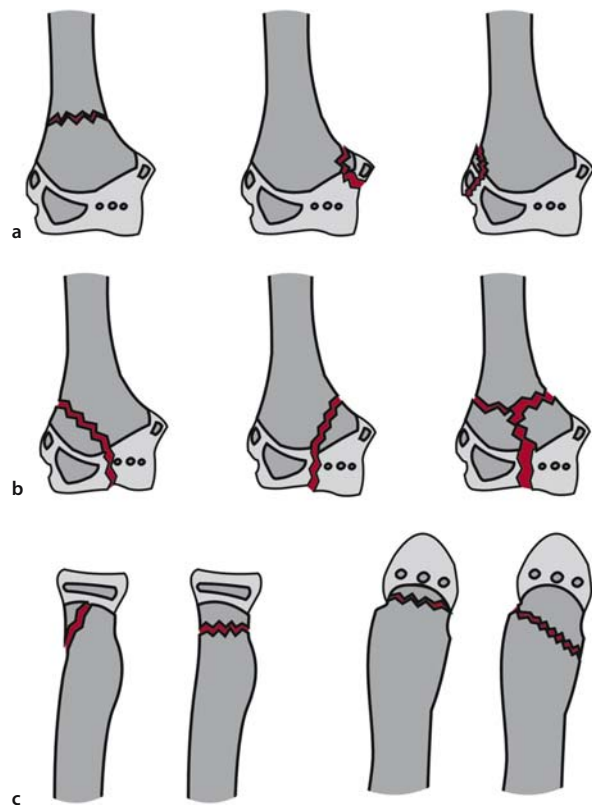
There are various reasons for this:

- The age-related – particularly in view of the various ossification systems – complex radiological anatomy (■ Fig. 3.503). Correct diagnosis is often a problem for unskilled practitioners, as evidenced by the numerous unnecessary side-comparing x-rays, which do not usually allow any conclusions to be drawn.
- The overlooking of radial head dislocations in connection with a Monteggia lesion.
- The biology of the radial head with posttraumatic circulatory disturbances.
- The biomechanics of the elbow, which, in the case of a fracture of the radial condyle, leads to progressive dislocation.

### Fracture types

We distinguish between the following types (■ Fig. 3.504):

- supracondylar humeral fractures (extra-articular),
- epicondylar fractures (extra-articular),
- transcondylar fractures (intra-articular),
- fractures in the area of the proximal end of the radius (extra-articular),
- fractures in the area of the proximal ulna (intra- and extra-articular).



■ **Fig. 3.504a–c.** *Fractures of the elbow:* **a** *Extra-articular distal humerus:* The commonest of all elbow fractures is the supracondylar humeral fracture (*left*). A fracture of the ulnar epicondyle (*center*) occurs more often in association with the elbow dislocation, while a fracture of the radial epicondyle (*right*) is a less common concomitant injury. **b** *Intra-articular distal humerus:* Of the joint fractures affecting the distal humerus, the fracture of the radial condyle is the commonest (*left*), followed by the rare ulnar condylar fractures (*center*) and transcondylar Y-fractures. **c** *Proximal forearm:* Almost as common as fractures of the radial condyle are those of the proximal end of the radius (*left*), while olecranon fractures (*right*) are fairly rare

### 3.5.7.6 Supracondylar humeral fractures

#### Occurrence

Supracondylar humeral fractures typically occur in 5- to 10-year olds and account for approx. 5% of all pediatric fractures, but 80% of all childhood elbow fractures [42]. Their equivalent in the over 10-year olds is an elbow dislocation.

The cross-sectional anatomy of the distal humerus is the reason for the high *complication rate* after displaced supracondylar fractures: Between the rounded contours of the condyles, the olecranon fossa is separated from the coronoid fossa only by a thin wall of bone. Even minor rotational deformities can lead to instability and slipping of the distal fragment into a varus deviation. This is aggravated by the fact that, even after an ideal reduction, the percutaneous insertion of fracture-cross-



ing, stabilizing wires is particularly demanding in view of the predominantly cartilaginous structure of the distal humerus.

❗ **Almost all complications occurring after supracondylar fractures are primarily iatrogenic in origin.**

### Diagnosis

#### Clinical features

A completely displaced fracture is usually accompanied by extensive swelling of the elbow. In hyperextension fractures, the sharp proximal fragment is displaced anteriorly into the brachialis muscle and subcutaneous tissues, producing an anterior subcutaneous hematoma, or even penetrating the skin in the case of an open fracture. Although a radial pulse is often not palpable, this is not necessarily a sign of a vascular lesion. In most cases the cubital artery is merely kinked over the proximal fragment. The same applies to the median nerve, which most often shows a primary deficit [13, 53]. Correct and complete documentation is essential, particularly in order to enable a primary neuropathy to be differentiated from a secondary, iatrogenic neuropathy. This includes the recording of unclear findings, which generally tend to be present in under 5-year olds.

We perform *Doppler ultrasound* only if the radial pulse is still not palpable after reduction. If no vascular signal is shown on the ultrasound scan then vascular revision is indicated.

#### Imaging investigations

The whole supracondylar area shows extensive intra-articular hemarthrosis after a fracture. Two thick fat pads are located at the front and back between the fibrous and synovial layers of the capsule, resulting in a contrasting »fat pad sign« on the x-ray in the event of the intra-articular accumulation of fluid.

Standard AP and lateral x-rays are arranged only if no obvious deformity is clinically apparent. In order to avoid unnecessary manipulations, the x-ray is recorded in this case with the arm in the most comfortable position. In

addition to the medial-lateral translation of the fracture, one should look for a flexion or extension deformity, particularly with fractures that are minimally displaced in the lateral projection.

❗ **Normally, a line extending distally along the anterior humeral cortex passes through the center of the capitulum in the ratio of 1/3 anterior to 2/3 posterior to the line.**

#### Fracture types

Three types of fracture can be distinguished, depending on the degree of displacement in each case, according to the most frequently cited Gartland classification [23]. The types most usually seen are an extension fracture with bending (type II) or complete displacement posteriorly of the distal fragment (type III):

- Type I non-displaced,
- Type II displaced but with preserved continuity of the posterior cortex,
- Type III complete displacement (no cortical contact).

❗ **Signs of a rotational deformity include a rotation spur or a difference in the AP diameter between the proximal and distal fragment on the lateral x-ray (Fig. 3.505).**

#### Differential diagnosis

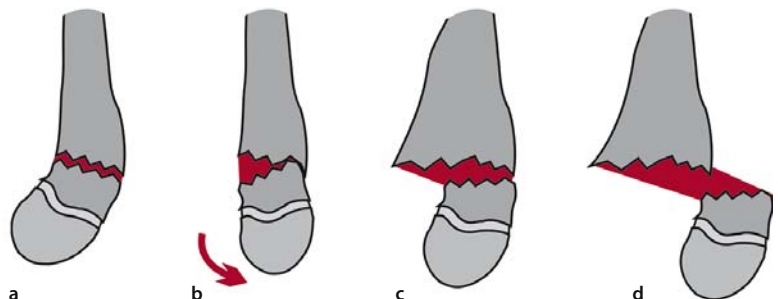
Supracondylar fractures must be differentiated from elbow dislocations and transcondylar fractures. The latter show a fracture line that crosses the growth plate in the lateral projection.

#### Treatment

##### Conservative

- Type I: Long-arm cast for 2–4 weeks, depending on the age of the patient. For initially non-displaced fractures, those at greatest risk of displacement are those in which at least one of the two condylar pillars is completely fractured. In this case, a check x-ray, without cast, is justified after 4–5 days.

❗ **Fig. 3.505a–d. Classification of supracondylar humeral fractures:** Since the rotational deformity and the resulting instability represent the central problem in these fractures, the only distinction required in such cases is between fractures without (a, b) and fractures with (c, d) rotational deformities



### — Type II:

Without primary rotational deformities: If a tolerable extension deformity is present according to the patient's age (see Prognosis/Spontaneous correction potential), a long-arm cast is fitted in the maximum tolerable flexion position and an x-ray, without the cast, is recorded 4–5 days later to rule out any secondary rotational deformity.

## Surgical

### Closed reduction

After closed reduction under anesthesia, the fracture is stabilized, unless a type II fracture without a primary rotational defect and/or unacceptable – in terms of age – extension deformity is involved.

The bony landmarks are often difficult to locate under the swelling, but this is usually possible if the elbow is flexed. An anatomically reduced fracture should satisfy the following criteria: The radial epicondyle is located dorsally in relation to the medial condyle. In the sagittal plane, the capitulum and trochlea are in 30–40° flexion. While flexion may be slightly restricted as a result of the swelling, it should be possible to approximate it to within approx. 10° of normal flexion subject to correct positioning in the sagittal plane and, in particular, correction of the rotational deformity.

As regards the elbow axes, the extended unaffected arm should be used for guidance purposes since considerable individual differences exist. The reduction maneuver starts with gentle traction in order to free the proximal fragment from the anterior soft tissues. If this proves unsuccessful, the brachialis muscle must be massaged away from the bone with »milking« movements in a proximal to distal direction [2]. We then eliminate the mediolateral translation while maintaining traction by holding the condylar block between thumb and forefinger. The rotational deformity can be corrected by supinating the forearm in a case of posterolateral displacement (and vice versa), during which the elbow is placed in increasing flexion by pressure exerted on the olecranon. This position is retained for subsequent stabilization.

### Open reduction

Open reduction is by no means simpler than the closed procedure. It is indicated only in the event of a vascular lesion requiring revision and after a failed closed reduction, which applies in approx. 10–20% of all type III fractures [2, 14].

### Stabilization

Unstable fractures, i.e. those with rotational deformities, should be stabilized surgically by a percutaneous method. The following options are available in order of decreasing stability: descending flexible nailing, crossed Kirschner wires inserted from the radial and ulnar sides, Kirschner

wire fixation from the lateral side [44, 78]. In the case of fixation from the ulnar side, a small incision should be made to check that the nerve is not directly located at the entry site to rule out the possibility of any iatrogenic ulnar neuropathy.

**! The younger the patient, the more likely it is that a constitutional anterior subluxation of the ulnar nerve occurs during flexion of the elbow, i.e. in the reduction position [77].**

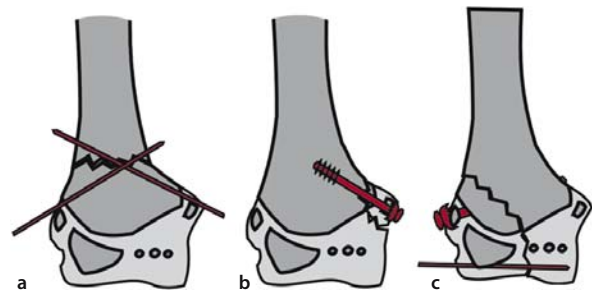
Fractures that are difficult to stabilize, particularly those with substantial metaphyseal comminution and extensive soft tissue damage, require alternative methods, e.g. the unilateral external fixator (■ Fig. 3.506, 3.507).

### Timing

The taboo of the delayed management of type III fractures is increasingly being called into question. Neurovascular complications are not more likely to be observed as a result of post-primary management, i.e. within 24 hours [58], provided there are normal neurovascular findings and repeated in-patient assessments. Since most complications after supracondylar humeral fractures are iatrogenic, these should ideally be managed by a well-rested, skilled team, but this is unlikely to be the case under emergency conditions.

### Follow-up controls

Once the function and elbow axes are the same on both sides, treatment can be considered as concluded. Long-term monitoring is only justified if there is a bony deformity in the sagittal plane, in order to verify spontaneous correction in younger patients or, in older patients, to discuss the possibility of a subsequent corrective osteotomy, depending on the persistence of functional restriction.



■ Fig. 3.506a–c. Treatment of distal humeral fractures. a Displaced supracondylar fracture with rotational deformity: All fractures with a rotational deformity should be reduced and stabilized with crossed Kirschner wires. b, c Epicondylar fractures: If a displaced epicondylar fracture – more commonly ulnar (b), less commonly radial (c) – occurs in connection with a dislocation (or also in isolation), it should, where possible, be managed by screw and washer fixation in order to facilitate spontaneous functional follow-up treatment



■ **Fig. 3.507a, b.** 4-year old boy with completely displaced supracondylar fracture (Gartland type III). **a** AP and lateral trauma x-rays: Complete periosteal tearing and shortening deformity. The proximal fragment has penetrated the brachial muscle into the subcutaneous tissues. **b** Intraoperative image converter view in two planes after closed reduction and crossed mediolateral Kirschner wiring and anatomical reduction. In relation to stability, note the correct position of the wires, which cross at a point proximal to the fracture

### Complications

— *Movement restriction.*

❗ **Even in ideal cases it can often take over 6 months before normal elbow mobility is restored. Premature physiotherapy usually proves to be useless or even counterproductive.**

— A *rotation spur* as an impediment to flexion remodels even in older children. More persistent cases of movement restriction are usually attributable to underlying incorrect bone healing. Translation deviations and deformities in the sagittal plane correct themselves according to the age of the patient. Common anterior bowing deformities of up to 20° can be left to correct themselves spontaneously in patients under 6 years of age. While little is known about the course of rotational deformities, they do not play a major role in functional respects as the shoulder is able to compensate for them. Spontaneous correction, which can take more than a year, should be awaited in children under 6. The possibility of a supracondylar correc-

tive osteotomy should be discussed in older patients if troublesome restriction is present. Stable fixation methods, e.g. an external fixator, with early functional, independent follow-up management are preferred. If a movement restriction is capsular in origin, we perform a distraction arthrolysis: In this procedure a joint-bridging external fixator stretches the humero-ulnar joint, while the joint is mobilized with the aid of physiotherapy.

— *Growth disturbances* are vanishingly rare. Deformities in the frontal plane, particularly cubitus varus, do not correct themselves, regardless of age, as growth continues.

— *Cubitus varus* remains the commonest complication after supracondylar fractures, with an incidence of up to 20% [13, 16, 37]. The underlying cause is inadequate elimination of the rotational defect and/or inadequate stabilization (■ Fig. 3.508). The unsightly cosmetic appearance is still the commonest indication for a corrective osteotomy, although sporting children (e.g. gymnasts, javelin throwers) can also feel func-



■ **Fig. 3.508.** Consequences of a rotational deformity after a supracondylar humeral fracture: Female patient with distinct cubitus varus after a supracondylar humeral fracture on the left arm. This deformity occurs as a result of the lateral tilting of the distal fragment towards the ulnar as a result of instability caused by a rotational defect

tionally restricted. In rare cases, a cubitus varus can be associated with a troublesome catching of the triceps tendon, a secondary ulnar neuropathy, an elbow or even a shoulder instability [1, 29, 62].

- **Neuropathies** usually show remission during the first 3 months after trauma.
- **Compartment syndrome:** The introduction of percutaneous fixation methods has dispensed with the need to place the elbow in a cast in a position of hyperflexion. As a result, the complication of serious ischemic contractures (Volkmann contractures) after isolated supracondylar fractures is no longer encountered. The risk of a compartment syndrome is significantly increased if a supracondylar fracture is combined with an ipsilateral forearm shaft fracture [4].

### 3.5.7.7 Epicondylar humeral fractures

#### Occurrence

The *medial epicondylar ossification center* appears as early as 4 years of age and is the last of the 4 ossification centers in the distal humerus to fuse, at the age of approx. 15. After supracondylar, lateral condylar and radial head fractures, a medial apophyseal avulsion fracture is the fourth commonest type of elbow fracture encountered in children, at an average age of approx. 11 years [22]. Avulsions of the *radial epicondyle* are rare. Ossification only starts at the age of 10–11 years.

#### Diagnosis

##### Clinical features

While the examination of medial joint stability is out of the question in the awake patient, the functioning of the ulnar nerve definitely must be clinically examined and documented.

##### Imaging investigations

Standard AP and lateral x-rays of the elbow. In younger children, the *medial* apophysis lies within the capsule, whereas the fracture line in the more frequently affected older patients always runs outside the joint, which means that the radiological sign of an elbow hemarthrosis, i.e. the fat-pad sign, cannot always be expected. A mere widening of the growth plate is often only apparent with non-displaced fractures. Usually, the epicondyle is displaced distally as a whole.

The physiological shell-like appearance of the *radial* epicondyle occasionally results in the incorrect diagnosis of a radial apophyseal avulsion. Ossification starts on the lateral side, making the plate look as if it has come loose. This is aggravated by the fact that the ossification can occur irregularly, giving the apophyseal center a fragmented appearance. The diagnosis of a radial epicondylar fracture can usually be substantiated only if clear soft tissue swelling is clinically and radiologically present or if the ossification center is distal to the lateral epiphyseal-metaphyseal junction.

##### Fracture types

- **Ulnar epicondyle:** The majority of the medial ligamentous apparatus, all of the flexor muscles and part of the pronator teres muscle originate from the medial epicondyle. Consequently, epicondylar fractures are equivalent to apophyseal avulsion fractures. They occur as isolated avulsions or, in around half of cases, in combination with an elbow dislocation. Fractures that appear to have occurred in isolation may have been preceded by a dislocation with spontaneous reduction that becomes visible at a later date in the form of periarticular calcifications. Other combinations are rare.
- **Radial epicondylar fractures** occur almost exclusively in connection with an elbow dislocation.

##### Treatment

- ! **With few exceptions, a conservative approach is indicated for epicondylar avulsions.**

##### Conservative treatment

Consensus on treatment prevails in the literature only in relation to non-displaced or minimally displaced (i.e. less than 5 mm) fractures: The cast immobilization should not continue for more than 3 weeks, and is followed

by independent elbow mobilization. Except in cases of incarceration, the radial epicondyle is managed conservatively with cast immobilization for approx. 2 to 3 weeks. [57].

### Surgical treatment

- Isolated epicondylar avulsions with more than 5 mm of displacement heal in the form of a pseudarthrosis in 50 percent of cases, which in itself is no indication for surgical stabilization. However, since stable screw fixation permits earlier and more active rehabilitation, this option should be discussed for youngsters who are keen on sports, particularly those involving throwing.
- The pseudarthrosis rate (approx. 10%) after open reduction and fixation can be minimized by freeing both the fragment and the avulsion site on the humerus of apophyseal cartilage with a sharp curette. Compression osteosynthesis with a small fragment cancellous lag screw on a toothed washer is preferable to Kirschner wires or absorbable pins.
- *Fractures accompanying elbow dislocations:* Epicondyles incarcerated in the joint represent an obstacle to reduction and therefore make open reduction a necessity [22]. Provided the fragments are not initially wedged, the guidelines for isolated avulsion fractures also apply in this case.

### Follow-up controls

Follow-up controls are continued until satisfactory mobility and confirmed joint stability are restored in the asymptomatic patient.

### Complications

- *Pseudarthroses* result in over 50 percent of cases after conservative treatment [22]. This rate is lower after surgical refixation.
  - ! **Since patients with or without an epicondylar pseudarthrosis are usually symptom-free, the primarily surgical treatment of the fresh avulsion does not greatly affect the clinical end result [21].**
- Cases of symptomatic pseudarthrosis can be managed by fragment trimming and fixation or excision with ligament refixation [25].
- *Movement restrictions* are rare after isolated avulsions and a short immobilization period, but are frequently encountered when the avulsion occurs in combination with an elbow dislocation.
  - *Neuropathies of the ulnar nerve* can be expected in 50 percent of cases involving fragment incarceration, but can also occur years after the trauma in cases of pseudarthrosis.
  - *Medial instability* with pseudarthrotic healing is rare and often leads to symptoms at a late stage [25]. Con-

sequently, medial joint stability should be thoroughly checked after the pain has subsided as soon as possible after the trauma.

- *Growth disturbances* do not occur since an apophysis is involved.
- *Deformities:* Doubling of the epicondylar contours and hypo- or hyperplasia of the epicondyle can occur regardless of treatment, but can usually just be classed as radiological phenomena [70].

### 3.5.7.8 Transcondylar humeral fractures

#### Occurrence

Around 5% of all pediatric fractures are intra-articular fractures of the distal humerus, over 80% of which involve the radial condylar pillar. Children between the ages of 4 and 8 are especially affected. At this age the trochlear ossification centers are not yet visible, and the capitulum radii is only ossified centrally. It can therefore be correspondingly difficult to establish the course of the epiphyseal fracture. In fact, distal, intra-articular humeral fractures are the ideal example illustrating the whole problem of the diagnosis of fractures that primarily involve the cartilaginous parts of the skeleton.

- ! **The challenge of condylar fractures lies in the correct identification of the fracture type. Failure in this respect can result, in extreme cases, in a pseudarthrosis that is difficult to treat (otherwise a rarity after pediatric fractures).**

#### Diagnosis

##### Clinical features

Swelling and hematoma over the lateral aspect of the elbow. Neuropathies are rare.

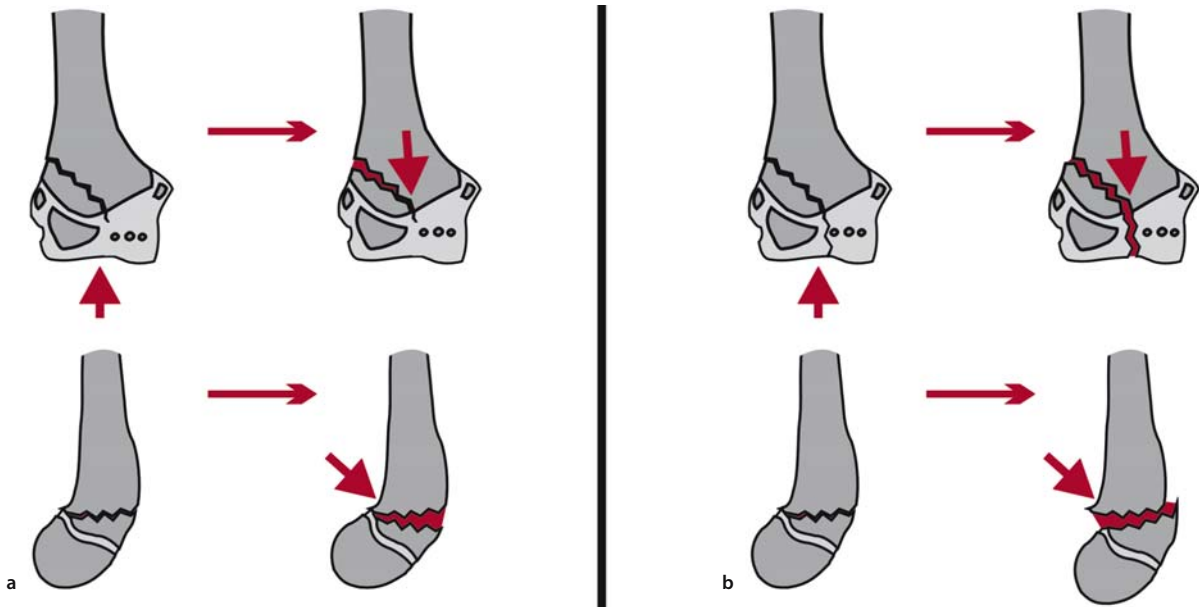
##### Imaging investigations

In addition to AP and lateral x-rays of the elbow, a view with internal rotation should be recorded if the findings are unclear.

##### Fracture types

The *radial condyle* is by far the most commonly affected. The fracture plane runs from a proximal-lateral-dorsal position in a distal-medial-ventral direction, corresponding to a Salter type IV epiphyseal-metaphyseal fracture. A crucial requirement for stability, and thus treatment and prognosis, is an intact cartilaginous bridge in the epiphyseal area: If the cartilaginous joint line between the trochlea and capitulum is intact, i.e. an incompletely intra-articular fracture is present, the fracture is stable. Although the lateral section may be hinged, the joint section remains in situ. Completely intra-articular fractures with rupture of the epiphyseal cartilage are unstable and therefore at risk of displacement.





■ **Fig. 3.509a, b.** Incomplete articular fracture of the radial condyle of the humerus (a): This so-called »hanging« fracture can be treated conservatively as secondary displacements are not expected. Complete articular fracture of the radial condyle of the humerus (b): The non-displaced,

complete articular fracture can even be displaced secondarily during cast immobilization and lead to pseudarthroses. The secondary displacement – as a sign of a complete articular fracture – must be identified as such and the fracture can then be managed surgically

❗ **If the widening of the central fracture sections at the cartilage-bone junction is less than 2 mm, it may be assumed that the cartilage is also intact.**

Employing this simple, radiological criterion, we have not missed any cases of primary or secondary displacement in our patients. The diagnosis based on the trauma x-ray is clear only if the fracture is completely displaced. If it initially appears non-displaced or minimally displaced, there is an approx. 15% probability that it will displace secondarily.

❗ **A cast-free check x-ray after 5–7 days is essential in order to check for secondary dislocation.**

*Ulnar condylar fractures and Y- or T fractures* are much rarer. The diagnostic considerations for radial condylar fractures also apply here (■ Fig. 3.509).

### Treatment Conservative

Non-displaced fractures undergo bone healing in a long-arm backslab, which is replaced by an encircling cast after the swelling has subsided, within 4–5 weeks (■ Fig. 3.510).

### Surgical

— *Radial condyle:* Primarily or secondarily displaced fractures are anatomically reduced in an open proce-

dure. We prefer a posterolateral approach. The posterior soft tissues over the condyle must be preserved so as not to jeopardize the circulation. Exposure of the posterior, metaphyseal fracture sections is essential for both the reduction and accurate implant placement. Compression osteosynthesis with a metaphyseal screw produces secure fixation, consolidation within 4 weeks and thus the prevention of partially stimulatory growth disturbances.

— Displaced *ulnar condylar fractures* are managed similarly from the medial side, and *Y- or T fractures* via a double incision or via a posterior approach.

### Follow-up controls

Consolidation x-ray after 4–5 weeks. Outpatient implant removal after 3–4 months. Follow-up controls for up to 2 years after the trauma are indicated in order to check for any growth disturbances. Full elbow mobility is usually restored only after several months. Physiotherapy is indicated only if movement restrictions persist for several months.

### Complications

— *Fishtail deformity:* After the distal epiphysis has ossified, one occasionally sees a central bony retraction in otherwise normal, but now more prominent, condyles. On the AP x-ray the distal humerus resembles a fishtail. Neither the origin nor the prognosis are



**Fig. 3.510.** Conservative treatment of a fracture of the radial condyle of the humerus: 5-year old girl with intra-articular non-displaced fracture of the radial condyle of the humerus. **a** Trauma x-ray. **b** Cast-free positional control during the first week with unchanged situation, the fracture gaps open on the radial and dorsal sides, but not centrally. This situation is interpreted as a »hanging« fracture and subsequently

treated conservatively. **c** After three weeks, the consolidation x-ray shows good incipient periosteal support, with a continued good fracture position. Start of spontaneous movement exercises. **d** At follow-up nine years later, clinical examination reveals identical elbow axes on both sides and free movement, while the x-rays show signs of slight condylar overgrowth laterally

clear, but it most probably involves local osteonecrosis resulting from instability during the consolidation phase [39]. We have never observed this change in shape in fractures that are stably fixed with compression screws [30, 31]. It is probably a radiological phenomenon with no serious long-term consequences.

#### — Growth disturbances:

- *Partially stimulatory growth disturbances* in the sense of unilateral stimulation during the consolidation phase occur in fractures with delayed healing and lead to a prominent condyle, also known as a *megacondyle*.
- A mild *cubitus varus* can occur as the result of a partially stimulatory radial growth disturbance after the delayed healing of a radial condylar fracture. The excessively large radial condyle often exaggerates the axial deformity. A more striking varization occurs after conservatively managed unstable Y fractures. Spontaneous correction cannot be expected. The indication for a corrective osteotomy is based on the patient's symptoms.

#### Posttraumatic deformities

Delayed healing with subsequent stimulated radial growth and a radial condyle that has consolidated too proximally, or even migrated proximally after pseudarthrosis, produce opposite effects on the elbow axis.

#### Pseudarthroses

Completely intra-articular fractures become displaced as a result of pressure exerted by the radial head following conservative treatment.

**! An established pseudarthrosis can result in a cubitus valgus that progresses even in adulthood, with the risk of ulnar neuropathy and restricted mobility.**

Considerable dispute exists as to the indication, and particularly the appropriate age, for a revision of the pseudarthrosis.

Lateral *condylar necrosis* may occur after delayed fracture management, lateral approaches with extensive posterior soft tissue removal from the radial condyle, inadequate fracture reduction and premature implant removal.

### 3.5.7.9 Radial head and neck fractures

#### Occurrence

After supracondylar and condylar fractures, those of the radial head are the third commonest elbow fractures during growth. They account for 10–25% of all elbow injuries and primarily affect the age group between 4 and 14 years [54]. They are the result of an excessive valgus stress in elbow extension during a fall onto the hand.

❗ **The primary determining prognostic factor is the impairment of the blood supply via the periosteal radial neck vessels caused by the initial trauma or even iatrogenically through an invasive therapeutic procedure.**

This vulnerability means that open reductions, unnecessary manipulations and transarticular wire fixations should generally be avoided. Otherwise, possible complications include avascular necroses, loss of the radial head shape and serious functional restrictions, particularly in respect of movements with forearm rotation.

#### Diagnosis

##### Clinical features

Local swelling, tenderness, painful restriction of a forearm turnover movement.

##### Imaging investigations

Standard AP and lateral elbow x-rays. The angulation of the radial head is determined by measuring the angle between the joint surface (or growth plate) and the longitudinal axis of the radius and subtracting from the normal figure of 90°. Small fragments close to the epiphysis may indicate epiphyseal involvement. The diagnosis can be aided by oblique views or – if uncertainty still exists – an MRI scan.

##### Fracture types

While the physes are still open, *radial neck fractures* and *epiphyseal separations* – with and without a metaphyseal wedge – are the rule. *Epiphyseal fractures* (Salter types III or IV) are rare.

##### Additional injuries

These fractures frequently occur in combination with other fractures of the elbow (particularly those of the olecranon, radial condyle, proximal ulna and avulsion fractures of the epicondyles, as well as elbow dislocations) [68]. The *radial head fracture* is the crucial prognostic factor.

##### Treatment

❗ **A conservative, non-invasive or (if reduction is required) minimally-invasive approach is essential. This means: avoid at all costs transarticular wire fixations, screws or plate fixations on the still growing proximal radius.**

##### Remodeling potential

❗ **Although the proximal radial epiphyseal plate only accounts for 20% of the growth in length, it possesses an impressive potential for spontaneous correction of deformities. Mechanical factors may play a crucial role in this remodeling process, for example it can be activated by early independent mobilization.**

##### Conservative treatment

Simple immobilization in a long-arm cast is appropriate in the following cases:

- before the age of 10 with angulations of less than 50–60° and ad latus displacements of less than half the shaft width,
- from the age of 10 with angulations of less than 20°.

##### Surgical treatment

A *closed reduction with subsequent Prévot nail fixation* is indicated if these criteria are not fulfilled. In this procedure, the fragment is reduced as far as possible by the application of external finger pressure and a concurrent pronation/supination movement. Any residual deformity is corrected via an elastic medullary nail advanced into the radius from the distal end. The implant is advanced as far as the epiphysis, which is then reduced by rotating the angled nail end [27] (■ Fig. 3.511). The nail is left in situ until consolidation is concluded.

##### The »joystick method«

If the fracture is severely, or even completely, displaced, it may not be possible to reduce the small epiphyseal disk manually by finger pressure. Nevertheless, an open, traumatizing procedure can be avoided by piercing the fragment with a percutaneous 1.6 mm Kirschner wire (»joystick«) and patiently guiding the fragment back to its original site under image converter control.

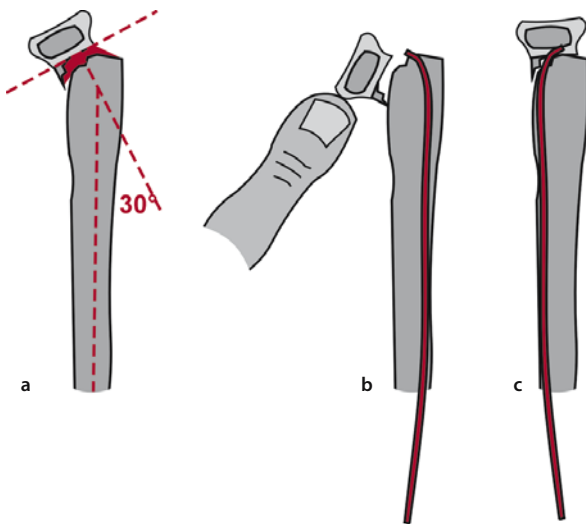
An *open reduction* is justified only in exceptional cases:

- complete displacement into the joint,
- after an unsuccessful, percutaneous reduction technique,
- epiphyseal, i.e. intra-articular fractures [48].

##### Follow-up management and controls

❗ **A maximum of 2 weeks' immobilization, followed by independent mobilization with complete avoidance of physiotherapy are the key features of follow-up management.**

After intramedullary splinting, movements should be allowed as soon as possible within the limits of pain. A consolidation x-ray is recorded 2–3 weeks after the trauma. Clinical controls should be continued for up to 2 years after the trauma in order to check for growth disturbances and avascular necrosis.



**Fig. 3.511a–c.** Treatment of displaced fractures of the proximal end of the radius: Up to the age of 9/10 years, and in order to minimize the trauma to the proximal end of the radius, tilt angles up to a maximum of 60° can, and should, be left alone but followed up with early functional treatment (a). Tilt angles >30° in patients older than 10 and completely tilted and displaced fractures should be reduced (b). An intramedullary nail can prove helpful for the reduction. If the head can be placed on top of the bone – by closed or open reduction – fixation will not be necessary. The nail should be removed again during the operation (c)

### Complications

- *Movement restrictions* are rare after a conservative or minimally-invasive treatment [73]. Any significant residual restrictions predominantly involve the forearm turnover movements after open reduction, after necrosis and in association with synostoses.
- *Avascular necrosis*: Clinically insignificant changes in the shape of the radial head, including an enlarged or ungainly appearance, are commonly observed on x-rays and probably represent past episodes of vascular crisis [54]. A loss of shape with, in many cases, severe restriction of forearm turnover movements and cubitus valgus is indicative of avascular necrosis, which often only becomes apparent on clinical and radiological examination several months after the trauma.
- *Pseudarthroses, radioulnar synostoses, periarticular ossifications and growth disturbances* as a result of partial physal closures are rare, but might be expected primarily after severe elbow trauma or an invasive procedure [75].

#### 3.5.7.10 Olecranon fractures

##### Occurrence

Less than 1% of all pediatric fractures. The average age of the affected patients is 9 years. Displaced fractures occur almost exclusively after the age of 10 [28].

### Diagnosis

#### Clinical features

Local swelling and pain.

#### Imaging investigations

Standard AP and lateral x-rays. The interpretation of the AP views is aggravated by the overlapping of the distal humerus. It is important to distinguish between a normal apophysis and a fracture: The ossification center in the area of the triceps attachment appears around the age of 9 and may be divided into two centers. The thin, bright line should not be confused with a fracture, particularly during physal closure around the age of 14. By contrast, the cartilaginous apophyseal section can result in an underestimation of the degree of displacement associated with fractures [20]. In cases of uncertainty, the fracture pattern should be evaluated by *ultrasound*.

#### Fracture types

A non-displaced or minimally-displaced *olecranon fracture* is present in over 80% of cases. The following fracture sites are involved in order of decreasing frequency [19]:

- metaphyseal *transverse, oblique and longitudinal fractures*,
- *apophyseal separations* with and without a metaphyseal wedge (Salter types I and II),
- fractures through the apophysis: *intra-articular* (Salter type III equivalent) and *extra-articular* as an avulsion of the tip,
- *additional injuries* are decisive for the prognosis and occur in approx. 50 percent of cases, particularly as proximal radial fractures, radial head dislocations and elbow dislocations [19, 28].
- *stress fractures* occur in athletes, particularly gymnasts, in the form of widening of the growth plate or fragmentation of the epiphysal center.

#### Treatment

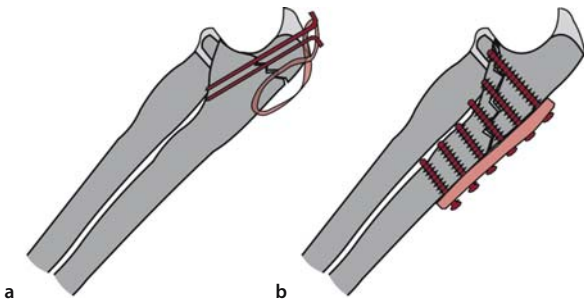
Fractures with less than 3–4 mm of displacement should be immobilized for 3–4 weeks in a long-arm cast. In most cases, fractures with greater displacement can, as in adults, be managed with tension-band wiring or, in the case of metaphyseal longitudinal fractures, by screw or plate fixation [28] (Fig. 3.512).

**! The detection and correct treatment of additional bone injuries, particularly of the proximal radius, are extremely important.**

#### Follow-up management and controls

From the functional standpoint, internally fixed fractures are managed at an early stage with independently implemented active and passive mobilization. Physiotherapy is indicated only if, after implant removal (3–4 months

## 3.5.7 · Fractures of the upper extremities



■ Fig. 3.512a, b. Treatment of displaced olecranon fractures: Displaced transverse olecranon fractures are (as in adults) managed with tension-band wiring (a). Oblique fractures are stabilized either with single screws or a one-third tubular or reconstruction plate (b)

postoperatively) a problematic movement deficit persists. Treatment is concluded when normal mobility is restored.

### Complications

The rare cases of moderate and poor results over the long term are based on *movement restrictions* and *axial deviations*, whether these occur after displaced fractures or as a result of concomitant injuries to the radial condyle or proximal radius [11, 19, 28]. *Growth disturbances* are not expected, even after wire piercing through the apophysis [28]. The rare cases of *pseudarthrosis* originate from underestimated displacements in younger children with fracture lines through the cartilaginous, radiologically invisible, part of the still minimally ossified olecranon. Minor intra-articular steps appear to remodel during the course of subsequent growth [38].

#### 3.5.7.11 Elbow dislocations

##### Occurrence

Elbow dislocations are rare during the growth phase. In contrast with supracondylar humeral fractures, which mainly occur between the ages of 5 and 8, dislocations do not usually happen until physeal closure, i.e. at the age of 13–14 years.

##### Diagnosis

##### Clinical features

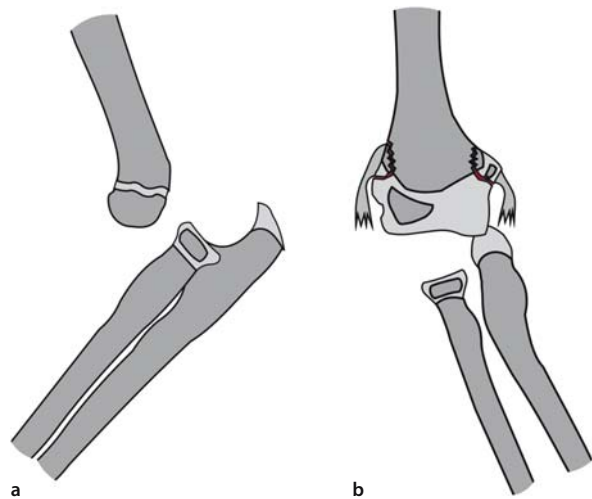
One objective of differential diagnosis is to rule out a supracondylar fracture. Since the swollen elbow is held in a semi-flexed position after both a fracture and a dislocation, only an x-ray, ideally in a lateral projection, can provide further diagnostic help (one plane is sufficient).

##### Dislocation type

A posterolateral dislocation with translation of the ulna and radius dorsally and laterally is the rule during childhood and adolescence. The physiological valgus position of the elbow promotes this direction of dislocation.

### Concomitant injuries

- *Concomitant ligament injury*: The elbow shows bone stabilization only in maximum extension. In all other positions, the joint is controlled by the capsule and collateral ligaments. Both the medial and lateral collateral ligaments are attached to the distal end of the ulna. The injury on the medial side inevitably involves either a ligament rupture and tearing of the flexor group or an avulsion fracture of the epicondylar apophysis (■ Fig. 3.513). Identical injury patterns on the lateral side are rarer.
- *Concomitant bone injuries* occur in over 50 percent of elbow dislocations, in decreasing order of frequency:
  - ulnar epicondylar avulsions,
  - proximal radial fractures,
  - radial condylar fractures,
  - fractures of the coronoid process,
  - radial epicondylar avulsions,
  - osteochondral fractures of the capitulum radii or the trochlea.
- *Concomitant neurovascular injuries* occur in 10% of dislocations. They are the result of forced ventralization of the distal humerus between the pronator teres and brachialis muscles into the subcutaneous tissues. This process primarily affects the ulnar and median nerves and the arterial anastomotic system on the medial side. Particular attention should be paid to cases with a concurrent ipsilateral forearm fracture because of the increased risk of concomitant vascular damage.



■ Fig. 3.513a, b. Concomitant injuries associated with elbow dislocation: lateral (a) and AP (b). The commonest concomitant injury associated with a, usually dorsally, dislocated elbow is a fracture of the ulnar epicondyle. At the same time, periosteal, chondral or bony avulsions of the radial collateral ligament very frequently occur, causing the ligament to slip in a dorsal direction. Habitual dislocations can occur if this injury is not recognized and the ligament refixed



### Treatment

Emergency reduction under anesthesia is simple and possible with gentle manipulation. A proven technique for closed reduction is to apply slight traction by pressing down on the anterior proximal forearm with the elbow flexed at 70–80° while simultaneously applying pressure to the tip of the olecranon with the other hand. If the ulnar epicondyle is incarcerated in the joint gap, which only occurs in around one dislocation in ten, then open reduction and internal fixation are indicated. In such situations, particular attention must be paid to the ulnar nerve, which is associated with a much higher deficit rate than after uncomplicated dislocations. If a medial epicondylar avulsion is combined with a lesion of the median nerve, the nerve usually becomes trapped, making open exploration essential [66].

After reduction, posterolateral stability must be checked by applying valgus stress and slight axial pressure to the forearm with the elbow slightly flexed. Only if there is a tendency to redislocate is open revision of the lateral ligamentous apparatus required, during which apophyseal avulsions or proximal ligament tears are found. Refixation with a screw is worthwhile in all cases in order to facilitate early functional follow-up treatment. Otherwise, the guidelines for isolated epicondylar fractures also apply to the treatment of apophyseal avulsions.

### Follow-up management

After *closed reductions* the elbow is immobilized for only two weeks in an above-elbow backslab, followed by independently performed active and passive elbow mobilization. After *open reduction* and screw refixation the elbow can be exercised from the very first days after the operation, under the guidance of the physiotherapist in the case of adolescents. If concomitant fractures are present, a consolidation x-ray is recorded four weeks after the trauma. Any implants can be removed after 2–3 months.

### Prognosis and complications

- *Growth disturbances* are possible in relation to concomitant fractures of the proximal radius.
- *Movement restrictions*: While early functional treatment reliably prevents any significant movement restrictions, experience suggests that a residual terminal extension deficit of 10–15° often remains [36].

❗ **If improvements in mobility are unexpectedly delayed or if the joint locks up repeatedly, the possibility of an overlooked (oste)chondral fracture should be considered [34].**

- The prognosis for *neurological problems* is good, as these usually involve neurapraxia, which recovers completely within a few weeks or months.
- *Heterotopic calcifications* are often observed in the capsule and ligaments. Fortunately, however, their im-

pact on mobility is minimal, in contrast with the rare condition of *myositis ossificans* which chiefly affects the brachialis muscle.

- *Radioulnar synostoses* often occur in association with concomitant fractures of the radial neck, regardless of the primary treatment.
- *Recurrent dislocations* are rare and require complex reconstruction procedures. Recurrences in the first two weeks after primary reduction are the result of the failure to detect a posterolateral instability after reduction and the omission of the subsequent lateral ligament revision. However, this situation can be rectified without adverse effects at this post-primary stage.

### 3.5.7.12 Radial head dislocations (Monteggia lesions)

#### Occurrence

Radial head dislocations are rare and occur mainly in the age group of 7 to 10-year olds. The existence of isolated dislocations, i.e. without a concomitant fracture of the ulnar, is greatly disputed [50].

❗ **The primary treatment of a radial head dislocation is simple and produces very good results. Overlooked dislocations, on the other hand, are very prone to complications, and their outcome after reconstruction is often uncertain.**

#### Diagnosis

##### Clinical features

A significant change in contours is lacking in the case of a ventral dislocation. Painful palpation maneuvers should be avoided.

##### Imaging investigations

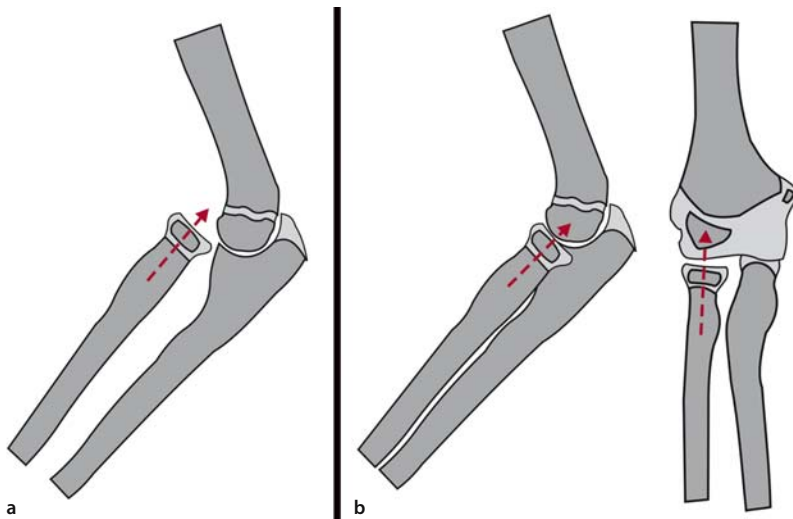
Radial head dislocations are detected only if:

- the orthopaedist insists on x-raying the elbow and the wrist in two planes in the event of a forearm shaft fracture,
- the position of the radial head in relation to the capitulum radii is correctly evaluated. Normally, the axis of the proximal radius projects onto the center of the capitulum. On the AP view, with the forearm pronated, this axis can be projected laterally onto the capitulum. On the lateral view, a line along the posterior ulnar cortex can help in identifying even slight deformations (■ Fig. 3.514).

#### Differential diagnosis

##### Congenital radial head dislocation

Differentiating between a congenital and traumatic etiology can prove difficult. The following factors suggest the presence of a congenital radial head dislocation:



■ Fig. 3.514a, b. *Diagnosis and treatment of radial head dislocation:* The axis of the proximal end of the radius must be centered over the middle of the capitulum humeri in all radiologically viewed planes (b). If this is not the case in one of the two x-ray planes (a), a radial head dislocation is present and must be reduced without delay

- lack of a trauma history,
- an excessively long radius,
- convex instead of concave shape of the proximal radial joint surface,
- bilateral occurrence,
- lack of deformation of the ulnar shaft.

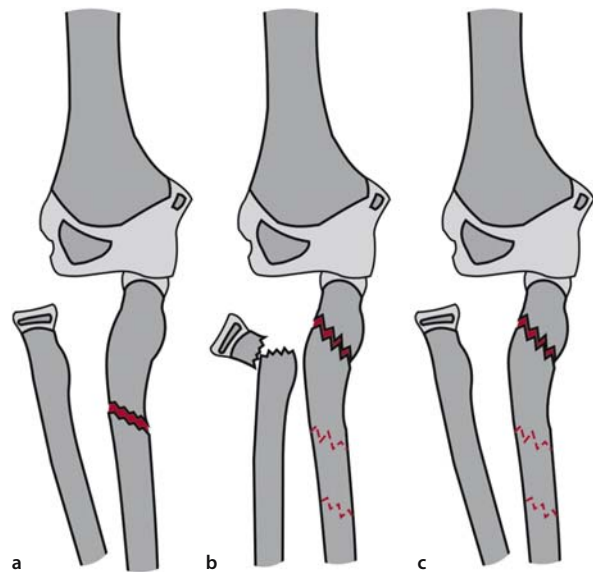
It should be noted that patients are often unable to recall any trauma and a dislocation is missed. In such cases the radius can continue to grow unhindered, the radial head changes its shape as a result of the missing joint partner and the ulnar shaft deformity can also remodel during the course of subsequent growth.

### Fracture types

The *classical Monteggia lesion* involves the combination of a dislocated radial head and an ulnar shaft fracture. The directions of the ulnar shaft deformation and the radial head dislocation correlate. The standard *classification* was proposed by *Bado* [3] (■ Fig. 3.515).

- *Type 1:* Extension deformity of the ulna, anterior dislocation of the radial head. Frequency: approx. 75% of cases.
- *Type 2:* Flexion deformity, posterior dislocation. Frequency: approx. 15% of cases.
- *Type 3:* Lateral dislocation. Frequency: approx. 5% of cases.
- *Type 4:* As for type 1, but additionally with proximal radial shaft fracture. Frequency: approx. 5% of cases.

So-called *Monteggia equivalents* are ulnar fractures in combination with radial head or neck fractures. In most cases the transition from the proximal to middle third of the ulnar shaft is fractured, less frequently the center



■ Fig. 3.515a, b. *Types of Monteggia lesion:* Apart from the classical Monteggia fracture (a), olecranon fractures with a radial head dislocation (b) and olecranon fractures with radial head dislocation (c) are considered to be Monteggia lesions, in addition to the radial head dislocation in isolation ( Fig. 3.514)

of the shaft, and rarely the distal or proximal thirds. The latter can even involve the humeroulnar joint intra-articularly or the olecranon extra-articularly. With increasing age, the ulna may merely suffer plastic deformation, a greenstick fracture or may be completely fractured. The radius is also fractured in rare cases. A slight bowing of the ulna is frequently overlooked, as a result of which the radial head dislocation also tends to be missed.

**!** This wide variety of injury patterns means that imaging investigations covering the wrist to the elbow are essential in all forearm fractures.

*Neurological concomitant lesions* are primarily associated with lateral dislocations, but can also occur with the other types. They usually involve cases of neurapraxia and show high spontaneous remissions rates.

### Treatment

The elimination of the bony deformity of the ulna is a precondition for stable reduction of the radial head. Since, in a case of a plastically deformed ulna or greenstick fracture, the elastic recoil force of the ulna usually prevents a reliable reduction of the radial head, completing the fracture is recommended.

**!** Full correction of the ulnar deformity in all planes is essential!

In most cases the correct position can be secured with an intramedullary flexible nail. Plate or screw fixation may be needed for very proximally located ulnar fractures or for rare multifragmented fractures. Closed reduction of the radial head by external manual pressure is usually successful, but can be obstructed by an interposed annular ligament or osteochondral shear fragments. A purely conservative approach, i.e. dispensing with ulnar stabilization, involves a fairly high risk of recurrent dislocation, since the ulna readily reverts to the primary abnormal position.

### Follow-up management

After internal fixation, spontaneous movement should be started within two weeks. A positional check after one week is advisable. A consolidation x-ray is recorded after 5–6 weeks, and implant material is removed after approx. 6 months.

### Complications

— Chronic radial head dislocation:

The proportion of missed dislocations cannot be determined as many patients may remain asymptomatic. At an interval ranging from a few weeks to several months, the head can often be reduced manually after osteotomy of the ulna. Possible problems can arise after more prolonged intervals, including restricted flexion as a result of the mechanically obstructing anteriorly dislocated radial head, restricted forearm pronation/supination, pain over the lateral elbow compartment or a cubitus valgus with/without ulnar neuropathy.

In such cases, the success rate of a surgical humero-radial joint reconstruction essentially depends on the appropriateness of the indication: An important factor is the shape of the radial head. If the joint surface is

already convex or if cartilage damage is present on the capitulum or radial head, the prospects of success are poor. On the other hand, good correction can be achieved for an excessively long radius or a deformity of the ulna. A proximal ulnar shaft osteotomy with an empirical search for the required degree of correction is a reliable way of achieving the objective. Empirical because the original deformity of the ulna has usually remodeled, at least partially, and also because a slight overcorrection is generally required to achieve stable head retention.

An ulnar external fixator can be helpful in this connection, since it facilitates the search for the correct adjustment of the ulnar osteotomy, the surgeon can test all movement combinations with the benefit of an open view of the radiohumeral joint, early functional follow-up treatment is possible and an excessively long radius can be compensated for by callus distraction of the ulna [32].

— *Periarticular ossification, myositis ossificans and radio-ulnar synostoses* can occur in isolated cases, particularly if there was severe initial trauma with substantial soft tissue damage, after an open surgical procedure or after repeated manipulations.

### 3.5.7.13 Forearm shaft fractures

#### Occurrence

Less than 5% of all pediatric fractures. The incomplete fracture types (greenstick, bowing fracture) dominate in under 10-year olds, and complete fractures in over 10-year olds. Fractures of the middle third are around 10 times more common than those of the proximal third [64].

#### Diagnosis

##### Clinical features

Well-documented checking of all 3 main nerve trunks and the radial pulse goes without saying. Failure to perform these checks will make it impossible to differentiate between a traumatic and an iatrogenic neuropathy. Firm swelling, unexpectedly intense pain, a high consumption of analgesics and exacerbation of pain on passive finger extension are clear signs of a compartment syndrome.

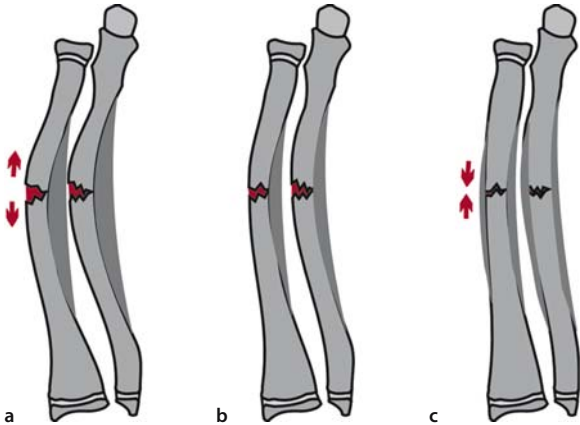
#### Imaging investigations

If a forearm deformity is clinically evident and reduction is clearly indicated, an x-ray in a single plane will suffice. The elbow and wrist must be included in an x-ray of *any* forearm shaft fracture so that concomitant injuries can be detected. In most cases, the latter are more crucial in terms of prognosis than the actual shaft fracture.

### Fracture types

#### Greenstick fractures

- *Compression fractures* («buckle fractures») in under 5-year olds show bending of the convex cortex and bulging of the concave cortex. These fractures are stable and not at risk of refracture.



■ **Fig. 3.516a–c.** *Healing mode of greenstick fractures:* If, in a diaphyseal greenstick fracture, the cortex on the concave side of the invariably present axial deviation is intact, periosteal bridging of the fracture gap occurs on the concave, but not the convex, side. This unbalanced consolidation involves the inherent risk of a refracture. If the axial deviation is greater, the amount of healing callus is increased and the risk of refracture is reduced, but functional problem is more likely because of the axial deviation (a). If the fracture is only «straightened», i.e. there is only slight axial deviation with preserved cortex on the concave side, the callus support is reduced and the risk of refracture increased, while the risk of functional restriction is correspondingly diminished (b). Only if the cortex on the convex side is approximated can the fracture consolidate adequately on all sides, and only then will the risk of further refracture be eliminated (c)

- *Bowing fractures:* fractures with mere bowing of both cortices are likewise stable.
- *Classical greenstick fractures* combine a bowing fracture on the concave side and a complete fracture on the convex side. The elastic resilience of the bowing side places the convex side under tension, which explains the tendency toward primary or secondary deformity, delayed healing and thus refracture (■ Fig. 3.516).
- *Complete fractures* can affect the ulna or radius in isolation or both at once. Combinations with greenstick fractures are possible (■ Fig. 3.517).

### Treatment

#### Spontaneous corrections

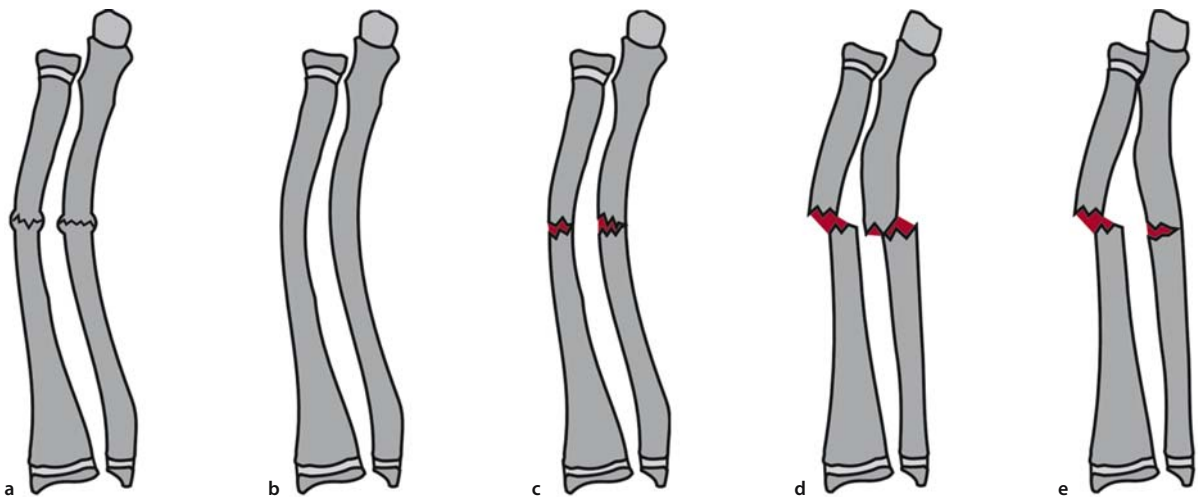
Ad latus deformities even out almost completely by adolescence.

- ! **The capacity for the spontaneous correction of axial deformities of the forearm shaft is relatively modest (10–20°) and limited to the age group of under 5-year olds. Since axial deformities of more than 10° usually lead to restriction of the forearm turnover movement, active correction is indicated [35, 56].**

#### Conservative treatment

The primary treatment involves the fitting of a long-arm backslab with neutral rotation of the forearm and a cast check after 24 hours and closure of the cast after the swelling has subsided, usually after 4 days.

- ! **An encircling cast should not be fitted to small children with stable fractures as this avoids the need to use the cast saw on the child when it comes to the removal.**



■ **Fig. 3.517a–e.** *a–c: types of diaphyseal fractures of the forearm: Greenstick fractures:* A distinction is made for diaphyseal greenstick fractures of the forearm between «compressed» (a) «bowed» (b) and «classical» (c) fractures. *d, e: types of diaphyseal fractures of the forearm: complete*

*fractures:* In these fractures all of the cortices of both bones are fractured (d), although mixed forms with a greenstick fracture of one bone and a complete fracture of the other can also occur (e)

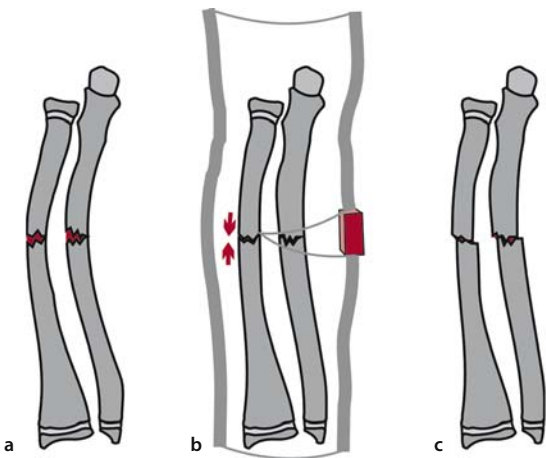
As a result of the trend toward surgical intramedullary stabilization a good plastering technique has been forgotten in many places. Shaping the cast to the forearm with an ellipsoid cross-section and the avoidance of excessive padding produces an excellent fit with reliable immobilization, thereby relieving pain and resulting in a reduced rate of secondary displacements. A good fit is particularly important for fractures of the proximal third.

Indications for a purely conservative primary approach:

- **Compression fractures**
  - *Under 5-year olds:* Fractures with axial deviations of less than 20°, excluding the classical greenstick fracture.
  - *Over 5-year olds:* Fractures with axial deviations of less than 10°, excluding the classical greenstick fracture.
- **Cast wedging:** A closed reduction is performed as a direct manipulation under regional or general anesthesia, or as an indirect manipulation in the form of cast wedging without anesthesia. Cast wedging in fractures of the proximal third of the shaft and isolated radial or ulnar fractures is less effective than in fractures of the radius and ulna in the middle third of the shaft (■ Fig. 3.518).

Indications for reduction (without subsequent intramedullary stabilization) – cast wedging:

- Radial and ulnar fracture with up to 20° axial deviation. For complete fractures, axial deviations of up to 10° are acceptable after cast wedging.



■ Fig. 3.518a–c. Treatment of diaphyseal greenstick fractures: Greenstick fractures always show axial deviation (a). Cast wedging on the 8th day after the accident can be employed in an attempt to place the completely fractured cortex on the opposite side under compression (b). If this fails, the cortex on the unaffected side of the greenstick fracture should be broken and both fractures reduced on a different day (c)

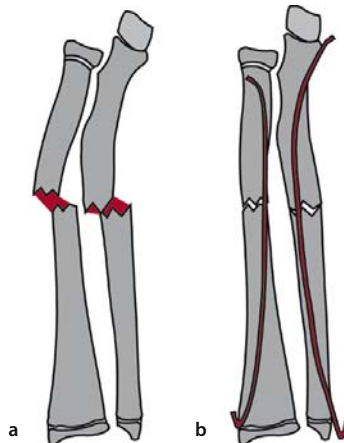
- In a greenstick fracture, compression of the fracture gap on the convex side is required, otherwise the closed reduction under anesthesia will need to be followed by fracturing of the concave cortex and Prévot nailing.

A check x-ray is recorded after cast wedging. If the residual deformity is unacceptable, closed reduction under anesthesia is recommended.

### Operation

- **Closed reduction under anesthesia**
  - Axial deviations of over 20° in under 5-year old children with a radial and ulnar fracture. Minor residual deformities noted on the positional check x-ray one week after reduction can be subsequently corrected with cast wedging.
  - Greenstick fractures with over 20° of primary displacement or after a failed wedging attempt with less than 20° of primary deviation. In rare cases, the fracture proves to be unstable after fracturing of the cortex and must be stabilized with intramedullary nails.
- **Intramedullary stabilization with flexible titanium nails (Prévot or Nancy nails)** has developed into a standard method for unstable shaft fractures (■ Fig. 3.519).

- ! **A sufficiently long skin incision of approx. 1.5 cm is recommended when inserting the radial nail, as this allows the surgeon to spare the superficial radial nerve and avoids the need to extend the primary skin incision when the nail is subsequently removed.**



■ Fig. 3.519a, b. Treatment of displaced, completely fractured, diaphyseal forearm fractures: If the growth plates are still open (a), the current treatment of choice is still the intramedullary nailing of displaced, complete forearm fractures (b). The nails must be strong enough to allow functional follow-up treatment without cast immobilization



In approx. 10% of cases, the surgeon is unable to insert the nail in to the other fragment, either because the reduction is obstructed by interposed soft tissues or because the medullary canal has been displaced by comminuted fragments [55]. A limited approach directly over the fracture is sufficient for an open reduction. Since the stabilization of just one bone in a complete forearm fracture involves a risk of further displacements and secondary operations, we perform a primary intramedullary stabilization of both bones in fractures of the radius *and* ulna [44]. Postoperatively, the patient requires only a forearm cast to reduce the pain and ensure wound healing over 7–10 days. The nail should not be removed before 3–4 months.

*Indication for intramedullary stabilization:*

- Completely displaced forearm shaft fractures,
- Radial and/or ulnar fracture with more than 20° of primary deviation or more than 10° after cast wedging in over 5-year olds.
- Greenstick fractures that prove to be unstable after deliberate fracturing of the cortex.
- We perform *plate fixation procedures* only in children over 10–12 years of age with fractures at the distal metaphyseal-diaphyseal junction, which are too distal for nailing and too proximal for Kirschner wire fixation. Over the radius we prefer the cosmetically less conspicuous volar approach and make it with 2 screws per fragment. The implant is removed after 4–6 months.
- *Immobilization periods*  
After intramedullary nailing or plate fixation we apply a volar forearm cast for one week to reduce pain and ensure wound healing. The consolidation periods are as follows: at the age of 5 years – 3 weeks, 5–10 years – 4 weeks, 10–12 years – 5 weeks, >12 years – 6 weeks. In view of the refracture risk, contact and ball sports should be avoided for 4 weeks in each case after cast removal and implant removal.

#### **Follow-up controls, complications**

As soon as full function has been restored, treatment can be considered as concluded.

- *Movement restrictions* primarily affect the forearm turnover movement. Axial deviations of over 10° regularly lead to functional restriction and should therefore not be tolerated [56]. Secondary corrective osteotomies for axial deformities that have persisted for a long time often fail to produce any significant functional improvement, probably because the shortening of the soft tissues, particularly the interosseous membrane, proves obstructive. Terminal limitations of pronation or supination of up to 10° can also occur after correct axial healing and early functional treatment [52].

- *Posttraumatic deformities:* Although rotational deformities cannot be reliably avoided either with conservative treatment or nailing, they do not lead to a restriction in the range of pronation/supination [72]. On the other hand, axial deviations of over 10°, particularly if they are counterrotating or affect the proximal part of the shaft, can lead to functional restrictions. Severe deviations are also cosmetically conspicuous.
- *Growth disturbances:* While increased length growth can occur after shaft fractures, this is usually of no clinical significance [10].
- *Neurological complications* of iatrogenic origin most commonly involve the superficial radial nerve and occur after incorrect insertion or removal of the radial Prévot nail [55]. Ulnar neuropathies are rare [55]. If the nerve fails to regenerate over 3 months, revision and a possible transfer of the ulnar nerve in a ventral direction are indicated.
- *Compartment syndromes* are rare and require immediate release of the compartment.
- *Refractures* occur after incorrectly treated greenstick fractures in up to 50% of cases. The uncompressed convex side with delayed healing represents a target fracture point. Other reasons include excessively short cast immobilization, early implant removal or premature weight-bearing after implant removal. Overall, the refracture risk is approx. 5%. Most fractures occur within the first two months after cast or implant removal [7].
- *Pseudarthroses or delayed consolidations* are very rare. The ulna is usually affected, particularly if an excessively large nail diameter was chosen and the fracture gap opened up as the nail advanced into the distal fragment. Delayed healing on the convex side of greenstick fractures can occur in the event of persistent axial deviations, including fairly minor deviations.

#### **3.5.7.14 Distal forearm fractures**

##### **Occurrence**

With an incidence of 20–25%, these are the commonest fractures during childhood and adolescence.

- ! **The highly active growth plates and the multidirectional mobility of the wrist signify an outstanding potential for the spontaneous correction of deformities. On the whole, these are very benign fractures that can be induced to heal with little effort and a low rate of complications.**

##### **Diagnosis**

##### **Clinical features**

The presence of angulation, particularly in a volar direction, is sufficient to raise the suspicion of a fracture on

clinical examination. Particular attention should be paid to swelling and pain in the carpal tunnel area because of the possibility of a manifest or threatened acute carpal tunnel syndrome.

### Imaging investigations

Standard x-ray in two planes, although one plane may suffice if a deformity is clinically obvious. Epiphysiolyse, in particular, are often not displaced, or minimally displaced, and therefore have to be diagnosed on the basis of physeal widening. As with the »fat pad sign« for distal intra-articular humeral fractures, the borderline between the volar periosteum, which is pushed up as a result of bleeding, and the overlying pronator quadratus muscle is radiologically visible in distal radius fractures.

### Fracture types

*Compression fractures of the radius* merely show bulging of both cortices and are therefore stable. *Epiphysiolyse of the radius* show a metaphyseal fragment in 80–90% of cases (Thurston-Holland fragment), and thus correspond to a Salter type II fracture [9]. *Metaphyseal radial fractures* without growth plate involvement are rare, as are *epiphyseal fractures of the radius* (Salter types III and IV). The classical *Galeazzi fractures*, i.e. distal radial fracture in combination with radioulnar dislocation are very rare during growth. Instead of the dislocation, epiphysiolyse of the distal ulna can occur, which can be described as a *Galeazzi equivalent* [47]. Concurrent involvement of the *distal ulna* is observed in approx. 50 percent of distal radial fractures: The commonest lesion is an avulsion of the styloid, followed by metaphyseal fractures and, in rare cases, epiphysiolyse. Isolated epiphysiolyse of the ulna are also rare.

*Stress fractures* in the area of the distal radial epiphysis and growth plate are described particularly in female gymnasts. Repetitive high axial stresses during dorsal extension of the wrist and possibly delayed bone mineralization in high-performance athletes lead to local overloading. Growth disturbances of the radius with subsequent advancing of the ulna and signs and symptoms of ulnar impingement are possible consequences [71].

### Treatment

#### Spontaneous correction

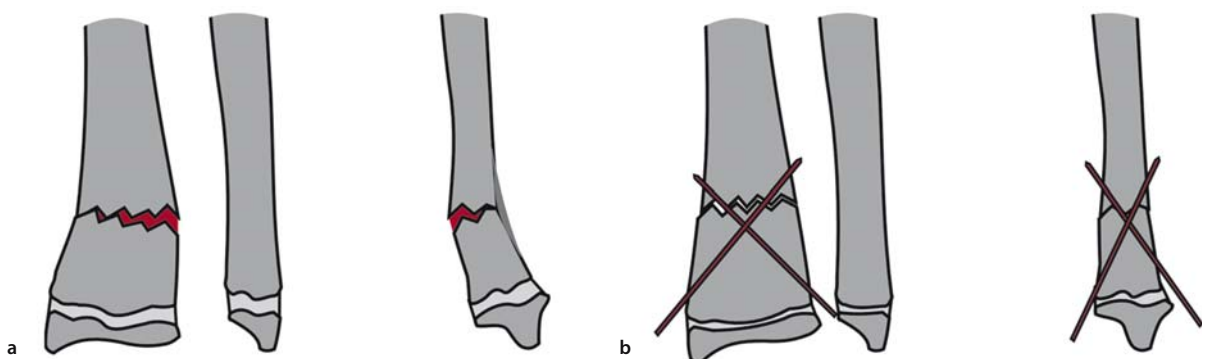
In addition to the correction resulting from subsequent growth, which takes several months, significant spontaneous reduction produced by mechanical factors is observed even after 1–2 weeks, particularly in cases of angulated radial epiphysiolyse.

**! The potential for the spontaneous correction of deformities of the distal forearm is substantial. This applies both to side-to-side displacements and axial kinks. Deformities in the sagittal plane of up to approx. 40° and in the frontal plane of up to approx. 30° correct themselves completely in girls under 10 years and boys under 12 years.**

Nevertheless, an attempt should still be made to reduce the extent of the deformity by cast wedging. If an unacceptable deformity still persists after cast wedging in a patient over 10–12 years, then closed reduction under anesthesia is indicated. Angulation in a volar direction is less amenable to spontaneous correction, but this is much rarer.

#### Conservative treatment

All fractures that are not completely displaced are initially immobilized in a long-arm cast (■ Fig. 3.520a). The sub-



■ Fig. 3.520a, b. Treatment of displaced fractures of the distal forearm. a Up until the age of around 10, axial deviations of up to 40° can sometimes be left to correct themselves spontaneously with further growth, although this should be discussed with the patient and the parents. The position can usually be corrected to less than 20° with

cast wedging. b A correct position should be attempted in children older than 10/12 years. If this cannot be achieved with cast wedging, the fracture must be reduced and, at this age, additionally stabilized with percutaneously inserted Kirschner wires

### 3.5.7 · Fractures of the upper extremities

sequent course of action depends on the stability of the fracture and the primary deformity:

- *Stable fracture, not displaced*, e.g. non-displaced compression fractures: No further check x-rays. Immobilization for 4 weeks.
- *Fracture at risk of angulation, primarily not displaced*, e.g. metaphyseal bowing fracture: Radiological positional check after one week. Secondary angulations are corrected by cast wedging.
- *Angulated fractures* are routinely reduced by cast wedging after 7–10 days. We perform a *closed reduction by cast wedging*, regardless of the patient's age, for all primarily angulated fractures. The outcome of the reduction after wedging is radiologically checked only in children older than 10 years, so that any unacceptable residual deformity can be reduced under anesthesia. In the case of epiphysealyses, displacements of up to half the shaft width can be tolerated.

#### Surgical treatment

- *Closed reduction under anesthesia and Kirschner wire fixation:*

In children over 10 years of age, completely displaced fractures, or fractures that are not adequately reduced by cast wedging, are reduced under anesthesia and stabilized by percutaneous *Kirschner wire fixation* in order to be certain of avoiding the need for secondary manipulations [24] (■ Fig. 3.520b). The wires are bent 90° approx. 1 cm above skin level and trimmed. Elevated circular cut-outs in the cast avoid any direct contact between the wires and the cast and thus prevent loosening and infection.

- *Plate fixation:*

Since angulated and displaced fractures at the *metaphyseal-diaphyseal junction* in adolescents cannot be adequately stabilized by Kirschner wires, we prefer a volar osteosynthesis with a small-fragment plate. Very rare *epiphyseal fractures* are reduced openly and stabilized by screw fixation in accordance with the principles applicable to joint fractures with more than 2 mm of intra-articular step formation. The *immobilization period* is four weeks for all fractures, with the exception of plated patients requiring early functional follow-up treatment.

#### Follow-up controls

Fractures with growth plate involvement should be followed up every six months for up to two years after the trauma in order to check for any growth disturbances.

#### Complications

- *Posttraumatic deformities* are conspicuous, particularly in cases of volar angulation. If angulated fractures are left to correct themselves spontaneously in younger children, the parents must be told that remodeling will

take from 6–12 months in order to avoid any unnecessary osteotomies [17]. On the other hand, the option of a corrective osteotomy should be considered in patients with little residual growth and consolidated deformities, depending on the functional and cosmetic impairment in each case.

- *Movement restrictions* are rare and usually the consequence of posttraumatic deformities in adolescents. A restriction in the forearm turnover movement can be expected in the event of consolidated ad latus deformities exceeding more than half the shaft width.
- *Growth disturbances:* Transient stimulation of radial length growth is usually of no clinical significance. The risk of a total or partial physeal closure after fractures with growth plate involvement is approx. 2–4% and applies both after the common condition of epiphyseolysis and the rare epiphyseal fractures of the radius.

The very rarely affected distal ulnar growth plate reacts much more sensitively. The risk of a growth disturbance in this case is 50% [9]. In a case of progressive deformation or shortening, and depending in each case on the patient's age, residual growth and the size of the closure, a bridge resection with fat interposition (Langenskiöld operation), a corrective osteotomy, a lengthening or shortening procedure, an epiphyseodesis or a combination of these methods should be considered.

- *Pseudarthroses* are the rule with avulsions of the styloid process, but are almost never symptomatic [9].
- *Acute carpal tunnel syndrome and compartment syndromes of the forearm* can occur in the event of the delayed management of epiphyseolyses or epiphyseolyses that are severely displaced in a dorsal direction. Intense pain, painful passive finger extension and, at a later stage, a median or ulnar neuropathy are warning signs that require immediate opening of the carpal tunnels, possibly supplemented by compartment release in the forearm [74].

#### 3.5.7.15 Fractures of the carpal bones

##### Occurrence

These are rare events, accounting for less than 0.5% of all pediatric fractures. The scaphoid is by far the most frequently affected bone, but occurs almost exclusively in adolescents.

##### Diagnosis

##### Clinical features

The pain in the »anatomic snuffbox« is a helpful indication in children as well. The volar radial tuberosity should also be palpated since fractures frequently affect the distal pole.

### Imaging investigations

- ❗ **If a scaphoid fracture is suspected, the standard AP and lateral projections should be supplemented by oblique views in supination and pronation-ular deviation. The latter are particularly valuable for the detection of fractures at the proximal pole.**

### Fracture types

#### Scaphoid

*Start of ossification:* at the age of 6 years in boys, 4.5 years in girls. *End of ossification:* 15 years in boys, 13.5 years in girls. The high cartilaginous proportion that persists for a long time ensures a high degree of elasticity up to adolescence, which explains the low incidence of fractures in children under 10 years of age.

Ossification progresses in a distal to proximal direction and is crucial for the fracture pattern: In contrast with adults, the *distal third*, often outside the joint on the volar side, is the most commonly affected and these fractures correspond to bony ligament avulsions [76]. Since fractures of the *middle third* involve a higher degree of force, the doctor should accordingly look for concomitant injuries to the carpus, metacarpus and distal radius and ulna. The rarest fractures are those at the *proximal pole*. Since this is the last part of the bone to ossify, such fractures probably remain undiagnosed not infrequently. Displaced fractures are rare.

*Fractures of the capitate, triquetrum, hamate, pisiform and lunate bones* are rare. They usually heal completely after 4 to 6 weeks of immobilization in a cast.

### Treatment

#### Scaphoid fractures

After initial immobilization in a long-arm cast that includes the thumb metacarpophalangeal joint, the situation is reassessed clinically after 7–10 days. If the findings are normal, treatment is considered to be concluded. If pain persists, further x-rays are taken and immobilization is continued for another two weeks even if the x-ray findings appear normal. If symptoms and normal radiological results still persist after these two weeks, an MRI scan of the wrist is indicated with the aim of ruling out any occult fractures, concomitant soft tissue injuries or tenosynovitis. A total immobilization period of 6–8 weeks is sufficient, except for initially overlooked and non-immobilized fractures. After two weeks, the long-arm cast can be replaced with a forearm cast [76].

### Follow-up controls

Once consolidation and free function have been achieved, the treatment can be considered as concluded.

### Complications

- *Pseudarthroses* after scaphoid fractures during growth are only mentioned in isolated cases in the literature [15] and are usually the result of overlooked fractures or excessively short immobilization periods. However, a lengthier period of immobilization will usually produce consolidation.

### 3.5.7.16 Fractures of the metacarpals and phalanges

#### Occurrence

Accounting for approx. 20% of all pediatric fractures, hand fractures represent the second commonest group after distal forearm fractures. The accident mechanism reflects the age-specific activity: small children typically suffer from entrapment injuries during their exploratory forays, while sports injuries become more frequent with increasing age, particularly in male adolescents [51]. Special attention must be paid to hand injuries in neonates or small children as these are a possible indicator of child abuse [59].

#### Diagnosis

#### Clinical features

- ❗ **Particular attention should be paid to axial deviations in the frontal plane and to rotational defects as spontaneous correction cannot be expected in these cases. Rotation is tested at 90° flexion in the metacarpophalangeal joints and the proximal interphalangeal joints. During normal rotation the fingertips converge in the direction of the scaphoid.**

### Imaging investigations

Dorsopalmar and oblique x-rays of the hand. In contrast with the long bones, all the metacarpals and phalanges possess only one epiphyseal plate which, with the exception of the 2nd–5th metacarpals, are always at the proximal end. A so-called pseudoepiphysis on the 1st distal metacarpal and, less frequently, the proximal 2nd–5th metacarpals, can be mistaken for a fracture. The same also applies to a congenital radial deviation of the little finger, also known as Kirner's deformity.

### Fracture types

The digits at either end of the hand, i.e. the thumb and little finger, are most frequently affected. Phalangeal fractures are more common than metacarpal fractures and the growth plate is also more likely to be involved, in up to 80% of cases, resulting in a Salter-Harris type II epiphyseal separation. The nature and site of the fractures show a clearly age-related pattern, with predominantly distal phalangeal fractures in 0- to 8-year olds, proximal phalangeal fractures of the little finger in 9- to 12-year

### 3.5.7 · Fractures of the upper extremities

olds and subcapital 5th metacarpal fractures in 13- to 16-year olds [65].

The following occur in decreasing order of frequency:

- *Metaphyseal compression fractures* are often not visible on the x-ray and are therefore often missed.
- *Epiphyseal separations*, usually with a metaphyseal wedge (Salter-Harris type II), should be reduced if necessary, except for epiphyseal separations of the 1st proximal metacarpal, which are occasionally irreducible by a closed procedure because of a periosteal »buttonhole« mechanism.
- *Volar lip fractures*, as small avulsions of the volar capsule plate, affect the proximal middle phalanx as a result of hyperextension trauma to the proximal interphalangeal joint, typically during ball sports such as basketball or volleyball.
- *Subcapital metacarpal fractures* occur as a result of punching, usually by male adolescents, against a hard object (boxer's fracture) with a flexion deformity of the head of the 5th metacarpal or, more rarely, the 4th metacarpal.
- *Shaft fractures*: Since the 3rd and 4th metacarpals are well stabilized by the suspensory effect of the deep intermetacarpal ligaments, severe shortening or displacements rarely occur.
- Fractures of the *phalangeal neck*, also known as supracondylar fractures, are usually highly unstable. The displaced phalangeal head can be confused with an epiphyseal separation.
- *Intra-articular fractures* account for less than 10% of all hand fractures and are seen almost exclusively in adolescents as transitional fractures (Salter-Harris type III) before physeal closure, either as a bony avulsion of the extensor tendon at the base of the distal phalanx or as a pediatric »Bennett fracture« of the 1st proximal metacarpal: the still open ulnar part of the epiphysis breaks away in the form of an ulnar ligament tear, with displacement of the radial epiphysis and shaft in a radial direction.

#### Treatment

The uniform sagittal movement plane of all finger joints also restricts the spontaneous correction potential accordingly to flexion and extension deformities. This is approx. 30° in under 10-year olds, gradually declining thereafter until expected growth plate closure at the age of 14 years in girls and 16 years in boys. The mobile 4th and 5th metacarpals even tolerate slight residual deformities. Deformities in the vicinity of the growth plates show the best remodeling outcome.

#### Conservative

Non-displaced fractures or those with an acceptable, i.e. remodelable, deformity, heal without complications with

simple immobilization. The latter should include at least two fingers, either in the form of a simple tape dressing for stable finger fractures or as a long finger cast for distal metacarpal fractures. The need for a correct functional position, with 70–90° flexion in the metacarpophalangeal joint and 10–20° in the interphalangeal joints, also goes without saying in children.

For subcapital fractures of the 5th metacarpal, closed reduction of flexion deformities of less than 30° does not offer any advantage over early functional treatment, even if the growth plate is closed. It should be noted, however, that the contours of the head of the 5th metacarpal may become less distinct during fist closure [41].

#### Surgical

##### Closed reduction

Extra-articular fractures with axial deformities in the frontal plane (ulnar or radial deviations) or rotational defects require a closed reduction and, if instability is present, fixation. An exception is the base of the 1st metacarpal, where the freedom of abduction and adduction can functionally compensate for slight deformities in the frontal plane. Only epiphyseal separations on the phalanges can be treated simply by reduction and cast immobilization as these are usually stable.

The fracture type and site determine the reduction technique. Thus, for example, flexed subcapital metacarpal fractures should be reduced with 90° flexion of the MP and IP joints, fully tensed collateral ligaments, and with the application of axial pressure on the proximal phalanx. In all other cases, and depending on the fracture pattern and the surgeon's preference, the outcome of the reduction should be stabilized with axial Kirschner wires, crossed Kirschner wires or a small fixator.

##### Open reduction and fixation

In addition to the above-mentioned fixations, osteosynthesis procedures with mini-screws and mini-plates represent an alternative for shaft fractures with a shortening tendency. Other indications are:

- unsuccessful closed reduction because of a soft tissue interposition,
- intra-articular fractures with correspondingly sized fragment and displacement; these are managed according to the guidelines for adult fractures,
- 2nd and 3rd degree open fractures.

##### Immobilization periods

For volar lip fractures, a maximum 7-day immobilization period is sufficient, followed by a finger tape dressing for 3–4 weeks during sport to guard against any further hyperextension trauma. Two weeks of immobilization is enough for metaphyseal compression fractures and non-displaced epiphyseal separations, while the consolidation



period for shaft fractures in adolescents can be up to 6 weeks.

### Follow-up controls

Positional check x-rays are indicated only for non-stabilized fractures at risk of displacement. Consolidation can be confirmed clinically on the basis of the absence of tenderness in the fracture area after 3–4 weeks. Further check-ups are not indicated once mobility has been restored as growth disturbances are rare.

### Complications

- *Growth disturbances and pseudarthroses* in the hand area are very rare. Isolated small fragments that have split off, particularly in the vicinity of the condyles, can heal as a pseudarthrosis, but are usually of no clinical consequence.
- *Movement restrictions* are not expected with short immobilization periods. Occupational therapy is indicated only in cases of delayed rehabilitation of surgically managed fractures in adolescents.
- *Posttraumatic deformities* can be safely avoided by the correct clinical recording of the rotational situation and observance of the limits of spontaneous correction potential.

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## 3.5.7 · Fractures of the upper extremities

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### 3.5.8 Tumors of the upper extremities

#### ➤ Definition

Bone and soft tissue tumors originating in the bony structures of the upper extremity or their surrounding soft tissues.

#### Occurrence

Around 20% of bone tumors in children and adolescents are located in the upper extremity, and around two-thirds of these in the proximal humerus (see ► chapter 4.5.1). No statistical data on soft tissue tumors are available.

Of the *benign tumors* that affect the scapula, clavicles, humerus, radius and ulna, the osteochondroma is the commonest, usually occurring in isolation, but occasionally in connection with »cartilaginous exostoses« (osteochondromas; ■ Table 3.83).

The commonest lesion of the proximal humerus is a unicameral bone cyst (■ Fig. 3.521). This site is particularly susceptible to this tumor-like lesion. It is, in fact, much more common than suggested by our statistical data, since a cyst at this site is not usually biopsied or treated

(■ Fig. 3.522). Another very common tumor is a chondroblastoma in the area of the humeral head. Aneurysmal bone cysts, in both the humerus and the clavicles and scapula, are also relatively common. An osteoid osteoma is also not infrequently seen in the humerus. Compared to the knee, however, non-ossifying bone fibromas of the upper limb are a rarity. The enchondroma is clearly the dominant tumor of the hand. In fact, almost half of all tumors of the hand are enchondromas and they very typically affect the phalanges in particular (■ Table 3.83, ■ Fig. 3.522).

Osteochondromas, aneurysmal bone cysts and osteoid osteomas also occur in the bones of the hand. All other tumors are rare. Osteochondromas on the forearm can be located at both the distal and proximal ends of the radius or ulna and often cause growth disturbances at these sites.

The dominant *malignant bone tumor* is the osteosarcoma, which particularly affects the proximal humeral metaphysis. The Ewing sarcoma is occasionally encountered in the humeral shaft area. Other malignant tumors and sites fortunately occur only as isolated cases.

The commonest *soft tissue tumors* are lipomas and fibromas, while desmoids are also not infrequently seen.

■ Table 3.83. Primary bone tumors of the upper extremities in children and adolescents (n=365) compared to adults (n=662). (Basel Bone Tumor Reference Center)

	Children and adolescents						Adults					
	Shoulder and upper arm		Forearm		Hand		Shoulder and upper arm		Forearm		Hand	
Osteochondroma	55	23.6%	13	24.1%	10	12.8%	26	10.3%	9	12.3%	9	2.7%
Chondromyxoid fibroma			1	1.9%	1	1.3%	1	0.4%	1	1.4%	1	0.3%
Chondroblastoma	12	5.2%			1	1.3%	3	1.2%			4	1.2%
Enchondroma	5	2.1%	2	3.7%	27	34.6%	41	16.3%	9	12.3%	225	66.8%
Aneurysmal bone cyst	19	8.2%	3	5.6%	10	12.8%	13	5.2%	6	8.2%	8	2.4%
Non-ossifying bone fibroma	8	3.4%	4	7.4%			1	0.4%	1	1.4%		
Giant cell tumor	6	2.6%	1	1.9%	1	1.3%	11	4.4%	17	23.3%	8	2.4%
Fibrous dysplasia	8	3.4%	4	7.4%	1	1.3%	7	2.8%	4	5.5%	4	1.2%
Osteoid osteoma/osteoblastoma	3	1.3%	7	13.0%	5	6.4%	6	2.4%	4	5.5%	9	2.7%
Other benign tumors	7	3.0%	6	11.1%	17	21.8%	25	9.9%	9	12.3%	27	8.0%
<i>Langerhans cell histiocytosis</i>	8	3.4%	1	1.9%			7	2.8%	3	4.1%		0.0%
<i>unicameral bone cyst</i>	45	19.3%	3	5.6%			1	0.4%	1	1.4%	1	0.3%
<i>Other tumor-like lesions</i>	12	5.2%	1	1.9%								
<b>Osteosarcoma</b>	<b>22</b>	<b>9.4%</b>	<b>3</b>	<b>5.6%</b>			<b>23</b>	<b>9.1%</b>	<b>3</b>	<b>4.1%</b>	<b>1</b>	<b>0.3%</b>
<b>Ewing sarcoma/PNET</b>	<b>9</b>	<b>3.9%</b>	<b>3</b>	<b>5.6%</b>	<b>1</b>	<b>1.3%</b>	<b>8</b>	<b>3.2%</b>	<b>1</b>	<b>1.4%</b>	<b>1</b>	<b>0.3%</b>
<b>Chondrosarcoma</b>	<b>9</b>	<b>3.9%</b>		<b>0.0%</b>	<b>2</b>	<b>2.6%</b>	<b>56</b>	<b>22.2%</b>	<b>4</b>	<b>5.5%</b>	<b>24</b>	<b>7.1%</b>
<b>Other malignant tumors</b>	<b>5</b>	<b>2.1%</b>	<b>2</b>	<b>3.7%</b>	<b>2</b>	<b>2.6%</b>	<b>23</b>	<b>9.1%</b>	<b>1</b>	<b>1.4%</b>	<b>15</b>	<b>4.5%</b>
<b>Total</b>	<b>233</b>	<b>100.0%</b>	<b>54</b>	<b>100.0%</b>	<b>78</b>	<b>100.0%</b>	<b>252</b>	<b>100.0%</b>	<b>73</b>	<b>100.0%</b>	<b>337</b>	<b>100.0%</b>



■ Fig. 3.521. AP x-ray of the shoulder in a 3-year old child with *unicameral bone cyst* in the proximal humeral metaphysis. This is the most typical site for this common tumor-like change in children. It does not generally require treatment at this site as it spontaneously disappears during the course of subsequent growth



■ Fig. 3.522. AP x-ray of the hand in an 11-year old boy with *enchondroma* of the 2nd and 3rd proximal phalanges. Since both tumors were causing the bone to protrude and were proving troublesome, curettage was indicated in this case, although enchondromas of the hand do not normally require treatment

A condition that specifically affects the hand is digital fibromatosis, which is observed in the first few months of life. Also typically encountered in small children is the likewise very rare condition of subdermal fibromatosis, which occurs in the shoulder, axilla and arm (see also ► chapter 4.5.4).

Although commonly seen in adults, ganglia are very rare in this age group. Giant cell tumors of the tendon sheaths occur even in adolescents in very rare cases, as do hemangiomas, which are among the commonest soft tissue tumors. Lipomas, fibromas and mucoid cysts are observed in the hand in very rare instances. All types of malignant tumors are equally rare in this age group and tend to affect adults. The commonest malignancy is a synovial sarcoma [3].

### Diagnosis

Since pain in the upper limb is extremely rare in children, it deserves our undivided attention. Only wrist pain is frequently unaccompanied by any visible pathology.

❗ **While commonly seen in adults, tendinopathies (such as »tennis elbow«) are rare in children and adolescents. Moreover, the »growing pains« that commonly affect the legs are never seen in the upper extremities. For these reasons, an x-ray should always be recorded if pain is present.**

However, most tumors or tumor-like lesions do not manifest themselves in the form of pain, but rather are observed as chance findings or following the occurrence of a pathological fracture. This particularly applies to unicameral bone cysts, which tend to affect the proximal humerus. Osteochondromas deserve special attention. On the scapula, osteochondromas are more likely to occur on the ventral aspect and can cause the scapula to protrude. During the differential diagnosis, this condition must be distinguished from a nerve lesion [2]. The diagnosis can be confirmed by an axial x-ray of the scapula or, in case of doubt, by a CT scan. On the proximal humerus, osteochondromas are usually located on the dorsomedial side, where they can be very large and disruptive. They may also cause nerve irritation.

On the *forearm*, *osteochondromas* usually cause *growth disturbances*, which are classified according to the system proposed by Masada [6] (■ Fig. 3.523, ■ Table 3.84). This classification can usefully serve as a basis for the decision concerning the appropriate treatment (see below).

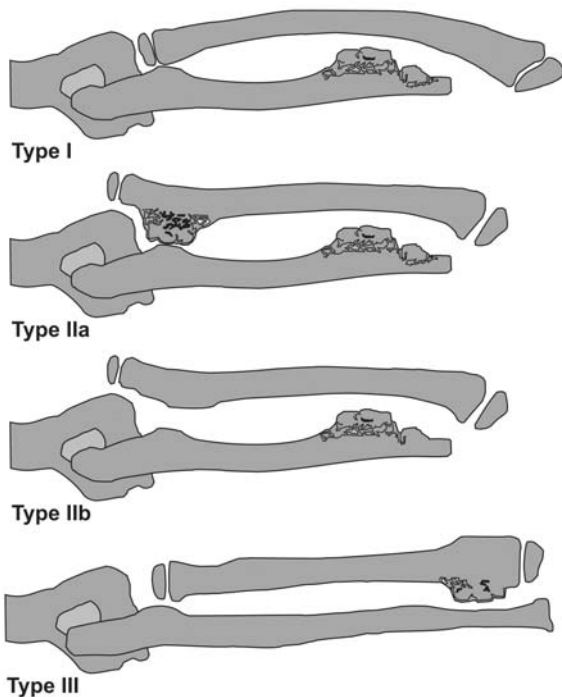
Other tumors affecting the forearm are enchondromas, osteoid osteomas, fibrous dysplasia and giant cell tumors.

On the *hand* enchondromas can cause troublesome swelling of the phalanges. But protuberances on the hand can also be caused by soft tissue tumors or tumor-like lesions. Tumors on the hand are usually diagnosed at an early stage since they prove troublesome even when small in size.



■ Table 3.84. Masada classification of growth disorders in osteochondromas of the forearm. (According to [6])

Type	Characteristic features
I	Distal ulna affected, shortening of the ulna
Ila	Distal ulna and proximal radius affected, shortening of the ulna and dislocation of the radial head
Ilb	As for Ila, but without visible exostosis on the radius
III	Distal radius affected and shortening of the radius



■ Fig. 3.523. Classification of growth disturbances due to osteochondromas on the forearm. (after Masada [6]; see text)

### Treatment

Since the therapeutic strategies for bone tumors are discussed in detail in ► chapter 4.5.5, only certain particular aspects will be highlighted at this point.

### Benign bone tumors

As a rule, surgery is not indicated for benign bone tumors and tumor-like lesions of the upper extremities if they show no aggressive growth locally and do not cause any pain. A more conservative approach is appropriate here than for the lower extremities. Since no weight-bearing is involved, the risk of deformation is much less than for the leg. This particularly applies to the unicameral bone cyst and fibrous dysplasia. In the upper extremity, neither lesion usually requires treatment. Any pathological fracture that occurs in

■ Table 3.85. Treatment of growth disorders in osteochondromas of the forearm based on the Masada classification. (According to [6])

Type	Treatment
I	Excision, ulnar lengthening and corrective osteotomy of the radius
II	Excision of the osteochondroma on the ulna, ulnar lengthening, poss. radial head resection
III	Only excision of the tumor on the radius



■ Fig. 3.524. Osteochondroma on the ulna (Masada type I [6]) with growth disturbance

a patient with a solitary bone cyst will generally heal completely and unproblematically with conservative treatment.

The situation is quite different, however, for locally aggressive tumors such as aneurysmal bone cysts. These should at least be marginally resected. Likewise, tumors that cause pain, such as the chondroblastoma, osteoblastoma or osteoid osteoma, should be treated. Osteoid osteomas affecting the upper extremity are often diagnosed only at a late stage [9]. The treatment is based on the usual guidelines.

*Osteochondromas* represent a special situation. Any that occur on the scapula, where they are usually located on the ventral aspect, should be removed, as otherwise the scapula will protrude and shoulder mobility may be hindered in the long term. The same applies to large osteochondromas of the proximal humerus, which can cause nerve irritation. On the forearm osteochondromas usually lead to growth disturbances (■ Fig. 3.524), as may be seen from the classification described above [6]. Based on this classification, the corresponding treatment shown in ■ Table 3.85 is recommended [6].



Decentering of the wrist also frequently occurs [4]. Other authors recommend radial epiphysiodesis, since a recurrence is possible even after ulnar lengthening [7]. Osteochondromas of the distal radius and ulna should be excised at a relatively early stage so as to minimize any impairment of growth. The ulna can be lengthened via an intramedullary Prévot nail, which prevents bowing of the ulna. Usually pronation and supination remain restricted in these patients.

Enchondromas on the hand can occasionally prove troublesome if they cause the bone to expand. Removal by curettage is indicated in such cases, and the defect can be filled with a cancellous bone graft. On the other hand, enchondromas that do not cause any problems should be left untreated.

### Malignant bone tumors

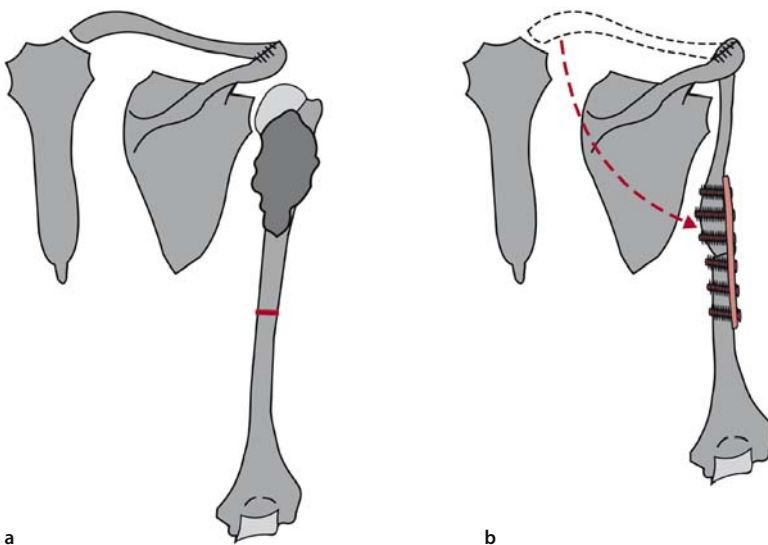
The treatment strategies for the relatively common osteosarcoma and the rare Ewing sarcoma follow the standard guidelines (▶ Chapter 4.5.6). Certain particular aspects concerning resection are worth mentioning: Osteosarcomas are usually located in the area of the proximal humeral metaphysis. Since the axillary nerve lies very close to the bone in its course from the posterior to the anterior side, a wide resection of the tumor is often not possible without also resecting this nerve. This will then lead to a failure of the deltoid muscle and thus of almost all active shoulder mobility. If very large, malignant, high-grade tumors are present in the shoulder area, the scapula may sometimes need to be removed completely together with the proximal humerus. In doubtful cases the surgeon can opt for the procedure of interscapulothoracic resection.

The reconstruction is also aggravated by the absence of the axillary nerve since it will not be possible to center

the humeral head prosthesis in the joint without the involvement of active muscle forces and the prosthesis will dislocate in a caudal direction. In young patients we use the clavicles as a replacement for the proximal humerus. The clavicles are released medially, folded down in the acromioclavicular joint and fixed to the residual fragment of distal humerus [8] (■ Fig. 3.525, 3.526).



■ Fig. 3.526. 16-year old female patient with *osteosarcoma* in the area of the proximal humerus. *Left* Preoperative view after 3 months of chemotherapy. *Right* After proximal humeral replacement by the *clavicula pro humero* technique



■ Fig. 3.525. Principle of proximal humeral replacement in the »*clavicula pro humero*« technique according to Winkelmann [8]. The clavicle is detached from the sternum and rotated downward in the acromioclavicular joint and fixed to the remaining section of the humerus

An alternative to this technique is an allogeneic bone graft (allograft). The muscles can be refixed to this graft, which provides a certain degree of purchase. This is not the case, however, for metal or plastic prostheses, which should be used only if the axillary nerve is preserved [10]. Poor results have been reported for extendable tumor prostheses [5]. Proximal dislocation of the prosthesis can occur during extension, although not all authors have observed this phenomenon [1].

Allografts can also be used on the forearm, although arthrodesis is usually the better and more lasting solution for the distal radius. A right-handed individual is able to manage functionally with arthrodesis, whether of the right or left wrist. However, arthrodesis can

prove problematic for a left-hander, as some left-handed individuals have to position their hand in extreme flexion when writing to avoid smudging the ink with the palm.

Fig. 3.527 shows the use of an allograft for a central, low-grade osteosarcoma of the distal radius in a left-handed female. A similar procedure can be employed for large giant cell tumors, which are not infrequently encountered at this site.

If malignant tumors occur on the hand, joint-preserving treatments are not usually possible (Fig. 3.528). Radiotherapy can help preserve the hand in some cases, although finger ray resections are usually more useful than likewise mutilating irradiation.

Fig. 3.527. Left AP and lateral x-rays of the left wrist in an 18-year old female patient with a central low-grade malignant osteosarcoma. Since the patient was left-handed, a primary arthrodesis after resection was out of the question since flexion in the wrist was absolutely essential for writing (this is less likely to be the case with a right-handed subject). A prosthetic replacement with a large bridging section was not a good solution for biomechanical reasons. We therefore chose a replacement with an allograft (right)

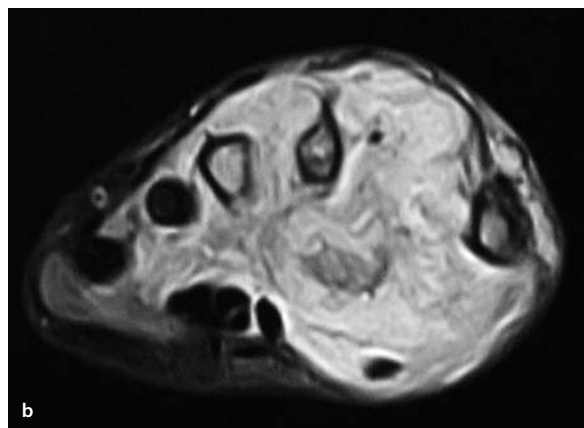
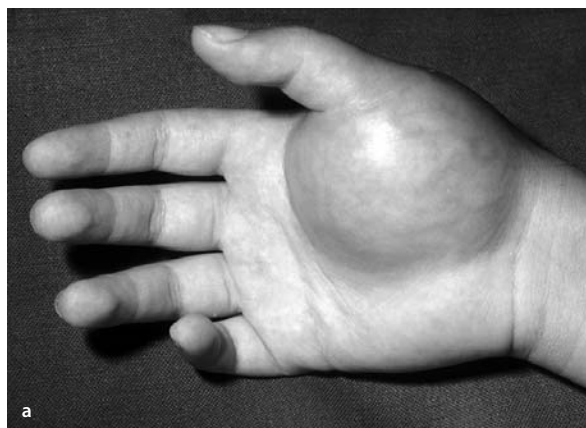


Fig. 3.528a, b. 8-year old girl with desmoplastic round cell tumor (a highly malignant tumor with a poor prognosis). a Clinical picture. b The soft tissues of the hand are infiltrated by the tumor. In such

cases a joint-preserving resection is out of the question. Since multiple metastases were present in this patient and the response to chemotherapy was poor, amputation was not performed

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# Systematic aspects of musculoskeletal disorders

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## 4.1 Traumatology – basic principles

C. Hasler

! **Modern traumatology is based on the needs of the children and parents and the prevailing social, economic, cultural and geographic circumstances.**

- A *child-based* approach means providing adequate analgesia, avoiding painful palpation and manipulation, outpatient treatment or the shortest possible hospital stays with a rapid return to play, school, sport and the family. All this requires an efficient therapeutic strategy based on the minimax principle: achieving the maximum result with minimum effort.
- The *parents* are the primary decision-makers, basing their decisions on the information given to them by the doctor in an open explanatory briefing.
- *Society*: Family structures have changed fundamentally over the last three decades. The traditional two-parent family with several children is now an outmoded archetype replaced by the single-parent and patchwork situations with changing educators or other innovative education systems. This requires a corresponding degree of farsightedness and individual clarification of the social situation by specialists in respect of hospital discharges and outpatient follow-up. The use of outpatient services means that care can be provided in the home, including physiotherapy, and that appliances can be rented, e.g. bath lifts, motorized splints, etc.
- *Economy*: The increasing restriction of available resources, bed capacities and care facilities, as well as itemized billing and modified remuneration systems significantly affect our room for maneuver and patient management.
- *Culture*: The spectrum ranging from the African medicine man to conventional western medicine offers considerable scope for one's self-image in medical terms. However, traumatology largely tends to avoid the discussion about alternative therapeutic approaches: The diagnosis is always too unambiguous, the objective too obvious. The route to the objective can vary, but is largely determined by the economic and social environment and by the individual preferences of the treating individual and by the standards of the local hospitals.
- *Geography*: The distance between the patient's home and the nearest basic healthcare provider or the nearest hospital unit influences the hospital stay of patients requiring close follow-up and possibly even the surgical procedure.

! **The greater the distances, the more extreme the climate, the less domestic care is provided and the more important the mobility of the patient at home, the more definitively the problem will need to be resolved by the time of hospital discharge!**

### 4.1.1 Epidemiology

! **Fractures are part of normal childhood development: Every other boy and every 4th girl suffers at least one bone fracture between birth and the age of 16.**

In any one year, one child in 50 will suffer a fracture. The incidence doubles in boys from under 10 years of age to puberty, whereas this puberty peak is not seen in girls. Fractures are most commonly caused by sporting accidents, followed by play-related and traffic accidents. Isolated fractures are usually involved. Multiple fractures or even polytrauma are much rarer than in adults, since children are less frequently involved in high-speed activities.

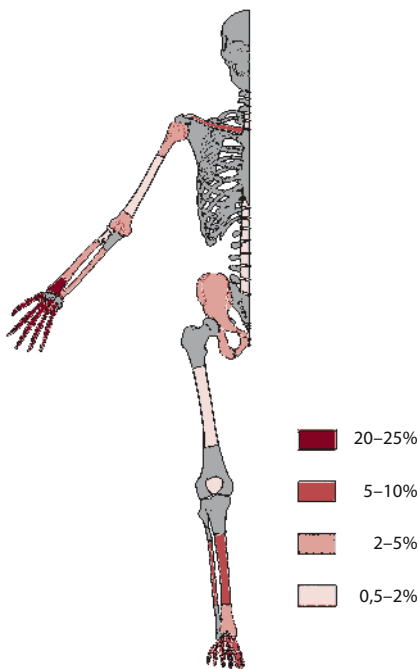
The relative frequency of the various fracture types varies according to sex and age group, geographical region (climate zones), the importance of sport and practiced sports, traffic volume and safety and is also subject to seasonal fluctuations. Last but not least, the reported figures depend on the size and importance of the recording institution: Large trauma centers are associated with a fundamentally different patient structure compared to e.g. small regional hospitals or even outpatient establishments. Accordingly, the following figures can be considered only as a rough guide, averaged across all age groups [24] (■ Fig. 4.1).

### 4.1.2 Communication with the parents and patients

! **Of all the problems associated with the musculoskeletal system in children and adolescents, fractures are by far the commonest reason for legal actions and expert reports.**

According to one American study, fracture treatments were involved in 77% of all lawsuits concerning children. In half of these cases the first contact was with the emergency room! The majority resulted from dissatisfaction with cast treatments. Diagnostic mistakes are particularly common with Monteggia lesions (missed radial head dislocation) and other elbow fractures [19]. But the medical problem is only superficially the trigger for a lawsuit. Much more important is the interaction between the doctor and patient/parents. Their psychological attitude is fundamentally different in an acute care environment compared to the elective situation of everyday pediatric

## 4.1.3 · Diagnosis



■ Fig. 4.1. Frequency of pediatric fractures according to site. (After [24])

orthopaedics, where most problems can be presented and discussed without any time pressure. In the latter situation, the family has the time to prepare the ground in an ideal fashion by conducting their own literature searches, making internet inquiries and obtaining second opinions.

In an accident the situation is completely different: The sudden violence of a trauma creates an emotional whirlpool of anxiety, powerlessness and helplessness that is even stronger if the injury was caused by third parties and the patient was completely blameless. The more acute the problem, the more restricted the room for maneuver in terms of time and therapeutic options, the more limited the free choice of doctor and hospital and the more likely that complications will result. The only true variable to emerge from this series of imposed circumstances is the doctor with his professional expertise and communication skills. He specifies the priorities and the speed of operations. Equally however, he can create an environment suggesting a relative freedom of choice to the parents through his calm demeanor, overall view of the situation, competence and openness. This will give the doctor room to pose questions, raise doubts and exert influence. This requires considerable skill bearing in mind that it is not only the parents and the patient who are emotionally stressed by the trauma. The duty doctor has probably been called away from some other task or is having to carry on through the night after a long day's work. Male colleagues appear to be less able than female doctors to cope effectively with this situation, since their risk of being at the receiving end of a complaint is three times that

for female doctors, who manage to produce more effective interaction with the parents and patients and thus create a healthier basis for trust [19].

Successful communication has many guises:

- Viewing the patients and parents as partners and taking them seriously.
- Providing an open, competent explanation as a basis for a common strategy.
- Answering parents' questions, including over the phone, and calling them back after their failed attempts to contact the doctor.
- Conveying an air of calmness and composure even under pressure of time.
- Recognizing one's own emotional stress.
- Documenting the content and time taken on discussions and formal written declarations of consent. These documents should not primarily be seen as a protective shield in the event of lawsuits, but rather as a basis for the therapeutic procedure established jointly with the parents.

In many cases this »arranged marriage« between the doctor and patient/parents resulting from the emergency situation is doomed to failure at an early stage because of irrational antipathy, professional shortcomings, lack of communication or other reasons. One possible way of defusing the situation is through postprimary management: Since few problems fulfill the criteria for a »genuine emergency«, the doctor is usually able to decide, depending on the local organizational and logistical circumstances in the individual case, whether the chances of success would be better under elective conditions outside the emergency situation.

### 4.1.3 Diagnosis

#### Clinical findings

Pain, additional swelling, hematomas and joint effusions, or even visible deformities are indicators of *fractures*, which still account for approx. 20% of pediatric surgical emergency admissions.

! Since the additional (pain-inducing) palpation of the painful site frightens the patients and does not provide any extra information it should be avoided.

The site of the pain can sometimes be difficult to locate in small children. However, with the keen perception of a detective, watching for spontaneous movements and possessing a knowledge of the commonest fractures in this age group, the doctor is usually able to decide on the correct x-ray projection even in these situations.

*Ligament injuries* of the knee and upper ankle do not require immediate clinical testing of stability. It is sufficient to arrange an x-ray on the day of the accident in order to rule out concomitant bone injuries and then immobilize the joint until the swelling and pain have subsided. After 5–7 days, the ligament stability can be investigated much more reliably in a patient with minimal pain.

## Imaging investigations

### Conventional x-ray

- If clinical examination shows a clearly visible deformity for which reduction under anesthesia is definitely indicated one projection plane will suffice.
- Side-comparing views are obsolete, since they involve additional costs without providing any additional information and suggest that the doctor is unaware of the normal age-matched radiographic anatomy.
- For shaft fractures the neighboring joints must also be x-rayed at the same time.
- Intra-articular distal femoral and proximal and distal tibial fractures are often not visible on standard AP and lateral views. Additional views in internal and external rotation are helpful.

### Ultrasound

The x-ray shows a fracture, and one that is usually non-displaced or minimally displaced, in fewer than one fifth of children with a suspected fracture. There is a need, therefore, for alternative, less stressful and more cost-effective imaging investigations. The advantages of ultrasound, i.e. functional examination on the awake patient, the lack of exposure to radiation, the inexpensive and mobile equipment, are countered by the disadvantages of investigator dependency and the difficult interpretation of the images. These drawbacks have limited its more widespread use.

Indications in which diagnosis by ultrasound is superior to radiographic investigation are lesions involving an epiphysis that has not yet ossified:

- epiphyseal separations of the proximal and distal humerus and of the distal femur in the neonate,
- in radial condylar fractures to differentiate between complete and incomplete intra-articular fractures [25],
- detection of intra-articular effusions,

Indications in which diagnosis by ultrasound is equivalent to, and can thus replace, radiographic investigation [6]:

- bone consolidation, which can be detected sooner with ultrasound,
- pseudarthroses,
- clavicular fractures,

- non-displaced supracondylar fractures,
- distal, metaphyseal radial fractures,
- compression fractures of the distal tibia.

Indications for radiographic investigation on its own:

- open fractures,
- unstable fractures,
- joint fractures.

### Bone scan

While this highly sensitive, though not very specific, investigation is not the first-line diagnostic technique, it is used if the following are suspected

- osteomyelitis,
- stress fractures,
- child abuse,
- tumors.

### CT scan

The *CT scan with 3D reconstruction* is suitable for visualizing complex fracture morphologies, particularly for operation planning or if the indication for surgery cannot be clearly established on the basis of the x-ray:

- unstable vertebral fractures,
- complex pelvic fractures,
- suspected vertebral fractures, facet dislocations, particularly in the upper cervical spine
- epiphyseal fractures,
- transitional fractures of the triplane type

### Magnetic resonance imaging (MRI)

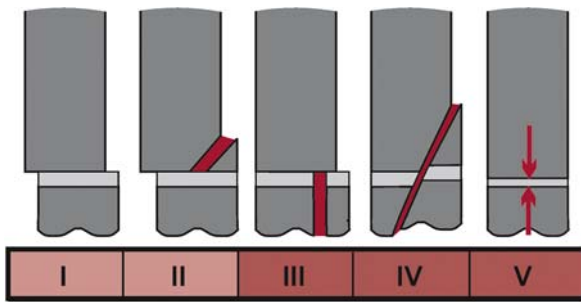
The disadvantages are the cost, the time involved and the fact that children of preschool age can only undergo an MRI scan under sedation or even anesthesia. The uncritical use of MRI for the detection of so-called occult fractures or internal joint lesions when the x-ray shows normal findings does not provide any therapeutic or prognostic benefit [3, 12]. On the other hand, with its radiation-free visualization of joint, epiphyseal and growth cartilage, ligament structures and nerve tissues, MRI is appropriate in the following indications:

- suspected osteochondral fracture with corresponding clinical signs and symptoms and normal x-ray,
- visualizing the site and degree of growth plate closures,
- spinal cord lesions in vertebral fractures,
- internal knee lesions if a corresponding clinical suspicion and potential therapeutic implications are involved,
- stress fractures,
- posttraumatic epiphyseal deformities, particularly in connection with avascular necroses [8].

### Fracture classifications

The fracture classifications used to date in pediatric traumatology are primarily simple morphological descrip-

## 4.1.3 · Diagnosis



■ Fig. 4.2. Classification of fractures in children according to Salter and Harris [21]. Type I and II lesions can also be described as »epiphyseal separations« or »shaft fractures«, and type III–V lesions as »epiphyseal fractures« or »joint lesions«. Type V (compression fracture) is initially undiagnosable

tions of injuries that affect the growth plates and are not particularly helpful as regards the choice of treatment or prognosis. The most commonly used classification is that according to Salter-Harris (■ Fig. 4.2), which subdivided trauma in the vicinity of the growth plate into four basic types, two describing pure separations and two describing fractures of the epiphysis [21].

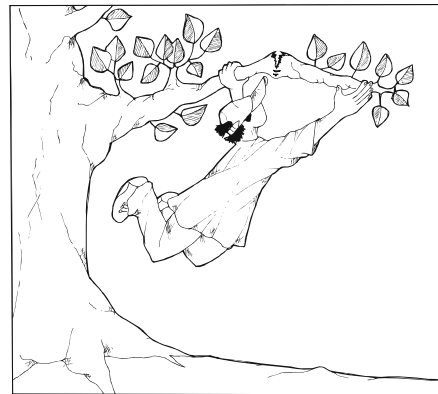
The original view that epiphysiolysees are not epiphyseal fractures but involve a high risk of physeal closure, is no longer justified. Epiphysiolysees are not just rather more common, they also lead, depending on the anatomical site and the displacement at the time of the trauma, to physeal bridges in a high percentage of cases. Since these form arbitrarily, they are difficult to influence by treatment. Some authors strongly dispute the possibility that a physeal bridge forms after axial trauma and an initially normal x-ray, i.e. a Salter type V trauma [18]. Nor does this additional type serve as a decision-making aid since it involves a retrospective evaluation. More recent, but less widespread classifications of pediatric fractures are more comprehensive since they also include fractures outside the growth plates [26].

#### Site

- **Diaphyseal:** Fractures of the shaft of a long bone, i.e. the section with a radiologically clearly visible cortex and medullary cavity and tubular in cross-section. The diaphysis is also the narrowest part of the bone.
- The **metaphysis** is located between the end of the diaphysis and that part of the growth plate on the shaft side. Epiphyseal separations (Salter I and II) are classified as metaphyseal fractures and run through the layer of hypertrophic chondrocytes (► Chapter 2.2.1).
- The **epiphysis** covers the section between the growth plate and the joint. Fractures in this part of the bone are termed epiphyseal fractures (Salter III and IV). Fractures not covered by the Salter classification include epiphyseal and ligament avulsion fractures, osteochondral fractures and transitional fractures.

#### Type of fracture

- **Compression fracture:** Axial trauma with bulging of both cortices.
- **Greenstick fracture:** Bowing of both cortices (bowing fracture) or bowing of the cortex on the concave side with complete fracture of the cortex on the convex side (classical greenstick fracture).
- **Complete fracture.**



The greenstick fracture is only broken on the convex side...

#### Displacement

- **Axis:** Establish the deviation from the normal position in degrees. For diaphyseal fractures: Measure the angle between the cortices of the main fragments and establish whether a varus/valgus deformity (AP plane) and extension/flexion deformity are present. In metaphyseal fractures, tangents drawn on the joint surfaces and knowledge of the physiological joint inclines are helpful. Alternatively, if the epiphysis is not very ossified, a straight line is drawn through the growth plate. Under normal circumstances this line runs parallel to the joint plane.
- **Length:** Measure any abnormal shortening in centimeters
- A **rotational deformity** can be recognized on the radiograph by means of the differing diameters of the fragments in the fracture area or the non-anatomical configuration of bony landmarks, although the extent of the deformity cannot be accurately described. Only on the lower leg can rotation be quantified to a precision of 10° in a direct comparison with the other leg by determining the angle between the malleolar axis and the transverse axis of the tibial head. At femoral level, any rotational defects in the acute situation can be determined only after surgical stabilization by comparing the passive internal and external hip rotation on both sides.
- **Side-to-side displacement:** Establish the displacement as a percentage of the bone width at the fracture level.



#### 4.1.4 Special injuries

##### 4.1.4.1 Birth traumas

The clavicle is the commonest site, followed by the humerus and femur [20]. Shoulder dystocia, a high birth weight and gestational age are risk factors [17, 20].

- **Clavicle:** Around 1–3 in 1000 neonates suffers a clavicular fracture in what would otherwise usually be an uncomplicated delivery. The expression »birth trauma« is not really appropriate in this case since a neonatal clavicle will break under a load of 5–16 kg, i.e. a level that is physiological in the birth canal. In addition to a pain-related reduction in spontaneous motor activity, a palpable, but not readily visible, spongy swelling is usually present over the fracture. An asymmetrical startle reflex, and the fact that the neonate can only be breast-fed on one side are further indicators. Since the findings are often minimal, however, many of these fractures remain undetected. Although it is rarely associated with a clavicular fracture, a plexus palsy should be ruled out [17].
- **Proximal and distal humerus, distal femur:** Epiphyseal separations due to birth trauma can, at best, only be suspected on an x-ray since the humeral head and distal end of the humerus and femur are completely cartilaginous at this stage. Any suspected fracture needs to be confirmed sonographically, although even the sonogram does not always clarify the situation. In such cases, only the subsequent clinical and radiological course will provide a conclusive result for the differential diagnosis. If a fracture has occurred, pain will be absent thanks to the rapid, and usually abundant, callus formation after just a few days. In view of the very high potential for spontaneous correction, even substantial deformities are acceptable. Partial closures of the medial physeal sections at the proximal end of the humerus are possible regardless of treatment.

#### Differential diagnosis: The infant fails to move an arm

- Clavicular fracture
- Epiphyseal separation of the proximal or distal humerus
- Humeral shaft fracture
- Plexus palsy
- Purulent arthritis, osteomyelitis
- Hemiplegia
- Child abuse

*Humeral shaft, femoral shaft and lower leg shaft fractures* are usually clinically obvious and simple to confirm radiographically. *Treatment* consists of 1- to 2-week immo-

bilization for pain relief, whether by splints, pelvic casts, plaster extensions or simple fixation of the arm to the thorax in fractures of the upper extremity.

Not infrequently the diagnosis is made only several days after the birth when an obvious reduction in spontaneous motor activity becomes apparent: In one of our little patients, a new-born girl, the left leg was affected. Any form of hip movement provoked pain. The neurologist confirmed a sciatic nerve palsy and the x-ray showed new bone formation in the area of the proximal medial femur. An MRI scan was arranged to exclude a tumor and revealed a *lesser trochanter avulsion*, compatible with sciatic neuropathia caused by excessive vertical traction on the leg during delivery.

##### 4.1.4.2 The abused child

Child abuse has many depressing facets, most commonly physical injuries caused by violence, followed by sexual abuse, psychological traumatization and neglect.

#### Occurrence

In over half of the cases, the parents themselves are the abusers. Particularly at risk are children in families with a low income/social status or single-parent families, firstborns, unplanned children, stepchildren, premature infants, handicapped children and children of drug-dependent parents.

**! In children under 1 year, 50% of the fractures are the result of abuse.**

Figures on the frequency are shocking, all the more when one considers that a substantial number of cases probably remain undetected: In the USA (290 million inhabitants), some 1.5 million children a year are affected, 2000 of whom die from the consequences of the abuse [22].

#### Diagnosis

##### History

A vague history, a trauma mechanism that is not compatible with the injury pattern, delayed consultation, repeated injuries, an »accidental« fall and placing the blame on siblings.

##### Clinical findings

Minor bruises on the front of the lower leg are an expression of the normal activity and playfulness of children. Abused children, on the other hand, particularly present with hematomas and burns on the trunk, back, groin area and thigh. Their shape provides an indication of the object used to inflict the injury. The child is examined by *ophthalmoscopy* (examination of the fundus of the eye) for retinal hemorrhages and detachments. The neurological status is investigated to rule out chronic subdural hemato-

#### 4.1.4 · Special injuries

mas. Abdominal examination. Urogenital status if sexual abuse is suspected. The specific investigations should be performed by corresponding specialists with accurate documentation.

##### Imaging investigations

- *X-rays*: in children under 5 years, supplementary views of the extremities, skull, spine and chest may reveal subclinical fractures, fractures of varying age, unusual sites, areas with pronounced callus formation and deformations. They also provide valuable information about the existence of any general underlying bone pathology.
- A *bone scan* is arranged if the radiographic findings are unclear but considerable suspicion remains. The scan will particularly reveal any rib fractures or occult fractures of the long bones.
- *CT scan* if a skull fracture, intracranial bleeding or intra-abdominal injuries are suspected.
- *Photographic documentation* of hematomas, burns, general and nutritional condition.
- *Laboratory tests*: clotting status, erythrocyte sedimentation rate, blood cell count.

##### Injury pattern

Skin injuries are the commonest, followed by bone fractures, usually shaft fractures. Half of the children have several fractures, in some cases of varying age. *Highly-specific* indicators of children abuse are metaphyseal lesions, particularly on the posteromedial aspect of the distal femur, and fractures of the ribs, scapula and sternum. *Moderately specific* indicators are multiple fractures, bilateral occurrence, differing healing stages, epiphyseal separations, vertebral body fractures, hand and foot fractures and complex skull fractures. Factors with *low specificity* include simple skull fractures, subperiosteal new bone formation, clavicular fractures and fractures of the long bones [9].

- ! **The possibility of child abuse must be considered when femoral shaft fractures occur in small children, particularly before the start of walking: »femur fracture – the ‘smoking gun’ of child abuse«.**

However, they can also occur in falling, running toddlers, when falling down the stairs or when adults fall over while carrying their child on their arm.

*Humeral shaft fractures*: In under 3-year olds, abuse is the cause of almost two-thirds of all acutely discovered fractures [23].

*Hand injuries* in neonates or toddlers who are not yet able to walk are likewise suspicious [16].

##### Differential diagnosis

»Is the fracture adequately explained by the history?« is a key question. A »no« indicates an imbalance between

the trauma energy and bone strength, and can therefore mean a non-accidental event or a bone pathology (► Chapter 4.1.4.2). The main condition to consider in the differential diagnosis is a previously undiscovered *osteogenesis imperfecta*, usually type I. In the *Münchhausen syndrome by proxy* parents invent symptoms, inflict injuries on the child or induce illnesses by poisons or drugs, thus provoking hospital admissions, costly investigations and unnecessary treatment [11].

##### What next?

- ! **The combination of risk factors, history, clinical findings and analysis of the injury pattern provides an overall picture that forms the basis for deciding whether a child abuse is classified as definite – probable – possible – unlikely.**

If abuse is suspected, a comprehensive multidisciplinary assessment and full written documentation are essential. The subsequent course of action, such as hospitalization to protect the child, ongoing diagnostic investigations, the involvement of guardianship authorities, child protection groups, etc. requires considerable tact and sensitivity. Since the personnel who initially treat the child often only have limited knowledge and little experience of the problem of abuse, it is worth consulting specialists at an early stage in order to establish the best course of action for the benefit of the child in need of protection and to act correctly according to the legal requirements. In doubtful cases, it is advisable to admit the child to hospital, as this ensures that the child is protected, permits rational and cautious planning and, not least, protects the doctor from the accusation of neglect.

#### 4.1.4.3 Pathological fractures

##### F. Hefti

Pathological fractures occur with minimal bone loading, either locally or as a generalized condition.

- *Local causes*: primary bone tumors, metastases, osteomyelitis, pronounced bowing of the bone (e.g. in crur varum congenitum, which can then develop into »congenital« tibial pseudarthrosis).
- *Generalized causes*: Osteogenesis imperfecta, Stuve-Wiedemann syndrome, rickets, juvenile osteoporosis, storage disorders such as Gaucher disease.

##### Local causes

Pathological fractures resulting from localized lesions occur during processes of rapid growth in which the bone does not have sufficient time to react. These can be benign lesions (usually cysts) or malignant processes. While most benign changes weaken the bone, the latter produces a sclerotic response in the immediate vicinity.

Since this prevents any overall weakening, pathological fractures are very unusual. This applies to giant cell tumors, enchondromas, non-ossifying bone fibromas, etc. One particular phenomenon concerns extensive processes which, although their growth may be slow, cover such a large area (particularly in the cortical bone) that the strengthening response is inadequate. Instead of fracturing, the affected bone then bows out (typically seen in fibrous dysplasia).

Pathological fractures in children and adolescents most commonly involve the proximal humerus or proximal femur as a result of solitary bone cysts. This lesion is usually simple to diagnose at both sites, as a clearly-defined osteolytic area of varying size is present in the fracture area and the bone is widened. It is essential to note the clear demarcation of the lesion, the septum formation, the widening of the bone and possible fallen fragments (▶ Chapter 4.5.2.7), since other benign (aneurysmal bone cyst, giant cell tumor, enchondroma) or malignant tumors (chondrosarcoma, osteosarcoma, Ewing sarcoma; ■ Fig. 4.3) can also lead to osteolysis at these sites. If the situation is not clear, further investigations (MRI, CT) and possibly a biopsy should be arranged before the fracture is treated. Outside the proximal humerus or femur, solitary bone cysts are extremely rare. Fractures in the vicinity of fairly major osteolysis should therefore always be investigated carefully before the fracture is stabilized with a contaminating measure.



■ Fig. 4.3a, b. Pathological fracture of the femoral shaft in a 16-year old boy with extensive Ewing sarcoma

### Generalized causes

Fractures associated with a generalized bone disorder occur with or without adequate trauma. In some patients the diagnosis of osteogenesis imperfecta (▶ Chapter 4.6.3.2) is made even at birth or during early childhood. In others, it is only the frequency of fractures that raises the suspicion of reduced bone strength. The most important differential diagnosis in patients with a high incidence of fractures is child abuse (▶ Chapter 4.1.4.2). A diagnosis of osteogenesis imperfecta (particularly of the clinically unclear late form) can be confirmed only by electron microscopic analysis of a biopsy. A characteristic feature is bowing of the bone as a result of microtraumas and the consequent appearance on the x-ray of adjacent bone resorption and formation processes. The treatment for frequent fractures is internal splinting with telescopic nails (▶ Chapter 4.6.2.24). In a case of juvenile osteoporosis it is the increased incidence of vertebral fractures without adequate trauma which particularly leads to the diagnosis.

#### 4.1.4.4 Stress fractures

*F. Hefti*

##### Pathophysiology

The natural remodeling process in the bone is based on microfractures after loading, which then trigger osteoclastic resorption. The resorption cavities are subsequently filled with lamellar bone. This is a natural process that enables the bone to be strengthened during corresponding loading. On the other hand, the sustained absence of loading will lead to a predominance of the resorption process and thus to osteoporosis. Any subsequent local overloading that occurs will lead to an imbalance between resorption and new bone formation and thus to a stress fracture. Periosteal and endosteal proliferation in the surrounding area attempt to compensate for the weakening of the bone, which leads to characteristic sclerosis of the bone surrounding the resorption zone.

##### Etiology, history

A triggering factor is repetitive trauma, usually caused by the excessive practicing of a particular sport. Load-related symptoms that worsen as the load increases and the absence of pain at night are characteristic of stress fractures. This point is extremely important for the differentiation from an osteoid osteoma, primarily chronic osteomyelitis or even a Ewing sarcoma, any of which can appear very similar in the imaging investigations.

##### Imaging investigations, differential diagnosis

Several weeks usually elapse between the onset of the first symptoms and the appearance of the stress fracture on a plain x-ray. The primary changes in the cortical bone are an ill-defined cortex and/or intracortical striation. At a

#### 4.1.4 · Special injuries

later stage, there will be localized osteolysis, usually in the form of an ill-defined fracture line (■ Fig. 4.4), but occasionally also as small cysts. In the cancellous bone, slightly blurred trabecular margins appear next to sclerotic and radiodense areas. The best diagnostic evidence is provided by a Tc-99m bone scan, which can show increased uptake even if the x-ray findings are negative. While the bone scan is highly sensitive, its specificity is less impressive. The increased uptake is usually not as pronounced as for an osteoid osteoma, but is similarly locally restricted.

In a primarily chronic osteomyelitis (which can also show sclerosis as well as fine osteolysis on the plain x-ray), the – usually only moderately pronounced – uptake generally covers a broader area. A (fine-slice) CT scan is ideally suited for visualizing the fracture line. An MRI is extremely sensitive and usually shows marked edema or an accumulation of fluid (often outside the bone as well), which readily raises the suspicion of a Ewing sarcoma. If the history is typical, the MRI scan should therefore be performed only after a failed attempt at conservative treatment, otherwise the risk of an unnecessary biopsy is very high.

#### Treatment

If the history is fairly typical and imaging investigations reveal the appropriate findings, then treatment with cast fixation should be initiated without further investigation (MRI) if this is permitted by the site of the fracture. Simply imposing a ban on sports will, on

the one hand, only be irregularly observed and, on the other, will not reliably rule out further overloading. The rapid disappearance of the pain and the regression of the radiographic findings confirm the accuracy of the diagnosis. If the symptoms have not subsided after four weeks, an MRI scan should be arranged. If the findings are doubtful, a biopsy is indicated (to investigate for Ewing sarcoma, primarily chronic osteomyelitis). If the findings are strongly positive, the cast fixation may need to be extended to eight weeks.

#### 4.1.4.5 Sports injuries

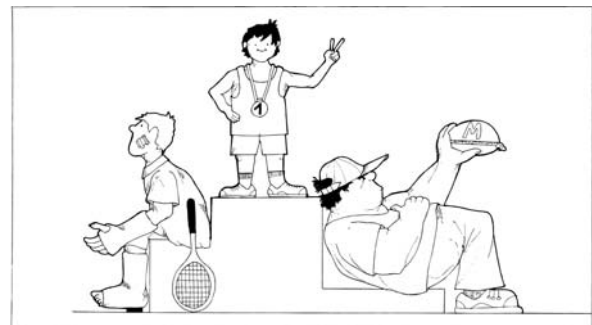
##### Epidemiology

Sports injuries have become increasingly common in recent decades in children and adolescents as well. According to recent surveys, and averaged across all age groups, they constitute the commonest cause of injury [24]. Male adolescents in particular appear to be exposed to a fairly high risk in sport. However, the approximately five-fold increase since 1950 in the relative proportion of sports injuries has been brought about by a reduction in traffic accidents and relatively fewer accidents during play [10].

**! The increasing significance of sports traumatology should not obscure the fact that it is *not* sport that poses the main health risk to children and adolescents, but rather the increasing *lack of exercise* and the associated obesity and declining physical fitness.**



■ Fig. 4.4. Stress fracture of the tibial shaft in a 14-year old boy (lateral x-ray). Note the fine, ill-defined fracture line and the slight thickening of the cortical bone anteriorly



The main health risk to children and adolescents is not sport, but rather the increasing lack of exercise...

#### Sport – a health policy issue

Despite the glamour of top-class sport, our hypokinetic society with over one-third of »exercise-neglecting« adults is a striking reflection of our difficulty in conveying to children the idea of sport for life. Only when sport can be successfully associated with positive emotions will it be possible to create the basis for lifelong sport

practiced for pleasure. The best way of achieving this in the long term is with daily, playful exercise lessons during the first years of school or, even better, at preschool age, when the motor learning skills are at their peak. More generally, sport and the promotion of exercise must also become an important political health issue in the context of primary and secondary prevention, particularly in a society that focuses on repair-based medicine. The health benefits far outweigh the risk of injury or the risk of suffering sudden cardiac death during sport. Moderate life-long exercise, even if practiced for just 2–3 hours a week or involving the additional expenditure of 1000 calories, leads to a significantly reduced risk of suffering cardiovascular illnesses, type II diabetes mellitus and certain tumors [13].

Also undisputed is the benefit of sporting activity in terms of psychological development: social learning, dealing with frustration and stress, self-esteem, goal-oriented thinking, physical and emotional competence are the key themes in this context.

#### Risk profile during growth

**! The biomechanical situation from birth until the conclusion of growth is characterized by complex changes in body size and proportions, leg axes, rotational configurations, body weight, muscle power and lengths and the lever relationships.**

The growth plates mean that the young are susceptible to specific injuries and overloading. This particularly affects those reaching puberty, at a time when they are exposed to an increased training intensity and show a greater willingness to take risks. Overload reactions between the tendons and growth zones, chronic separations of growth plates or fractures through growth zones are possible consequences.

Such reactions occur especially in sports

- with high eccentric muscle loads, particularly »stop-and-go« sports such as football, tennis and basketball,
- with high shear forces on the joint cartilage, particularly in the knee: football, skiing,
- with high axial forces on the spine: skiing, downhill mountain biking,
- with high rotational forces and forced lordosis of the lumbar spine: gymnastics, rhythmic gymnastics, ballet,
- with a risk of falling: skiing, snowboarding, riding, inline skating, motocross, climbing.

The basis for treatment and counseling is our understanding of age-specific loading capacity, trainability in terms of endurance and power and development-related stress (in)tolerance, all areas in which significant gaps in our knowledge still exist. Nor is any reliable data available on possible long-term damage. To ensure that the needs of

young athletes are properly met, the orthopaedist must, on the one hand, provide sound medical follow-up care in relation to their particular sport and, on the other, include parents, and possibly trainers and teachers, in the treatment and rehabilitation process. Not infrequently the promising young athlete must be protected from the excessive ambitions of parents who seek to find their own fulfillment in the impressive achievements of their offspring.

#### Sports-associated and overload injuries

- *Tendon-bone junction*
  - Sinding-Larsen-Johansson disease
  - Osgood-Schlatter disease
  - Calcaneal apophysitis
- *Growth cartilage*
  - Scheuermann disease
  - Distal radius: gymnast's wrist
  - Slipped capital femoral epiphysis
- *Apophyses*
  - Superior/inferior iliac spine, sciatic tuberosity
  - Apophyseal avulsion fracture/herniated disk in the lumbar spine
- *Joint cartilage*
  - Osteochondrosis dissecans, distal femur and talus
- *Stress fractures*
  - Spondylolysis L5
  - Diaphyseal stress fractures, particularly the tibia
- *Functional*
  - Femoropatellar pain syndrome
  - Functional back pain
- *Acute trauma*
  - Salter type I and II epiphyseal separations
  - Anterior cruciate ligament rupture, intraligamentous
  - Anterior cruciate ligament rupture, bony avulsion

#### 4.1.5 Therapeutic principles

##### Principles

- Include parents and patients in the decision-making process
- Immediate pain management by analgesia and immobilization
- No palpation or manipulation on the awake patient
- Try to provide a definitive treatment initially



- Prompt mobility and independence
- Incorporate spontaneous correction through further growth in the treatment regimen
- Correct axial deformities of less than 20° with cast wedging
- Respect fracture biology through closed reductions
- Use percutaneous fixation systems
- As few check x-rays as possible, as many as necessary

### Timing of treatment

The definition of an »emergency« means that the fracture must be managed as soon as possible, otherwise a high complication rate (circulatory disturbances, compartment syndrome, etc.) can be expected. This, in turn, means that the fasting period of at least six hours cannot always be observed. The dogma of emergency management of all fractures and dislocations that require reduction requires a discriminating appraisal [14]. Even completely displaced fractures can sometimes be managed in the postprimary period: absence of neurovascular signs and symptoms, no impending compartment syndrome, adequate pain control and close in-patient clinical monitoring are essential preconditions. Particularly with emergency cases at night, the doctor should carefully consider, on a case-by-case basis, whether the patient would benefit from delayed management by a rested, and possibly more professionally competent team.

- **Absolute emergencies:** Dislocations/displaced joint fractures/second- and third-degree open fractures/compartment syndrome.
- **Relative emergencies:** Tilted fractures for which reduction under anesthesia is definitely indicated can be managed immediately or within 24 hr depending on the psychosocial situation in each case.
- **Non-emergencies:** Age-matched acceptable deformities, or those that can be rectified with cast wedging.

### Conservative treatment

#### Cast immobilization

During the first few days, the purpose of cast immobilization is to rest the affected area and reduce swelling. The longuette technique with white plaster satisfies these requirements and is easy to apply, and thus convenient for the patient. After the swelling has subsided, approx. after 4 days from experience, the longuette cast is converted into an encircling cast, producing an ideal fit without any unpleasant and time-consuming change of plaster. In small children with stable fractures that do not require correction, e.g. compression fractures, an encircling cast is not fitted, thereby avoiding the need to use the cast saw for subsequent removal.

### Cast wedging

Cast wedging is a tried-and-tested means of reducing axial deformities of 10–20° indirectly without anesthesia. The parents and the patient should be comprehensively informed about the procedure and possible complications. Immediate reduction under anesthesia must be mentioned as an alternative, as should the possibility that the cast wedging may not lead to the desired result and that manual reduction may still be required. Experience has shown that most families opt for cast wedging which, subject to the requirements outlined below, represents a well-tolerated, low-complication and cost-saving corrective method for tilted fractures that are not completely displaced:

**Timing:** After 7–10 days the swelling of the limb has subsided and the immature callus stabilizes the fracture, resulting in freedom from pain in the cast, but still allows further bending, which is produced by the wedging.

**Technique:** On the concave side of the deformity, a semi-circular opening is made in the cast, but not the padding, with the cast saw. The cast spreader is used to gradually expand the cast until the patient notices slight pressure. Pain must be avoided at all costs. Excessive pressure involves the inherent risk of a pressure sore. This position is maintained with a small cube of wood that is wedged in the expanded gap. Under no circumstances should this spacer exert pressure on the underlying soft tissues. Window edema and slippage of the spacer are prevented with a plaster bandage.

Cast wedging is particularly *suitable* for:

- forearm and lower leg shaft fractures (complete and greenstick),
- metaphyseal fractures of the distal forearm and tibial metaphyses.

The effect of cast wedging is *limited* in:

- fractures of the proximal third of the forearm,
- epiphysiolyse of the distal radius,
- isolated radial or ulnar shaft fractures, since the intact partner bone acts a block.

Cast wedging is *unsuitable* for:

- humeral fractures,
- joint fractures,
- after the application of plastic casts as these are too inflexible.

### Cast removal

The noise of the cast saw and the heat and pressure induced by the saw blade provoke anxiety, sweating and a raised blood pressure and pulse. Proven stress-reducing measures include a calm explanation of the procedure, comfortable positioning, quieter cast saws, slow, safe operation, and the use of headphones [7].

### Extension treatment

We only use this in exceptional cases: rare cases of shortening deformities >2 cm in femoral fractures in small children.

### Surgical treatment

**!** *Percutaneous fixation methods are preferable as they allow closed reduction and thus respect the biology of bone healing in the best possible way. Moreover, implant removal is a trivial procedure.*

#### Closed manual reduction under anesthesia and cast immobilization

Cast wedging may be appropriate if secondary displacement remains.

#### Closed reduction under anesthesia, percutaneous wire fixation and cast immobilization

This method is recommended for metaphyseal fractures which prove to be unstable after reduction and which, in view of the patient's age, do not allow any remodeling of secondary deformities.

The diameter of the wire is selected on the basis of the patient's age and the fracture site and ranges from approx. 0.8–2 mm. The wires are bent at 90° above skin level and trimmed, leaving space for postoperative swelling. Wire-cast contact is avoided by means of circular cut-outs in the cast, approx. 4 cm in diameter, the walls of which extend slightly above the wire ends. These cut-outs help reduce the loosening and infection rates. When the bone has consolidated the wires, which have generally loosened in any case, are withdrawn without anesthesia.

#### Monolateral external fixator

*Method:* percutaneous insertion, away from the fracture, of 2–3 pins in each main fragment, onto which fixator clamps are mounted approx. 1–2 cm above the skin. Connect the clamps by a rigid rod. Reduce the fracture indirectly and tighten the system. Check and document the axial configurations clinically and radiologically and check the rotational situation clinically. Move the adjacent joints passively through their full range at operation.

##### *Indications:*

- Unstable shaft fractures of the femur and tibia (spiral fractures, long oblique fractures, multifragmented fractures, fractures with bone loss).
- Humeral shaft fractures that cannot be controlled by conservative treatment.
- 2–3 open fractures.
- Corrective osteotomies for posttraumatic deformities.

*Advantages:* Simple and quick assembly, closed reduction, immediate weight-bearing, dynamization, simple metal removal.

*Disadvantages and complications:* The external frame is not readily accepted by children of preschool age; transverse fractures are not an ideal indication – despite the dynamization, delayed healing or refractures can occur after removal of the fixator; impatient children want to rid themselves of their fixator as soon as possible with the risk of premature metal removal/refracture; pin care involves daily cleaning of the pin entry sites with cotton buds/hydrogen peroxide, daily showers or baths. Weekly checking of the pin entry sites; pintrack infections – these are usually only superficial and respond well to a 5–7-day treatment with a broad-spectrum antibiotic; keloid scarring after metal removal.

#### Intramedullary flexible nails (Prévot nailing)

*Method:* Expose the nail entry site with a short skin incision. Open the medullary cavity with the bone awl. Introduce one (radius and ulna) or two oppositely curving, flexible titanium nails, ascending from one side (radius, humerus), descending (ulna) or ascending from the medial and lateral sides (femur) or descending (tibia). *Ascending* = nails are inserted at the distal end and advanced in a proximal direction. *Descending* = nails are inserted at the proximal end and advanced in a distal direction. Biomechanical principles for optimal stability: sum of the nail diameters approx. 70–80% of the medullary cavity. Bend the nails so as to produce 3-point support. Double nails should have identical diameters. The growth plates should not be pierced.

##### *Indications:*

- Femur and tibia: Unstable transverse and short oblique fractures.
- Proximal pathological femoral fractures associated with a juvenile bone cyst.
- Unstable forearm fractures.
- Indirect reduction and stabilization of displaced radial head fractures.
- Unstable transverse humeral shaft fractures.
- Unstable proximal humeral fractures.

*Advantages:* Early functional follow-up treatment; simple, minimally invasive implantation; good cosmetic result.

*Disadvantages and complication:* In the case of obese patients or unstable fractures with a shortening tendency, reduced weight-bearing capacity with delayed mobilization may be observed because of the initial instability that is often present; irritating nail end, particularly in the distal femur or proximal ulna, with restricted movement at the knee; neuropathy at the entry site: almost always affects the superficial branch of the radial nerve at the distal radial entry site. Metal removal can be difficult if the nail ends have been trimmed too short.

#### Solid intramedullary nail

For femoral and tibial shaft fractures shortly before or after physal closure. There is a risk of femoral head ne-

#### 4.1.7 · Follow-up controls

crisis if the growth plates are still open. More recent and less widely used semi-rigid nails attempt to avoid this risk by using an entry point on the lateral aspect of the trochanter. Rigid tibial nails should not be used if the growth plates are still open as they have to be inserted directly through the proximal tibial plate.

#### Plate osteosynthesis

We have almost completely abandoned this technique because of the lack of immediate weight-bearing stability, the unsightly scarring and the second procedure needed to remove the plates from shaft fractures. We only use it these days for adolescents with displaced forearm fractures at the distal metaphyseal/diaphyseal junction (small-fragment plate) and for subtrochanteric femoral fractures (angled blade plates). A minimally invasive form of internal fixation based on adult traumatology techniques, has been used as an alternative method in isolated cases: closed reduction and fixation with a fracture-bridging plate advanced beneath the muscle and fixed with fixed-angled screws [1].

#### 4.1.6 Follow-up management

##### Physical therapy

**! The general prescribing of physiotherapy after fracture treatments is not particularly useful in growing patients. Children's enjoyment of exercise results in an autonomous, natural rehabilitation process, although this may take several months depending on the extent and nature of the injury.**

If progress is slow or even absent, the services of a physiotherapist accustomed to working with children should be engaged after bone deformities have been ruled out. Active, playful exercises supplemented by coordination training and instruction in a home-based program are preferred options. The latter saves time and costs and also encourages the patients to take responsibility for themselves.

##### Resumption of sports

Confirmation of the full resumption of sports activities depends on the bone consolidation and the restored range of motion. The functional deficits resulting from a post-traumatic loss of maximum power and stamina, muscle imbalances and partial losses of proprioceptive abilities, are not generally evident under everyday conditions. But under the increased demands of sport, they can involve the not inconsiderable risk of further injury. Accordingly, professional, graduated and individually adapted rehabilitation of motor skills in sporting youngsters is useful. This should be based on the nature of the injury, the age of the patient, the practiced sport, the desired level of training

and the ambitions, particularly in respect of participation in competitions.

#### Metal removal

In view of the risk of refracture, a period of 4–6 weeks after metal removal should elapse before the patient can resume jumping sports, contact sports or sports involving an increased risk of falling (skiing, snowboarding, inline-skating, riding).

#### 4.1.7 Follow-up controls

##### Consolidation

After undisplaced fractures of readily palpable bone, bone consolidation can be confirmed solely on the basis of painless palpation of the callus. The position can also be evaluated clinically in such cases. This particularly applies to:

- clavicular fractures,
- proximal humeral fractures,
- extra-articular olecranon fractures,
- distal radial fractures,
- tibial shaft fractures,
- toe and finger fractures,
- fractures of the distal fibular metaphysis.

For all other fractures, bone consolidation must be checked by radiography: The fracture is considered to have consolidated if the fracture gap in the area of three of the four cortices is bridged with callus in two standard projections at right angles to each other.

In practical terms, the verdict of a »consolidated or healed fracture« signifies the following for the patient:

- *Conservative treatment of the upper extremity:* Full movement-related stability. Increasing spontaneous axial loading in everyday activities.
- *Intramedullary nailing of the upper extremity:* The patient has already been moving his arm, without weight-bearing, during the consolidation phase and can now progress spontaneously to full axial weight-bearing.
- *Conservative treatments of the lower extremity:* Full movement-related stability. Increasing axial loading in everyday activities. Crutches for 1–2 weeks after cast removal.
- *Intramedullary nailing of the lower extremity:* As a rule, the patient has already been fully weight-bearing, usually with the aid of crutches for longer distances. Progress to walking without crutches.
- *External fixator on the lower extremity:* Schedule the metal removal.

As a rule, and depending on the fracture type, site and age of the patient, jumping sports, contact sports, ball sports

and sports involving an increased risk of falling should be avoided for some time after clinically or radiologically confirmed consolidation in order to avoid the risk of a refracture.

#### Growth disorders

Six-monthly check-ups should be arranged for up to two years after the trauma in order to detect any changes in length or axial alignment. This does not apply however to fractures without any risk of a growth disorder, e.g. compression fractures. If a growth disorder is already present, the control should continue until the end of growth.

#### Deformities

Possible causes are consolidation in an abnormal position, secondary deformities caused by growth disorders or a combination of these. The configuration of the affected extremity in terms of length, axes and rotation is described in comparison with the unaffected side. After vertebral fractures, the function and statics of the spinal column must be established clinically in three dimensions.

- *Frontal plane*: Shoulder and pelvic tilts, lateral deviations of the spinal column, abnormal vertical alignment, waist triangles.
- *Sagittal plane*: Profile (kyphoses and lordoses), abnormal vertical alignment, finger-floor distance, segmental mobility.
- *Transverse plane*: Rib or lumbar prominences, shoulder and pelvic rotation.

#### Mobility

Joint mobility is checked at the earliest 2–4 weeks after cast or metal removal and documented for both the affected and healthy sides by means of the zero-crossing method/neutral-zero method (► Chapter 2.1.1.3). The subsequent follow-up intervals depend on the site and extent of the deficit and the subjectively experienced functional restriction, which should be queried in detail, and documented, in relation to everyday and sporting activities.

#### Neurological disorders

Peripheral neuropathies are common particularly after elbow fractures. In most cases, these involve a prognostically benign neurapraxia that recovers completely within a few weeks or months. Clinical assessment and documentation of the sensory and motor nerve functions in the emergency room and subsequently during outpatient check-ups is essential since it allows any iatrogenic causes to be identified and the course of the condition to be monitored. Even age-related or pain-related unreliable findings should be documented as such. If no recovery is apparent by four weeks after the trauma, the patient should be referred to a neurologist for examination and the determination of nerve conduction rates. Occupational or physiotherapy is also useful.

#### 4.1.8 Prognosis

! **The fracture- and age-related interaction between the abnormal position at consolidation, spontaneous correction potential and any growth disorder will determine the bone shape on completion of growth.**

The *abnormal position at consolidation* can primarily be influenced therapeutically. This situation is known as *deformation* if spontaneous remodeling is expected. The term *deformity*, on the other hand, describes persistent axial and torsional defects that cannot be compensated for functionally and/or which the patient feels to be unsightly [5].

*Remodeling* cannot be influenced therapeutically (the administration of hormones and other drugs or foods do not have any detectable effect). On the other hand, an awareness of the limits of spontaneous correction of abnormal positions will enable the remodeling to be anticipated and integrated in the therapeutic strategy in a targeted manner. Deformities that result from inadequate remodeling should therefore be viewed as iatrogenic.

*Growth disorders* can be prevented or therapeutically influenced only to a limited extent.

#### Spontaneous corrections

A fundamental philosophical aspect of effective therapeutic strategies is the integration of the corrective growth potential as the fourth dimension of pediatric traumatology.

! **The exploitation of the spontaneous correction of deformities is linked to the following basic principles:**

- Sufficient residual growth: The remodeling potential is at its peak before the age of 10 and then declines until the onset of physiological physal closure.
- Only abnormalities in the main planes of movement of the nearest joint are corrected.
- The more active the adjacent growth plate, the greater the corrective potential.
- The parents and patients should be informed about the expected time taken to straighten the deformity, in order to avoid unnecessary second opinions and ill-considered corrective osteotomies.

The extent of the age- and deformity-related limits of correction are addressed in the corresponding sections on the various parts of the body.

But there are some deformities that can substantially correct themselves in a few days or weeks. Such occurrences are probably based on mechanical phenomena (ligamentotaxis or reduction in muscle tone as the pain

#### 4.1.8 · Prognose

subsides), e.g. in proximal humeral fractures, radial head fractures and epiphysiolyse of the distal radius.

#### Growth disorders

This term incorporates complete or partial inhibitions or stimulations of *epiphyseal* growth whose consequences are closely linked to residual growth and the nature and duration of the problem.

#### Completely stimulatory

Uniform stimulation of the whole growth plate with consequent excessive growth in length.

*Occurrence:* To a slight extent after every fracture, but particularly during the remodeling of deformities. The more pronounced the deformity, the longer the remodeling process and the greater the increase in length.

*Duration:* Limited to the end of the healing and remodeling processes, usually from a few months to approx. 1–2 years.

*Diagnosis:* Clinically.

*Consequences:* Clinically unimportant in the upper extremity. At femoral and tibial levels, increases up to approx. 2 cm and 1 cm, respectively, are possible. Significant changes in terms of statics can occur in the lower limbs, depending on pre-existing leg length discrepancies.

*Prevention:* Fractures of the lower extremity must be caused to heal correctly in terms of alignment and length in order to prevent significant spontaneous correction processes. Avoid secondary manipulations on the callus.

*Treatment:* Depending on the age of the patient, the extent of the difference and the change in spinal statics, conservative equalization of any leg length discrepancy, epiphysiodesis or lengthening osteotomy.

#### Partially stimulatory

Unilaterally increased physeal activity in connection with delayed healing on one side.

*Occurrence:* a) Bowing fractures of the proximal tibia, b) Radial condyle fractures.

*Duration:* Until bone consolidation, with a maximum of several months.

*Diagnosis:* Clinically and radiologically.

*Consequences:* Progressive axial deformity: a) clinical apparent valgus deformity, b) slight cubitus varus.

*Prevention:* a) Detection and elimination of the initial deformity, b) stable internal fixation.

*Treatment:* a) Corrective osteotomy.

#### Completely inhibitory

*Occurrence:* Rare. After comminuted fractures, secondary osteomyelitis.

*Duration:* Until completion of growth.

*Diagnosis:* Clinically, radiologically, possibly by MRI.

*Consequences:* Depending on the affected plate and residual growth. An annual loss in growth of approx. 1 cm can be expected for the distal femur. Thus, in an 8-year old girl, a total length loss of 6 cm can be expected by the time of expected physeal closure at the age of 14.

*Prevention:* None.

*Treatment:* Depending on the age of the patient, the extent of the difference and the change in spinal statics, conservative equalization of any leg length discrepancy, epiphysiodesis or lengthening osteotomy.

#### Partially inhibitory

*Occurrence:* In contrast with the conventional school of thought, physeal closures occur not only after Salter-Harris types III and IV *epiphyseal fractures*, but also after Salter-Harris types I and II *epiphyseal separations*.

*Duration:* Until physeal closure.

*Consequences:* Dependent on residual growth, site and extent of physeal bridges. Peripherally occurring ossification can quickly lead to deformities, whereas small central bridges often remain unnoticed or are spontaneously broken up as a result of continuing growth.

*Diagnosis:* Clinically during the six-monthly check-ups. If a disorder is suspected, radiological investigation with measuring of the joint angles is indicated. If a radiological deformity is present, the local residual growth must be estimated clinically (secondary sex characteristics, height) on the basis of sex, chronological age, bone age (x-ray with hand plates) and the affected growth plate. If a residual growth of more than 1–2 years is expected the possibility of a bridge resection or epiphysiodesis should be considered. The surgeon will need to know the precise location, size and composition (bony – cartilaginous – fibrous) of the bridge beforehand, ideally with the aid of magnetic resonance imaging with 3D reconstruction.

*Prevention:* The development of growth plate closures cannot be influenced to any great extent and occurs as a result of local plate destruction and the traumatic circulatory disturbance. In epiphyseal fractures the size of the bridge can be limited by watertight, stable internal fixation. If pronounced plate destruction has occurred, local fat interposition should be considered as part of the fracture management [2].

*Treatment:* Possible procedures, either alone or in combination, include corrective osteotomies, epiphysiodeses and bridge resections. The purpose of the latter operation is to restore normal growth activity. The basic preconditions are a residual growth of at least 1–2 years, a bridge of less than 40% of the total area and an experienced operator. Incomplete resections, bridge recurrences and overestimated residual growth as a result of premature physeal closure or restricted functioning of plate sections that appear normal on imaging investigations are possible reasons for the unsatisfactory results that are not infrequently observed [4].



## Posttraumatic deformities

### Indications for correction

- *Urgent* because worse preconditions are expected if a wait-and-see approach is followed.
  - Pseudarthrosis of the radial condyle of the humerus with/without cubitus varus and ulnar neuropathy.
  - Radial head dislocation.
  - Intra-articular fractures that have consolidated in an incorrect position.
  - External rotation of the distal fragment after femoral fractures with consequent femoral neck retroversion (= pre-arthritis).
  - Femoral neck pseudarthroses.
  - Deformity of the forearm shaft with restricted turn-over movement.
- *Not urgent. The patient determines the indication and the timing depending on the functional and/or cosmetic impairment.*

These indications deformities which, whether intrinsically or because of the extent of the deformity and age of the patient, are unlikely to undergo spontaneous correction or which are felt to be unacceptable.

- Cubitus varus after supracondylar humeral fractures.
- Extension deformity after supracondylar humeral fractures after the age of 6.
- Internal rotation deformity of the distal fragment after femoral fractures.
- Rotational deformity after lower leg fractures.
- Rotational and axial deformities after finger and metacarpal fractures.
- Axial deformities of the upper arm.
- Axial deformities of the femur.
- Valgus deformities of the proximal tibia. If the condition is left untreated for a prolonged period in children under 6 years there is a risk that the deformity will grow in the middle of the shaft, whereas the proximal and distal epiphyses return to the horizontal, which would make a double osteotomy necessary. Alternatively, the deformity is corrected at an earlier stage, albeit with an increased risk of recurrence.
- Axial deformities of the tibial shaft and distal metaphysis.

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## 4.2 Axes and lengths

F. Hefti, C. Hasler

### 4.2.1 Are children twisted when they have an intoeing gait or warped if they are knock-kneed or bow-legged?

*The intoeing gait of a child demonstrates that we are of animal rather than divine origin. As with all mammals, the hips of the human fetus are in a flexed position in the womb. The centering of the femoral head during increased*



*anteversion is better in flexion than when the femoral neck is only slightly rotated forwards. In animals, the hip remains in the flexed position throughout life. The human child, however, starts walking with an »unnatural« extended hip position and must compensate for the increased anteversion by adopting an intoeing gait.*

» **Even with crooked legs it is possible to walk straight.** «

(Jakob Lorenz)

The intoeing gait of toddlers often gives the parents much cause for concern. Sometimes the mother and father do not spontaneously decide to consult the doctor, but only do so after being alerted by a well-meaning grandmother or neighbors, or even a shoe sales assistant. They bring their child to the office and ask anxiously whether their child with the twisted feet is really normal. Occasionally, the child may also be bow-legged or, if slightly older, have pronounced knock knees, which just serves to deepen the worry lines on the parents' faces even more. For this reasons, a detailed review of the rotational and axial relationships in children is appropriate.

#### Terminology

Since terms are often mixed up and used incorrectly, **■** Table 4.1 lists relevant terms with corresponding explanations.

#### Normal development of axial and rotational relationships in the lower extremities

A knowledge of the normal development of axial and rotational relationships is crucial in order to be able to

**■ Table 4.1.** Terms associated with axial and torsional deformities

Term	Meaning
Torsion	Rotation of the anatomical axes of the two end points of a bone in the frontal plane in relation to each other
Rotation	Movement of a joint around a fixed axis of rotation
Valgus	Axial deviation towards the central axis of the body in the frontal plane
Varus	Axial deviation away from the central axis of the body in the frontal plane
Femoral anteversion	Angle between the femoral neck axis and the frontal plane towards the front
Retroversion	Pathological torsion posteriorly of the femoral neck in relation to the frontal plane
Genu valgum	Valgus deviation of the lower leg in relation to the upper leg (knock knees)
Recurvated knee	Hyperextensibility of the knee by >10°
Genu varum	Varus deviation of the lower leg in relation to the upper leg axis (bow legs)
Bowed leg	Medial bowing of the distal part of the lower leg
Medial torsion of the tibia	Torsion of the lower leg with a malleolar axis of less than 10° at the age of over 5 years
Lateral torsion of the tibia	Lateral torsion of the lower leg with a malleolar axis of more than 40° in relation to the knee condyle axis
Derotation osteotomy	Usual term for a correction of the torsion of the upper or lower leg; a more correct term would be derotation osteotomy

establish whether a child's situation is pathological or can still be considered within normal limits. Axes and torsions undergo typical changes as the infant develops into a toddler, child and adult. These changes mutually effect each other in the thigh, lower leg and foot. Thus the characteristic flat valgus foot position of the toddler depends greatly on the extent of anteversion of the femoral neck. Expressed in rather simplified terms, the flat valgus foot represents an attempt by the child to correct the inward-facing position of the foot resulting from the increased anteversion of the femoral neck. During fetal development the whole lower extremity rotates internally from a pronounced position of external rotation.

**Anteversion**

The average anteversion at birth is approx. 30°. This decreases during growth to a final angle of 15° in adulthood (Fig. 3.142, Chapter 3.2.3). Slightly higher angles are found in girls compared to boys. The rotatability of the hip is an indirect expression of the degree of anteversion. Increased anteversion also increases the ability to rotate the hip internally. But the progression of rotatability does not completely parallel the development of anteversion. At birth, external rotation is usually higher than internal rotation, whereas the opposite is the case after the child starts walking.

**Femoral Neck-shaft angle in the frontal plane**

The femoral neck-shaft angle is approx. 150° at birth, decreasing to 120° by adulthood (Fig. 3.140, Chapter 3.2.3).

**Tibial torsion**

Tibial torsion refers to the rotation of the malleolar axis in relation to the back of the tibial condyle at knee level. Torsion is not usually present at birth. A lateral torsion of 15°, on average, develops during the first few years of life. Tibial torsion can also be expressed by the angle between the axes of the foot and thigh (Chapter 3.4.1). The foot should show a lateral torsion of 10–20° (Fig. 4.5).

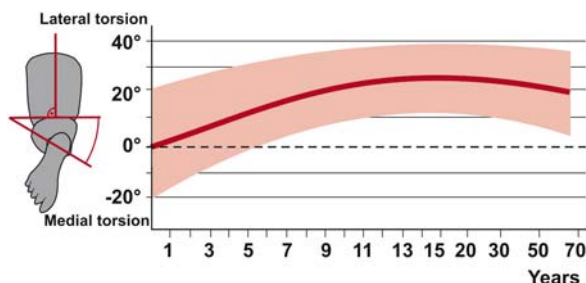


Fig. 4.5. Malleolar axis compared to the femoral axis (as an expression of the tibial torsion) during growth. (After [14])

**Knee axis**

A physiological varus leg axis exists at birth, in the sense of a bowed leg rather than a genu varum. The average varus angle is 15°. The knee should be in a neutral position at the start of walking, but then subsequently develops a valgus position of approx. 10°, producing an intermalleolar distance of 2–4 cm. The exaggerated valgus position corrects itself by the age of 10 to the physiological valgus position of 5–7°, which we experience as a »straight« leg axis, with both the femoral condyles and malleoli touching each other (Fig. 4.6).

**Clinical features, diagnosis**

We first observe the gait pattern. Small children often show an intoeing gait. A more specific distinction is possible between »kneeing-in« and »toeing-in«. We then observe the axes in standing. In a case of genu varum we measure the intercondylar distance and, in genu valgum, the malleolar distance in centimeters. The position of the patella should also be assessed in the neutral foot position (approx. 10° external rotation in relation to the median) (Fig. 4.7).

Next, we measure the range of motion of the hip. External and internal rotation are determined on the prone patient with the hip extended (Chapter 3.2.1). Anteversion can also be estimated by clinical examination in this position. With the patient still in the prone position we measure the torsion of the malleolar and foot axes compared to the femoral axis. Note that the foot should not

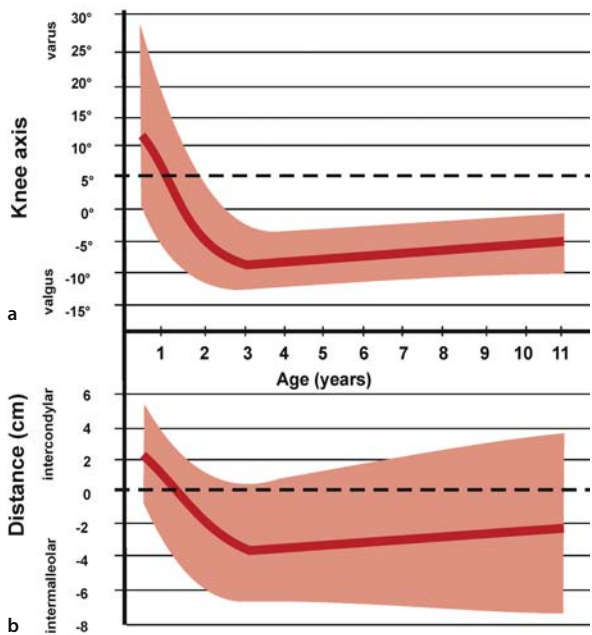


Fig. 4.6a, b. Knee axis (a) and intermalleolar / intercondylar distance (b) during growth. (After [14])

## 4.2.1 · Are children twisted when they have an intoeing gait or warped if they are knock-kneed



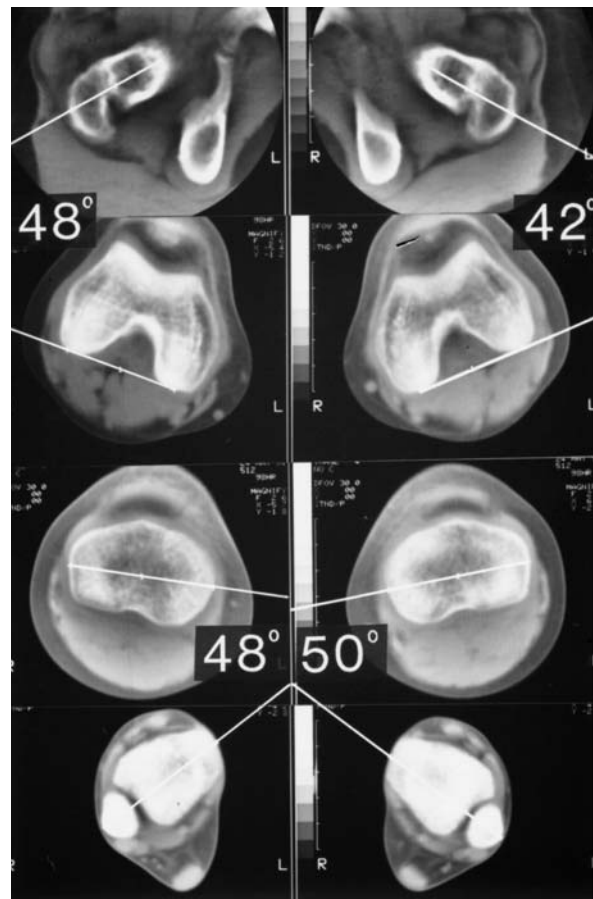
■ Fig. 4.7. Legs of an 8-year old girl with increased lateral torsion of the tibia. *Left* When the feet point straight ahead the kneecaps are turned inward. *Right* When the feet are rotated outwards the kneecaps point straight ahead

be allowed to rotate, since it can easily rotate inwardly or outwardly at the ankle. It should be at right angles to the lower leg and should adopt its spontaneous position in respect of rotation.

### Imaging procedures

*Anteversión* (AV) can be determined by various methods. X-rays can be recorded in two planes (AP and Dunn view, ► chapter 3.2.2) and the project angles measured. The true figures for the CCD and AV angles (■ Table 4.2; also ► Chapter 3.2.2) must then be determined from the conversion table shown opposite. Anteversión can also be determined with almost equal precision by means of ultrasound [2]. However, if an abnormal condition requiring treatment is not suspected, clinical measurement will also suffice (► Chapter 3.2.1) [15]. Quite accurate measurement is also possible with CT. To this end, slices must be recorded through both femoral necks and both femoral condyles at knee level (■ Fig. 4.8).

It should be noted, however, that the anteversión angle on the CT scan is not measured in space but rather in the horizontal plane, which is not exactly the same. Both CT and sonography can also be used, in addition to clinical examination, for measuring *tibial torsion* [14]. The main problem with this measurement is that the back of the tibial condyle at knee level is rounded, thereby preventing any clear axis to be determined. The range of normal values is relatively large. But since therapeutic measures only need to be considered if the values are very abnormal, we believe that the inaccuracy of this measurement is not particularly problematic. The *knee axes* can be measured radiographically on AP x-rays, with smaller children standing on both legs and older children standing on one leg.



■ Fig. 4.8. Rotation measurement with a CT scan (same patient as in Fig. 4.7): On both sides one slice in each case has been recorded through the femoral necks, the femoral condyles, the tibial condyles and the malleoli to enable the axis to be determined. Increased femoral anteversión and increased lateral torsion of the lower leg are present at the same time

■ **Table 4.2.** Conversion Table for determining the actual AV and CCD angles. Top figure = true AV angle, bottom figure = true CCD angle. (From [11])

Projected CCD angle	Projected AV angle							
	9°	10°	30°	40°	50°	60°	70°	80°
100°	9 100	20 100	30 99	40 98	50 96	60 94	70 93	80 91
110°	10 110	21 108	32 106	42 105	52 103	61 99	71 97	80 93
120°	11 119	22 117	33 115	44 112	53 108	63 104	72 101	81 95
130°	12 129	24 126	35 124	46 120	55 116	64 109	73 104	82 96
140°	13 138	27 135	38 132	49 127	58 120	67 114	75 107	83 97
150°	15 147	29 144	42 141	52 136	61 129	69 120	76 112	84 100
160°	18 158	34 155	46 151	57 144	65 134	73 128	79 116	85 103
170°	27 167	46 164	58 159	67 154	73 145	78 134	83 122	87 113

### Pathology

Some children show even greater anteversion than normal at birth. We consider the anteversion to be pathological from an angle of 50° and above. Several authors have shown that the increased anteversion usually returns to normal during the course of growth [17]. However, derotation fails to occur in isolated cases, i.e. the anteversion angle still remains 50° and above at completion of growth. This type of persisting *anteverted hip* has two principal causes:

- the presence of a (minimal) cerebral palsy,
- compensation of the increased anteversion at the femoral neck by increased lateral torsion of the tibia.

The *physiological correction* of the increased anteversion is attributable to the (unconscious) need to place the feet on the ground in parallel (or in slight external rotation), as this is the only way of achieving efficient forward motion. If motor coordination is impaired, this impulse is lacking. The derotation of the femoral neck can be described as a »physiological slip of the capital femoral epiphysis«, since the direction of movement of the femoral head in relation to the shaft corresponds to that in epiphyseal separation, which shows that the dynamic forces during upright walking produces this alignment of the femoral neck. A recent study has shown a correlation between increased anteversion, reduced hip extension and motor development [7].

Anteversion also decreases in patients with cerebral palsy, but not to the same extent as in healthy individuals. The derotation is much better in children who are able to walk than in severely disabled children [9]. Gait investigations have also shown that the load transfer differs greatly during an intoeing gait compared to a normal gait [10]. If the increased anteversion were offset by increased tibial torsion, then the impulse for further correction of anteversion would no longer be present since the feet strike the ground in parallel. Various investigations have shown a positive correlation between femoral and tibial torsion [14].

The problem, however, lies in the fact that the knee is rotated in an intoeing gait and is not aligned with the direction of walking. This is not just an esthetic problem. Although there are no scientific investigations on the consequences of this kind of deformity, it is striking that adult patients with this condition tend to consult the orthopaedist with knee problems relatively frequently. On the other hand, the increased anteversion does not have any long-term consequences for the hip [18]. No increased incidence of osteoarthritis of the hip has been observed, for example. And this is logical, since the internal rotation of the femur during walking produces a physiological position at the femoral neck. It is only the position of the knee that is pathological. In contrast with the increased anteversion of the femoral neck, retroversion is a sign of pre-arthritis [8, 18] and requires



#### 4.2.1 · Are children twisted when they have an intoeing gait or warped if they are knock-kneed

treatment, since such patients frequently experience pain even in young adulthood because of impingement [8, 18] (► Chapter 3.2.3).

As already mentioned, the impulse to correct the anteversion is lacking in *increased lateral torsion of the tibia*. We consider that a lateral tibial torsion of 40° or above requires correction [6, 16]. The best age for this is between 8 and 10. Up until the age of 8 we await the outcome of spontaneous developments, although the lateral torsion tends to increase rather than decrease. The supramalleolar tibial derotation osteotomy can be carried out at this age. This is a minor and safe procedure associated with minimal morbidity and gives the child the chance to derotate the femoral anteversion during the pubertal growth spurt. This operation should not be performed after the age of 10. Efficient derotation is possible by this point only if the fibula is osteotomied as well. Fixation is more complicated and spontaneous derotation of the femur can no longer be expected. An intoeing gait would then persist in such cases unless the torsion of the femoral neck were also corrected, which – when performed bilaterally – is quite an elaborate procedure.

A *pathological medial torsion of the tibia* is rarer than lateral torsion. This occurs regularly in connection with a genu varum, but is very atypical in clubfoot. The internal rotation of the foot is not caused by the medial torsion of the tibia, but rather by the foot position itself. Consequently, the externally rotating tibial derotation osteotomy is rarely indicated in clubfoot. On the contrary: the malleolus usually stands well back. In fact,

an increased lateral torsion is generally present despite the inward-facing position of the foot. Only in very rare cases is correction indicated at this site, and the correction must usually be performed on the foot itself (► Chapter 3.4.3).

**! We therefore consider that the supramalleolar tibial derotation osteotomy is indicated if there is a lateral torsion of more than 40° or a reduced tibial torsion of 5° and under.**

*Genua vara* are always pathological. They occasionally occur after the start of walking, particularly in children who start walking at a very early age, i.e. at a time when the knees are still in varus alignment. In such cases this varus axis can take on dramatic proportions at the age of 1–2 years. It is usually associated with pronounced medial torsion of the tibia, making the genua vara appear even more extreme. Nevertheless, we would warn against overtreatment at this stage. The prognosis for these idiopathic cases of genu varum is very good in small children provided there is no underlying pathology. A recent investigation has shown that the risk of progression only applies if the varus is located predominantly in the tibia rather than the femur [1]. Pathological forms occur in Blount disease, rickets and after trauma.

*Blount disease* is extremely rare among the white population (■ Fig. 4.9). This condition involves a necrosis in the area of the proximal medial tibial epiphysis, and possibly the medial distal femoral epiphysis. Blount disease is much commoner among the black population



■ Fig. 4.9. AP and lateral x-rays of the left knee in a 3-year old boy with osteonecrosis of the medial femoral condyle (*Blount's disease*)



■ Fig. 4.10. 5-year old girl with *vitamin-D resistant rickets* and pronounced *genu vara* bilateral: **a** from the front, **b** from the back

[5]. In addition to the infantile form, there is a juvenile variant, which can involve the spontaneous formation of a medial bridge across the epiphyseal plate and necrosis of the proximal medial tibial epiphysis. This variant is also known as the late form of Blount disease. One sign of *rickets* (■ Fig. 4.10) is a widening of the epiphyseal plate (■ Fig. 4.11). *Rickets* can be related to the diet or occur as a vitamin D-resistant condition (▶ Chapter 4.6.2.1). *After trauma*, *genu varum* occurs on medial physeal closure. An increased varus axis is a clear sign of pre-arthritis. A varus position with an intercondylar distance of more than 2 cm should be corrected, particularly if a rotational deformity is also present in the lower leg.

*Genu valgum* is physiological in small children (■ Fig. 4.12). Up until the age of 8–10 years a gap between the malleoli is apparent in most children when the knees are approximated. The persistence of *genu valgum* beyond the age of 10 is rare and almost always caused by relatively pronounced *overweight*. Treatment should consist primarily of weight reduction. *Genu valgum* is much less commonly associated with pre-arthritis compared to *genu varum*, and the need for treatment is likewise reduced and indicated only in severe forms.

*Recurvation* of up to 10° in the knee is an expression of general ligament laxity and commonly occurs in children. If the knee can be hyperextended by more than 15° (■ Fig. 4.13), then the condition is pathological. The cause can usually be found not just in the capsular ligament apparatus, as the physiological inclination of the tibial plateau is also missing, whether as a result of idiopathic, posttraumatic or iatrogenic factors (after surgically-induced damage to the apophysis on the tibial tuberosity).



■ Fig. 4.11. Same girl as in Fig. 4.10 at the age of 11 years. Correction of the pronounced *genu vara* required *osteotomies on the upper and lower legs* in order to keep the knee horizontal

## 4.2.1 · Are children twisted when they have an intoeing gait or warped if they are knock-kneed



■ Fig. 4.12. 6-year old boy with physiological *genua valga*



■ Fig. 4.13. 16-year old girl with pronounced *recurvated knees* (by 25°) with general ligament laxity and (idiopathic) missing inclination of the tibial plateau

The alignment of the lower extremity in the frontal and sagittal planes and the joint position have major consequences for the function and loading of the hip, knee and ankle. There is a normal range for the position of these joints in respect of the mechanical and anatomical axes of the femur and/or tibia. In the frontal plane we use both the anatomical and mechanical axis lines in therapeutic planning. Since the mechanical axis is less relevant in the sagittal plane, only the anatomical axis is used for planning. Angulation deformities are characterized by four parameters:

- level of the apex of the angulation,
- plane of the angulation,
- direction of the apex in the plane of angulation,
- extent of the angulation [12].

In order to correct the angulation deformity, all of these parameters must be determined before the level and type of osteotomy to be performed is selected. The apex of the angulation is measured as the intersection between the proximal and distal axis lines. The extent of the angulation is determined at the level of the apex as a transverse angle. This is usually less than 90°. A line bisecting this angle is drawn through the apex, thus dividing the longitudinal angle (which is normally greater than 90°) into two equal halves.

### Treatment

#### Conservative treatment

Although numerous measures have been proposed for correcting axial and rotational deformities, none has proved completely effective to date. The list of measures starts with the instruction that the child should not be allowed to adopt a »reverse cross-legged« sitting position. However, we consider this ban to be absurd. In a child with increased anteversion, the hip is well centered when the legs are internally rotated. If the legs are placed in a position of external rotation, the femoral head subluxates anteriorly. For the purposes of derotation, the dynamic forces during walking are far more effective than the static forces during sitting. »Twister cables« have been used for many years to correct the anteversion. These extend laterally on the leg from a hip strap to a lower leg orthosis and force the foot to twist outwards. However, the inefficiency of this rather unpleasant measure for children has since been confirmed. Nor has the treatment with diagonal inserts proved effective in influencing the anteversion. Attempts to treat genua vara or genua valga with splints are also doomed to failure. Such splints are usually worn only at night when no dynamic forces are involved. Since the knee ligaments are elastic, the correction takes place in the joint instead of the bone. The only recommendation we give to parents of children with axial deformities is to insert a wedge in the shoe. For genua valga and vara we recommend 3 mm medial and lateral wedges respectively,

with the aim of modifying the loading axis. We do not know whether this measure is actually effective or not. But since it is harmless and does not bother the child we can nevertheless recommend it.

### Surgical treatment

#### Correction of femoral neck anteversion

If an anteversion of more than  $50^\circ$  is present at the age of 12 years, the possibility of surgical correction can be considered, particularly if the ability to rotate the hip externally in the extended position is restricted to  $20^\circ$  or less. In unilateral cases we correct this deformity by means of an intertrochanteric osteotomy and fix the result with an angulated blade plate (■ Fig. 3.169). After the operation immediate mobilization with crutches is possible. If the osteotomy is performed on both sides at the same time at the intertrochanteric level, a 6-week period of bedrest would have to be expected, even with the use of modern implants. An alternative is to perform the osteotomy at the supracondylar level above the knee and insert low-contact plates with fixed-angle screws (■ Fig. 4.16). Immediate mobilization with weight-bearing is possible after this procedure. This is not only due to the type of implant, but also to the fact, that at this level (unlike the intertrochanteric level) the bending momentum is much smaller. We employ this procedure if surgery is required on both sides since it avoids a prolonged period of bedrest. This procedure can also be employed at the subtrochanteric level.

#### Correction of tibial torsion

Up to the *age of approx. 10 years* the torsion can be corrected at supramalleolar level on the tibia alone. The osteotomy is performed above the epiphyseal plate through an approx. 2 cm long vertical incision. The tibia can be derotated externally or internally by approx.  $30^\circ$  in each case, without the need to osteotomize the fibula as well. The result is fixed with two crossed Kirschner wires inserted from the outside through the skin (■ Fig. 4.14). A lower leg non-walking cast is applied for four weeks. Since the correction is usually performed on both sides, the child must be mobilized for four weeks in a wheelchair. At the end of this time, a check x-ray is recorded, the Kirschner wires are removed without anesthesia and lower-leg walking-casts are applied for a further two weeks. This operation leaves a barely visible scar and is associated with low morbidity and few complications.

#### Correction of genua vara and genua valga

On the basis of the previously mentioned measurements with the determination of the apex of the angulation, the intersection of the angle-bisecting line with the concave bone edge is the location for a closing-wedge osteotomy. The intersection with the convex bone edge is the site for an opening-wedge osteotomy. If an osteotomy is performed



■ Fig. 4.14. AP x-rays of the lower leg 4 weeks after *supramalleolar tibial derotation osteotomy* for a pathological lateral torsion of the tibia in a 10-year old boy

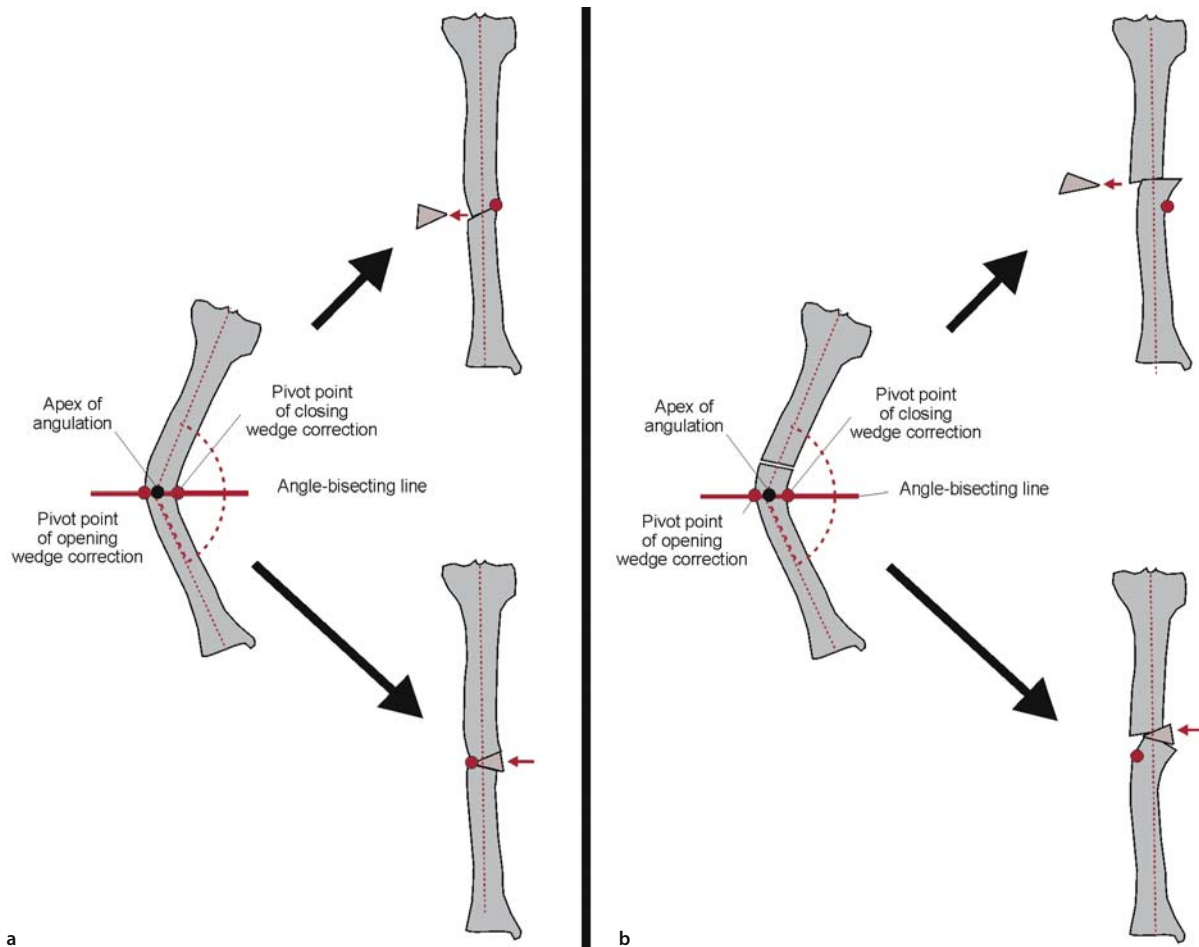
through the apex, the angulation alone will completely restore the proximal and distal bone axes (osteotomy rule 1; ■ Fig. 4.15a). If the osteotomy is not performed at apex level, the angulation alone will result in a translation of the proximal and distal bone axes, and an additional translation in the opposite direction will be needed to restore the axis (osteotomy rule 2; ■ Fig. 4.15b) [12, 13].

In addition to these rules, the status of the growth plates must also be taken into account. Accordingly, the operation is usually performed at infracondylar level in *small children*, i.e. below the tibial tuberosity. We generally perform a transverse osteotomy, produce the desired correct position (including any derotation) and fix the result with crossed Kirschner wires and long-leg cast. It may also be possible to remove a wedge, including in an oblique plane, so that an axial correction occurs at the same time as a result of rotation. However, the inclination of this plane must be calculated very carefully. Another option is a dome-shaped osteotomy with a rounded cut surface. This allows for subsequent cast correction. We do not use this method, however, since we never fix with plaster on its own.

In *older children* we fix the result with an external fixator or low-contact plates with fixed-angled screws. The Tomofix plate is particularly suitable for this purpose (see also below for further details). Here too, if the physes are still open the osteotomy is performed at infracondylar level and, if they are closed, at transcondylar level. These types of stabilization permit immediate weight-bearing, which is a significant advantage if the osteotomy has to be performed on both sides. The young patients can be mobilized and walk with crutches after just a few days,



## 4.2.1 · Are children twisted when they have an intoeing gait or warped if they are knock-kneed



■ **Fig. 4.15a, b.** Osteotomy rules according to Paley [12, 13]. **a** Osteotomy rule no. 1: If the osteotomy runs through the apex of the angulation, only a change in the angle is required to restore the proximal and distal bone axes. If the apex of the angulation on the convex side of the angle-bisecting line is selected, this results in an opening-wedge cor-

rection, compared to a closing-wedge correction on the concave side. **b** Osteotomy rule no. 2: If the osteotomy is not performed at apex level, a change in the angle alone will result in a translation of the proximal and distal bone axes. An additional translation in the opposite direction will therefore be needed to restore the axis

when the initial pain has subsided. The correction can be performed either by the removal or insertion of a wedge. Continuous correction by unilateral lengthening is also possible.

In the English-speaking literature, the procedure of partial *physeal closure (hemiepiphysiodesis)* is recommended for the correction of axial deformities. We do not use this method since it is not very reliable. The subsequent extension of the bridge is difficult to predict and a rotational deformity often occurs (if the bridge is eccentric). Overcorrection can also occur, thereby necessitating a physeal closure on the other side of the tibia which, in turn, can cause shortening. Undercorrection is more common, however, in view of the inadequate growth potential before completion of growth. For these reasons, we consider that a corrective osteotomy is more appropriate.

### Complex corrections

Young patients with systemic diseases (e.g. vitamin D-resistant rickets, achondroplasia etc.; ► chapter 4.6) can sometimes suffer from complex deformities with axial defects, both in the upper and lower leg, with concomitant rotational deformities. In such cases, the orthopaedist must always ensure that the knee is horizontally aligned after any correction. This condition often means that correction is required in both the upper and lower leg. The procedure is usually performed at supracondylar level on the femur and at infracondylar level in the lower leg (■ Fig. 4.11). If axial and/or rotational corrections are associated with length differences, we currently use the »Taylor Spatial Frame« developed by J.C. Taylor in 1997« [3] (see ► chapter 4.2.2. and the website [www.jcharlestaylor.com](http://www.jcharlestaylor.com) for further details).





■ Fig. 4.16. 16-year old girl postoperatively after *bilateral supracondylar femoral derotation osteotomy* with AO-low contact plate (LCP) with fixed angled screws. This fixation allows immediate weight bearing.

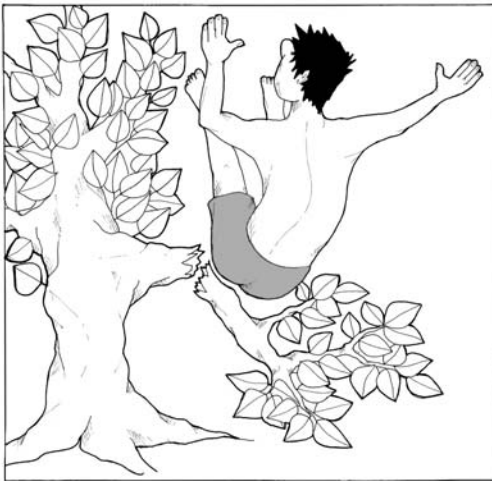
! **Axes and torsions of the lower extremities undergo major changes during growth, and it is important to be aware of this fact in order to be able to distinguish pathological forms. Deformities only require correction in extreme cases, when surgery is always essential as conservative measures are ineffective.**

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### 4.2.2 Do children go »off the straight and narrow« when the pelvis is oblique? – or: Causes and need for treatment of pelvic obliquity?

Leg length discrepancy is one of the commonest conditions seen in the pediatric or orthopaedic clinic. Uncertainty often prevails concerning the pathological significance and need for treatment of such leg length discrepancies. Since two-thirds of the population are thought to have a pelvic obliquity of up to 1 cm [14], this condition must be considered as »physiological«.



#### ! Definition

Directly or indirectly measured difference in leg length of 1 cm or more.

#### Occurrence

Minor leg length discrepancies are extremely common, although exact epidemiological figures are not available simply because the measuring methods are very imprecise. Leg length discrepancies of less than 1 cm are clinically irrelevant. Dispute exists as to whether a leg length discrepancy of 1 cm is clinically significant [28]. What is clear, however, is that a length difference of 2 cm certainly is relevant in view of the effects on the spinal column [37]. Figures relating to differences between 1 and 2 cm are not available in the literature, therefore, precisely because the indications for leg length equalization are defined very differently. In our clinic around 5 patients out of 100 fall into this group. Better frequency figures are available for length discrepancies of more than 2 cm. One epidemiological study in France calculated a prevalence of 1/1,000 for leg length discrepancies requiring equalization, with a male:female ratio of 2 : 1 [16]. The authors suspect that their study design has resulted in figures that tend to be on the low rather than the high side.

#### Etiology, classification

We make a basic distinction between:

- *true leg length discrepancy*, which is caused by the shortening or lengthening of individual sections, and
- *functional leg length discrepancy*, which is caused by knee or hip flexor contractures or adduction or abduction contractures of the hip or by a fixed equinus position of the upper ankle.

■ Table 4.3 presents the causes of true leg length discrepancies during childhood and adolescence [15]. The commonest cause of differences of more than 1 cm is probably growth stimulation after metaphyseal, and possibly diaphyseal fractures. Posttraumatic shortening, on the other hand, is rare. Injuries to the epiphyseal plate usually lead primarily to an axial deformity and only secondarily to shortening. A primary reduction in growth would only be expected if the injury affected the whole of the plate, which is extremely rare.

The healing of shaft fractures in a shortened position leads to leg lengthening rather than shortening because the growth stimulation resulting from the remodeling process overcompensates for the shortening [19]. Of the congenital forms, a *hemihypertrophy* is more common than *hemihypotrophy* [4]. *Hemihypertrophies* occur in the Klippel-Trenaunay-Weber syndrome, Proteus syndrome (► Chapter 4.6.6.3) [18], Albright syndrome (► Chapter 4.6.2.31) [22] and neurofibromatosis [39] (► Chapter 4.6.6.2), although they can also be associated with primitive tumors of the liver or kidney [4].

Scoliosis and impaired intelligence are fairly rare in *hemihypertrophy*, in contrast with *hemihypotrophy*, where these symptoms are often observed. *Hypotrophy* is sometimes associated with chromosomal mosaic forms or the Russell-Silver syndrome [4]. Axial deformities can lead to leg length discrepancies for purely geometric reasons if they are one-sided. This occurs relatively frequently in Legg-Calvé-Perthes disease (► Chapter 3.2.5). Since knock knees and bow legs are usually bilateral, no pelvic obliquity arises. They can occur unilaterally however, e.g. in connection with osteochondromas (► Chapter 4.6.2.31).

#### Clinical features, diagnosis

The procedure for *indirect leg length measurement* (evaluation of the iliac crest) is described in ► chapter 3.1.1. For the *direct measurement* we proceed as follows: We mark the knee joint space on the skin. Using a flexible tape measure graduated in centimeters we measure from the anterior superior iliac spine down to the external malleolus (or to the heel) to establish the full leg length, down to the knee joint space for the upper leg length and from the mark at knee level down to the ankle for the lower leg length. Despite the use of the tape measure the direct measurement is less accurate than the indirect measurement because of the skin mobility. If contractures are

present, however, the direct measurement can provide a more realistic assessment of the length relationships than the indirect method. For the *radiographic measurement* a semi-opaque ruler is fastened to the skin, in line with the bone, over the full length of the leg. Bilateral x-rays of the hip, knee and ankle, with inclusion of the ruler, are used to calculate the lengths. An even more precise method is the use of the *computed tomogram*. Marking the joint heights with *ultrasound* with the aid of a calibrated ruler also produces a very precise measurement [36].

### Prognosis

An awareness of the basic principles and factors that influence *growth* is essential for the treatment of leg length discrepancies. The basic processes are described in ► chapters 2.2.1–2.2.3, which address the growth rates and the difference between chronological age and skeletal age. The following *parameters* must also be known before leg length discrepancies can be treated:

- the *relative* growth of the affected sections of the extremity compared to the other side,
- the *expected* growth of the affected sections of the extremity,
- the effect of *shortening* or *lengthening measures* on growth.

Knowledge of the growth potential of the femur and tibia is particularly important. ■ Fig. 4.17 shows the *average length* of these two bones in boys and girls with the corresponding single and double standard deviations [2]. ■ Fig. 4.18, on the other hand, shows the *residual growth* in the femur and tibia in girls and boys according to the skeletal age in each case. Follow-up monitoring is very important for establishing a growth prognosis. Three length measurements at minimum intervals of 18 months are required for a reasonably reliable prognosis. The growth disturbance can be calculated according to the following *formula* (as a percentage) [13]:

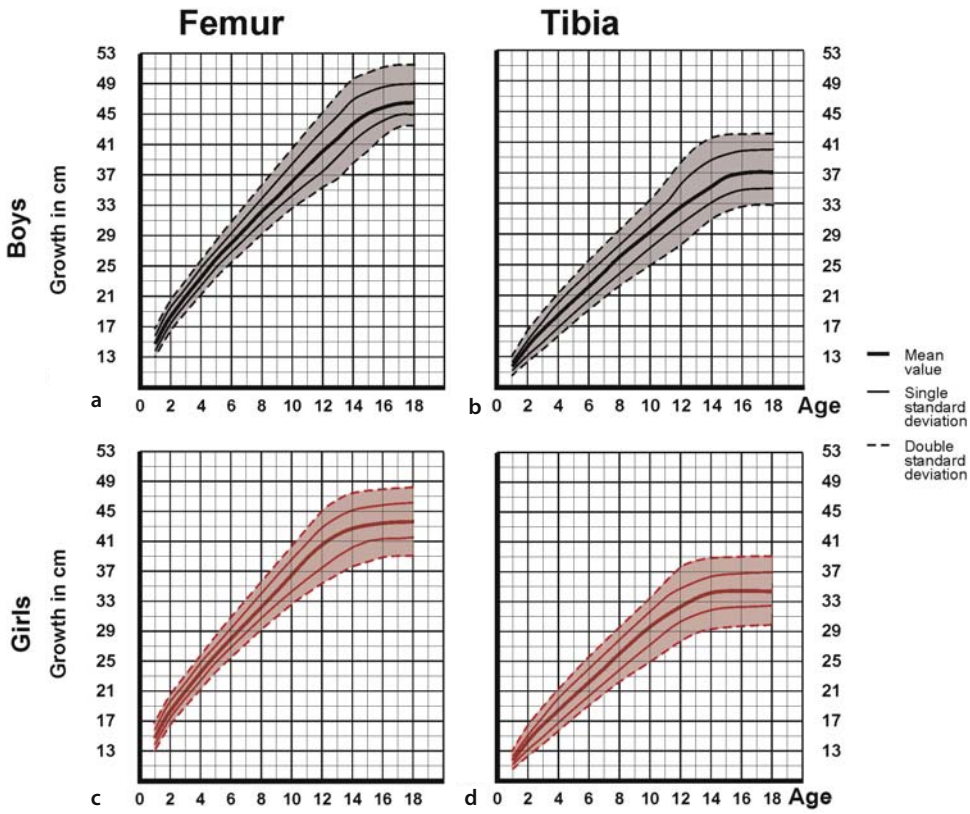
$$\text{»Growth disturbance«} = \frac{(\text{Growth normal side}) - (\text{Growth short side})}{\text{Growth normal side}}$$

The *growth disturbances* can then be classified into *levels of severity* (■ Table 4.4). *Prognoses* always assume that the growth disturbance progresses proportionally throughout the growth period. But this only applies to a limited extent and also depends greatly on the clinical situation. Thus, a traumatically impaired growth plate does not lead to a disproportionately increasing discrepancy since the plate no longer grows at all. On the other hand, the percentile at which the patient is located in terms of growth plays a major role. A 14-year old boy, for example, has only 5 mm

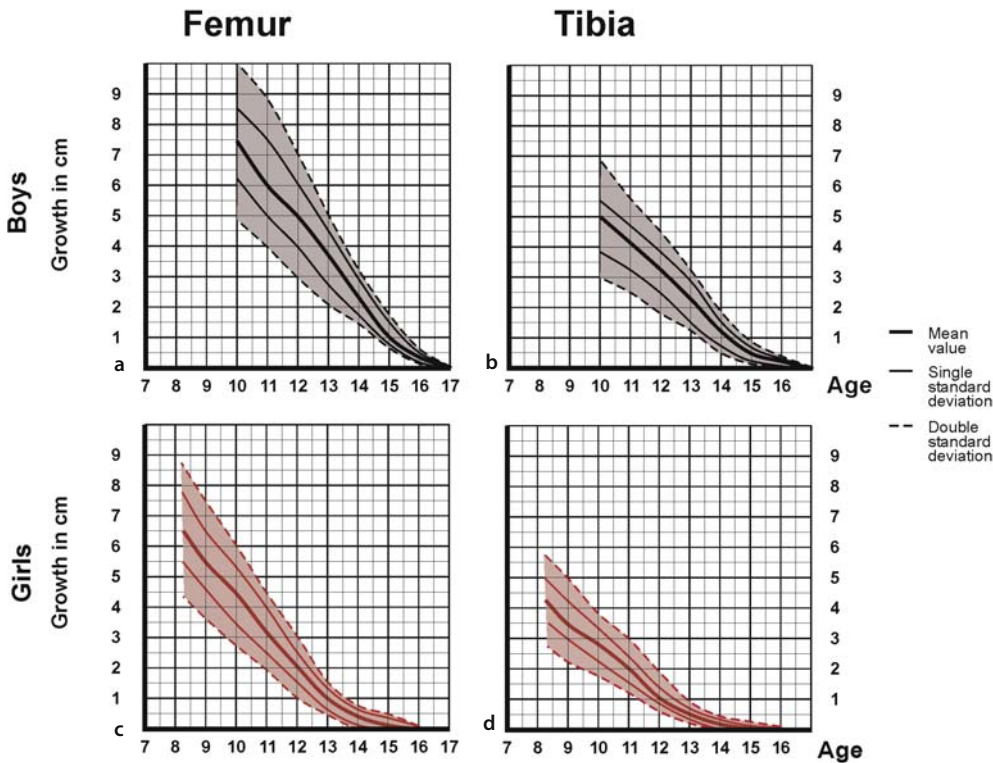
■ Table 4.3. Causes of leg length discrepancies during growth [19]

	Due to growth retardation	Due to growth stimulation
<b>Congenital</b>	Congenital hemiatrophy (essential hypoplasia) Congenital atrophy with skeletal anomaly (fibular aplasia, femoral aplasia, coxa vara etc.) Ollier enchondromatosis Dysplasia epiphysealis hemimelica, hereditary multiple exostoses Congenital hip dislocation Clubfoot	Partial gigantism with vascular anomalies (Klippel-Trenaunay-Weber syndrome; hemarthrosis in hemophilia, Proteus syndrome)
<b>Infections</b>	Destruction of the epiphyseal plates by osteomyelitis (femur, tibia), Tuberculosis (hip, knee, foot) Purulent (septic) arthritis	Osteomyelitis of the femoral and tibial diaphyses, Brodie abscess, Tuberculosis of the femoral and tibial metaphyses (tumor albus genus), Elephantiasis after soft tissue infections thrombosis of the femoral and iliac veins
<b>Paralytic conditions</b>	Poliomyelitis, other (spastic) paralytic conditions	–
<b>Tumors</b>	Osteochondromas (solitary exostoses) Giant cell tumors Osteitis fibrosa cystica Neurofibromatosis	Hemangiomas Lymphangiomas Giant cell tumors Osteitis fibrosa circumscripta Neurofibromatosis Fibrous dysplasia (Albright syndrome)
<b>Traumata</b>	Injuries to the physeal plate (separations, operations, etc.) Severe burns	Diaphyseal and metaphyseal fractures of the femur and tibia (osteosynthesis) Surgery to the metaphysis (periosteal separation, graft harvesting, osteotomy etc.)
<b>Mechanical</b>	Prolonged immobilization	Traumatic arteriovenous aneurysms
<b>Other causes</b>	Legg-Calvé-Perthes disease Slipped capital femoral epiphysis Irradiation of the femoral and tibial physeal plates Axial deformity (knock knees or bow legs, genu antecurvatum, genu recurvatum)	–

## 4.2.2 · Do children go »off the straight and narrow« when the pelvis is oblique?



■ Fig. 4.17a–d. Growth of femur (distal physis) and tibia (proximal physis) in boys (a, b) and girls (c, d). (After [2])



■ Fig. 4.18a–d. Residual growth for the femur (distal physis) and tibia (proximal physis) in boys (a, b) and girls (c, d). (After [2])



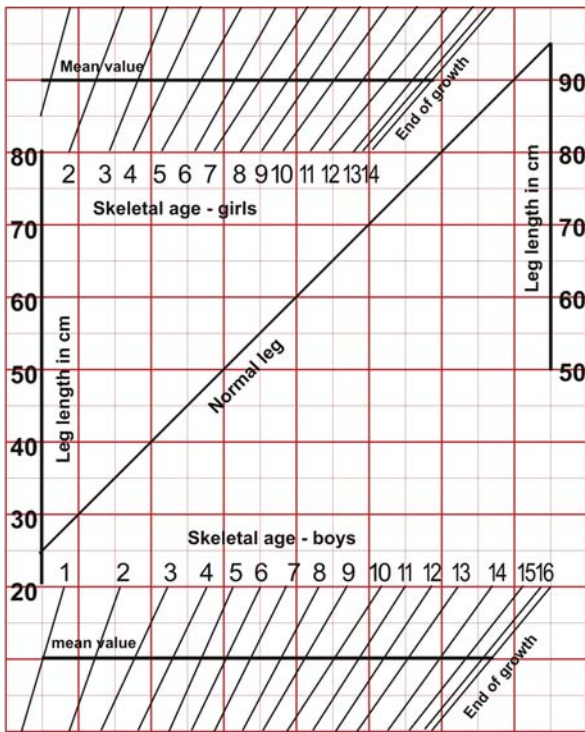


Fig. 4.19. Moseley chart (after [36]): The final length can be derived by plotting the length on three dates over a minimum period of 1.5 years against the respective skeletal age and extending the resulting line at the corresponding inclination. The lengths of the normal and shortened legs are entered one below the other

**Table 4.4. Levels of severity of growth disorders**

Severity	Growth disorder
Mild	0–10%:
Moderate	11–20%:
Severe	21–30%:
Very severe	>30%

of residual growth in the distal femur if he is on the 10th percentile in terms of height, but as much as 2.5 cm on the 90th percentile. We must also take account of the fact that the linear progression of growth is influenced by our surgical measures. If a proportionately increasing length discrepancy is present, a relatively good prognosis for the difference on completion of growth can be provided on the basis of three measurements at intervals of 18–24 months (in the natural course of events without external influencing factors). The *straight-line graph* according to Moseley is particularly suitable for this purpose [27] (Fig. 4.19).

Even though this method also has its own shortcomings it remains the most accurate way of predicting a leg length discrepancy. It requires radiographic leg length measurements and skeletal age calculations at three dif-

**Table 4.5. Multipliers for upper extremities.** (After [11])

Age (years)	Boys	Girls
Birth	3.14	3.00
1	2.37	2.38
2	2.08	1.97
3	1.89	1.79
4	1.74	1.65
5	1.63	1.54
6	1.54	1.45
7	1.46	1.37
8	1.39	1.31
9	1.33	1.25
10	1.28	1.20
11	1.23	1.15
12	1.19	1.09
13	1.15	1.05
14	1.10	1.03
15	1.05	1.02
16	1.02	1.00
17	1.01	1.00
18	1.00	1.00

**Table 4.6. Multipliers (M) for lower extremities of boys.** (After [11])

Age (years + months)	M	Age (years + months)	M
At birth	5.080	7+6	1.520
0+3	4.550	8+0	1.470
0+6	4.050	8+6	1.420
0+9	3.600	9+0	1.380
1+0	3.240	9+6	1.340
1+3	2.975	10+0	1.310
1+6	2.825	10+6	1.280
1+9	2.700	11+0	1.240
2+0	2.590	11+6	1.220
2+3	2.480	12+0	1.180
2+6	2.385	12+6	1.160
2+9	2.300	13+0	1.130
3+0	2.230	13+6	1.100
3+6	2.110	14+0	1.080
4+0	2.000	14+6	1.060
4+6	1.890	15+0	1.040
5+0	1.820	15+6	1.020
5+6	1.740	16+0	1.010
6+0	1.670	16+6	1.010
6+6	1.620	17+0	1.000
7+0	1.570		



## 4.2.2 · Do children go »off the straight and narrow« when the pelvis is oblique?

■ **Table 4.7.** Multipliers (*M*) for lower extremities of girls. (After [11])

Age (years + months)	M	Age (years + months)	M
At birth	4.630	6+0	1.510
0+3	4.155	6+6	1.460
0+6	3.725	7+0	1.430
0+9	3.300	7+6	1.370
1+0	2.970	8+0	1.330
1+3	2.750	8+6	1.290
1+6	2.600	9+0	1.260
1+9	2.490	9+6	1.220
2+0	2.390	10+0	1.190
2+3	2.295	10+6	1.160
2+6	2.200	11+0	1.130
2+9	2.125	11+6	1.100
3+0	2.050	12+0	1.070
3+6	1.925	12+6	1.050
4+0	1.830	13+0	1.030
4+6	1.740	13+6	1.010
5+0	1.660	14+0	1.000
5+6	1.580		

■ **Table 4.8.** Multipliers (*M*) for height in boys. (After [11])

Age (years + months)	M	Age (years + months)	M
At birth	3.535	8+6	1.351
0+3	2.908	9+0	1.322
0+6	2.639	9+6	1.298
0+9	2.462	10+0	1.278
1+0	2.337	10+6	1.260
1+3	2.239	11+0	1.235
1+6	2.160	11+6	1.210
1+9	2.088	12+0	1.186
2+0	2.045	12+6	1.161
2+6	1.942	13+0	1.135
3+0	1.859	13+6	1.106
3+6	1.783	14+0	1.081
4+0	1.731	14+6	1.056
4+6	1.675	15+0	1.044
5+0	1.627	15+6	1.030
5+6	1.579	16+0	1.021
6+0	1.535	16+6	1.014
6+6	1.492	17+0	1.010
7+0	1.455	17+6	1.006
7+6	1.416	18+0	1.005
8+0	1.383		

■ **Table 4.9.** Multipliers (*M*) for height in girls. (After [11])

Age (years + months)	M	Age (years + months)	M
At birth	3.290	8+6	1.254
0+3	2.759	9+0	1.229
0+6	2.505	9+6	1.207
0+9	2.341	10+0	1.183
1+0	2.216	10+6	1.160
1+3	2.120	11+0	1.135
1+6	2.038	11+6	1.108
1+9	1.965	12+0	1.082
2+0	1.917	12+6	1.059
2+6	1.815	13+0	1.040
3+0	1.735	13+6	1.027
3+6	1.677	14+0	1.019
4+0	1.622	14+6	1.013
4+6	1.570	15+0	1.008
5+0	1.514	15+6	1.009
5+6	1.467	16+0	1.004
6+0	1.421	16+6	1.004
6+6	1.381	17+0	1.002
7+0	1.341	17+6	1
7+6	1.309	18+0	1
8+0	1.279		

ferent times. The effect of leg-lengthening measures during growth is difficult to estimate. Lengthening performed on the lower leg generally leads to a reduction in residual growth, i.e. to an increase in the discrepancy. This does not apply for the upper leg, where growth stimulation is even observed at times. A particularly problematic aspect is the fact that this effect cannot be accurately predicted in the individual case [21, 24].

A new method for producing growth forecasts is the use of Paley's *multiplier* [33]. Paley has developed formulas for calculating the length discrepancy on completion of growth, for selecting the correct timing of epiphysiodesis and for determining final height. However, the calculations are based on chronological age and take no account of skeletal age. The advantage of this method is that a forecast can be worked out immediately during the consultation using ■ Tables 4.5 to 4.9. Nevertheless the skeletal age and height percentiles should also be considered when establishing the indication for surgical measures.

Formula for forecasting the length discrepancy in *congenital deformities*:

$$\ddot{A}_m = \ddot{A} \cdot M,$$

where  $\ddot{A}_m$  = discrepancy on completion of growth,  $\ddot{A}$  = current discrepancy,  $M$  = multiplier.

Formula for forecasting the length discrepancy in *growth-related disorders* (e.g. Ollier disease, poliomyelitis, posttraumatic growth disorder):

$$\ddot{A}_m = \ddot{A} + I \cdot G,$$

where  $\ddot{A}_m$  = discrepancy on completion of growth,  $\ddot{A}$  = current discrepancy,  $I$  = growth retardation =  $1 - (S - S') / (L - L')$ ,  $G$  = residual growth =  $L(M - 1)$ .  $S$  and  $S'$  are measurements on the shorter extremity at two different times separated by at least 6 months,  $L$  and  $L'$  are corresponding measurements on the longer side.

Selection of the *timing of epiphysiodesis*:

$$M_{\hat{a}} = LM / (LM - \hat{a} / \hat{e}),$$

where  $M_{\hat{a}}$  = multiplier at the time of epiphysiodesis,  $\hat{a}$  = desired correction,  $\hat{e}$  = characteristic factor for epiphysal plate: 0.71 for distal femur, 0.57 for proximal tibia and 0.67 for femur and tibia together.

$A_{\hat{a}}$  = age at the time of  $M_{\hat{a}}$  based on the table.

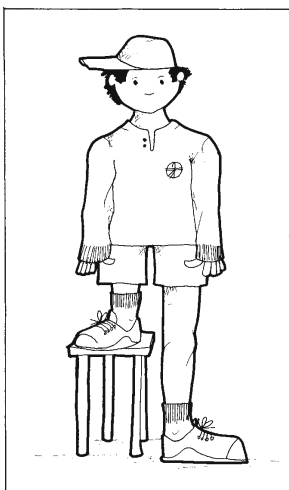
Example: The length of the femur in a 10-year old girl is 37 cm, the discrepancy to be corrected ( $\hat{a}$ ) is 3 cm. The multiplier, according to ■ Table 4.7 is 1.19. According to the formula  $LM / (LM - \hat{a} / \hat{e}) = 44.3 / (44.35 - 3 / 0.71) = 1.09$  for  $M_{\hat{a}}$ , which means that the epiphysiodesis should be performed when the multiplier is 1.1, i.e. at the age of approx. 11.5 years (■ Table 4.7).

The multipliers for the upper and lower extremities for boys and girls and for calculating final height are listed in ■ Tables 4.5 to 4.9.

### Treatment

The following *options* are available for treating leg length discrepancies:

- Conservative treatment with shoe elevation or orthoses,
- Epiphysiodesis,
- Surgical leg shortening,
- Surgical leg lengthening.



### Indication

! Leg length discrepancies of 1.5 cm or less do not require treatment.

We reduce leg length discrepancies *between 1.5 and 2 cm* (at least by half) by means of a heel or sole wedge in the shoe *during puberty*. After completion of growth, discrepancies of less than 2 cm do not require treatment, but during puberty we equalize the discrepancy because of the risk of scoliosis. Although it has not been proven that a discrepancy of this size can actually be responsible for the development of scoliosis, we believe that equalization is still useful since it is a trivial and low-cost measure. This equalization can be achieved with the aid of a heel wedge in the shoe, which can be purchased as a ready-made product from a shoe retailer.

A leg length discrepancy of 2 cm or more, including after completion of growth, should be equalized. The pelvic obliquity promotes the development of scoliosis and the one-sided loading of the muscles, including during adulthood. Although equalization via shoe-based measures alone is possible for a discrepancy of up to 3 cm, the heel wedge will not suffice for this purpose since it cannot be more than 1 cm high otherwise a standard shoe will no longer fit. Both the heel and sole must be raised. Even for a discrepancy of 2 cm, the heel on its own should not be elevated otherwise the shoe will produce an equinus foot position. The fact that the height is equalized on one side only, rather than both sides as is usual for example with high heels, is disadvantageous. The heel should not be raised by more than 1 cm compared to the sole in the forefoot area.

The question is whether patients are willing to continue wearing such inserts for the rest of their lives. If a patient would like to undergo surgical correction this can be indicated from a difference of 2 cm. More than 3 cm of correction with a standard shoe is often cosmetically unacceptable and also problematic in functional respects. The risk of supination trauma increases in line with the amount of elevation. Discrepancies of more than 4–5 cm must be equalized with an orthosis that also stabilizes the back of the foot and the lower leg. On the other hand, orthopaedic appliances can pose cosmetic and functional problems for patients that can be resolved by surgical measures. The discrepancy can be equalized either by lengthening the shorter side or shortening the longer side.

The following factors should be considered when deciding whether surgery is indicated:

- The patient should be aware of *all the options* and be involved in the decision-making process for the surgical procedure. It is particularly important that the patient is aware of the possible complications and the effort involved in terms of time, technical complexity and, in particular, psychological stress.
- A *shortening* can only offset relatively minor discrepancies, in contrast with a lengthening procedure, in

#### 4.2.2 · Do children go »off the straight and narrow« when the pelvis is oblique?

which (in theory) lengthening of over 30 cm is possible. Maximum acute shortening of 4 cm is possible in the femur and of 3 cm in the lower leg.

- An *epiphysiodesis* may only be implemented relatively shortly before completion of growth otherwise the calculation of the effect is too unreliable.
- It is always problematic if, when one leg is affected by a disorder, operations are performed on the *other leg*. This limits the options for shortening, since the shorter leg is usually the diseased one. Consequently, the shortening osteotomy almost invariably has to be performed on the healthy leg. Even if the risk of complications is lower with the shortening osteotomy compared to a lengthening procedure, this aspect must be taken into consideration. A complication in the healthy leg is much more problematic than one in the diseased leg. Epiphysiodesis on the healthy leg for correcting fairly small differences is relatively unproblematic, since the transcutaneous method rarely involves any complications, its morbidity is extremely low and no functional restriction is expected. It also produces an extremely satisfactory esthetic result.
- Another important factor is the expected *final length*. Tall people are more likely to accept a shortening procedure than short people, and the wishes of the patient should be respected particularly in this situation.
- For leg length discrepancies of *over 5 cm*, lengthening is the only option for equalization. If the expected difference is more than 8 cm, the lengthening must be performed in stages. We never perform lengthening of more than 8 cm at a time in a given stage as the complication rate rises sharply above this level.
- The leg length equalization should always be performed at the *site of the discrepancy* (upper or lower leg). This simple truth requires further elaboration. The objective is always to achieve knees at the same height. Let us assume that a patient has a leg length discrepancy of 3.5 cm with shortening at the femur of 2 cm and at the tibia of 1.5 cm. In this case it would be absurd to perform lengthening procedures on both the upper and lower legs. Since a maximum height difference of 2 cm is acceptable for the knees, we would only lengthen the femur in this case.
- If *substantial differences of over 20 cm* are anticipated, then one should consider very carefully whether lengthening is appropriate at all. Such patients usually suffer from a *longitudinal deficiency of the fibula* (i.e. a proximal femoral deficiency is combined with a fibular aplasia and the absence of the lateral rays on the foot; ► chapters 3.2.7, 3.3.6 and 3.4.5). These patients often have cruciate ligament aplasia, as well as major problems in stabilizing the ankle as a result of the absent or dysplastic lateral malleolus. We stick to a relatively simple rule: If three or more rays are present in the foot, the possibility of lengthening can

be considered. But if only two rays are present, we try to persuade the child and the parents that life-long orthotic provision is more useful than the lengthening procedure. Another problematic situation is lengthening in association with a proximal femoral deficiency and an unstable hip joint. Here, too, we advise against lengthening. In such cases, a rotation-plasty is a possible solution [1, 6] (► Chapters 3.2.7 and 4.5.5).

- If major discrepancies exist, a *combined approach* may be appropriate, with lengthening on the shorter side and percutaneous epiphysiodesis on the longer side shortly before completion of growth.
- An extremely problematic situation is lengthening in *poliomyelitis*. It should be borne in mind that the lengthening of the muscles in this situation is invariably associated with a loss of power. Patients with poliomyelitis already have muscle power problems and only just manage to walk. Any lengthening procedure involves the risk of a deterioration in, or even complete loss of, walking ability (► Chapter 4.7.4.5).
- Caution is required when assessing *patients with dwarfism* who express a wish for double-sided lengthening. This often involves a height gain of 25 cm or more. The desire to be taller is understandable in these patients, and it makes sense particularly if a normal height can be achieved, i.e. of at least 150 cm (4 feet 11 inches). Since the arms are almost always shortened as well as the legs, patients with lengthened legs and short arms look out of proportion. Consequently, the possibility of arm lengthening must also be considered. Although many such lengthening procedures have been implemented worldwide on patients with dwarfism (particularly with achondroplasia) [9], the fact should not be overlooked that this elaborate and very time-consuming process is associated with considerable, albeit temporary, psychological stress [35]. The complication rate is also high for such major lengthening procedures.

We also have experience with bilateral lengthening. The rule that lengthening should not exceed 8 cm in each stage also applies in these cases. We primarily lengthen both lower legs and only secondarily both upper legs. If the attempt has to be discontinued after the first stage of the attempt, disproportionately long lower legs are much more readily acceptable from the esthetic standpoint than correspondingly overlong upper legs. We therefore tend to discourage patients with dwarfism from undergoing lengthening surgery. Only if they still persist with their request are we prepared to perform this elaborate procedure. It is important for them to meet other patients who have already undergone the procedure so that they have a realistic idea of the impending mental and physical effort involved.

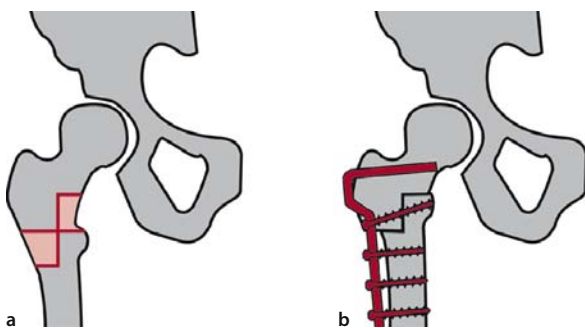
## Techniques

### Epiphysiodesis

Epiphysiodesis can be performed as a temporary or definitive measure. We no longer use the temporary stapling method proposed by Blount since it is not very reliable. Definitive epiphysiodesis cannot be performed until relatively shortly before completion of growth. For several years we have been using a very simple percutaneous method of epiphysiodesis. Through a stab incision the germinative layer of the epiphyseal plate is destroyed with a wide oscillating drill [11, 30]. The method is reliable with low morbidity and only leaves tiny, barely noticeable scars. We generally advise against weight-bearing for the first three weeks. If this is not possible however, full weight-bearing is also perfectly possible from the outset. We have also performed this epiphysiodesis on both sides at the same time (in patients with macrosomia) with immediate postoperative full weight-bearing. This method is also suitable for physal closure following a tumor resection on the other side. Here, too, full weight-bearing is required from the outset.

### Shortening osteotomy

Leg shortening of up to 4 cm for the femur and up to 3 cm for the lower leg is possible. The most reliable type of shortening procedure at femoral level is an intertrochanteric osteotomy (■ Fig. 4.20) although, for anatomical reasons, shortening only up to 3 cm or so is possible at this point. A higher figure is only possible if the osteotomy is performed in the shaft area, but the subsequent healing process is not as favorable here. Shortening in excess of 4 cm is not possible because the muscles would be weakened for a very long time postoperatively. It can take 1–2 years before normal muscle power is restored. In view of the relative overlength of the muscles, the risk of thrombosis is also fairly high. This also applies to the lower leg, where the osteotomy is usually performed through the diaphysis, followed by stabilization with plate osteosynthesis. In general, the complication rate for a shortening osteotomy is lower than that for the lengthening procedure, as we have confirmed in our own study [20].



■ Fig. 4.20a, b. Principle of intertrochanteric shortening osteotomy with Z-shaped osteotomy and fixation with 90° angled blade plate. **a** Before the operation, planned osteotomies, **b** postoperatively

### Surgical leg lengthening

Four basically different methods are available for surgical leg lengthening:

- diaphyseal osteotomy, lengthening with external fixator, followed by packing of the distracted segment with cancellous graft and plating (*Wagner method*).
- lengthening by an osteotomy (compactotomy) in the diaphyseal or metaphyseal area, callus distraction with an external fixator (*«callotaxis», «Ilizarov method»*).
- distraction of the epiphyseal plate with external fixator (*«chondrotaxis»*).
- externally-controlled lengthening by means of a diaphyseal osteotomy and the fitting of an intramedullary lengthening apparatus [3, 8, 17].

*Callotaxis* according to Ilizarov's method has gained the most widespread acceptance in recent years [5, 14, 20]. The Wagner method is associated with too many complications, as has been shown not only by a study in our own hospital [20], but also many other authors [15, 25, 29]. The difference between the Wagner method and the Ilizarov method concerns not so much the lengthening apparatus, but rather the fact that a cancellous bone graft is inserted and stabilized with a plate in the Wagner method after the appropriate length has been achieved in the distracted segment. This is a non-biological technique. Since the necrotic cancellous graft remodels itself into weight-bearing bone only very slowly, fractures and plate breakages were common.

*Distraction epiphysiolysis* has also failed to catch on, since premature physal closure often occurs as a result of the distraction of the epiphyseal plate. The advantage of the method is that no osteotomy is required. However, the premature physal closure means that the final amount of lengthening is extremely difficult to predict, since shortening then occurs after the lengthening. This method is unreliable and has not gained widespread acceptance.

One method that has established itself worldwide however is *callotaxis*, or the *«Ilizarov method»*. This involves the following steps:

- *Fitting of the external fixator.*
- *Compactotomy*, if possible in the metaphyseal, or possibly the diaphyseal area. This involves an osteotomy with the chisel, with preservation of the medullary vessels. Alternatively, a transcutaneous osteotomy can be performed with a drill. It is important to preserve the periosteum.
- *Compression of the osteotomy* for several days. The following rule applies: »age in years« = »number of days«.
- Followed by *lengthening* with the external fixator by 1 mm a day.
- After the desired length has been achieved the *external fixator is left in place until consolidation*, ideally with full weight-bearing.

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- As a rule of thumb the fixator should be fitted for the following length of time: 40 days per 1 cm of lengthening.
- Monitoring of callus formation by *ultrasound* [26].
- The external fixator can then be *removed* if there are radiographically visible incipient signs of cortex formation in the bone mass of the lengthened segment.
- The external fixator is removed first, leaving the screws in place. If the bone does not bend the screws can then be removed a few days later.

#### Medullary nails

*Internal medullary nails* represent an attractive alternative as they also allow the length to be controlled externally. Three such systems are currently in use. With the ISKD nail (Intramedullary Skeletal Kinetic Distractor), the rotational movement that occurs during walking is translated into the lengthening of a threaded medullary rod [8]. Rotational movements of 3° are sufficient for the lengthening. With the Albizzia nail (named for a fast-growing Mimosa tree), manual rotational movements of 20° must be made in order to produce the lengthening [17].

The most elegant solution is the Fitbone nail, system in which a telescopic motorized medullary nail is lengthened by remote control [3]. The attractive feature of this system is that it is much more comfortable for the patient compared to external fixators. The drawback of all intramedullary nails is that they can only be used after the plates have largely closed and if the bone structure is fairly normal. Nor should the risk of complications be underestimated (they differ from those associated with external fixators, but they are also not negligible). In cases of substantial lengthening, a weakening of the lengthening effect can occur as a result of increasing muscle resistance. Implant failures can also occur.

These implants are also very expensive. Only a small number of centers worldwide are authorized by the manufacturer to use the Fitbone nail. Recently we are now authorized to use the Fitbone nail in our hospital, our experience to date is limited but positive [23].

#### Fixator systems

We distinguish between the following basic options:

- Ring fixators (Ilizarov-type), Taylor Spatial Frame,
- rigid monolateral fixators (e.g. Wagner apparatus, Orthofix, Monotube),
- monolateral fixators with angulation (e.g. Heidelberg fixator, ExFiRe).

Each system has its own *advantages and disadvantages*. *Ring fixators* offer unlimited options for correction. Angulations in all directions can be corrected, and even rotational correction is possible. Joints can also be bridged. The disadvantages are the discomfort (particularly in the upper leg) and edema formation. The esthetic shortcom-



■ Fig. 4.21a b. 8-year old girl with longitudinal deficiency of the fibula and *tibial lengthening* with circular fixator (Ilizarov apparatus). a During the lengthening procedure. b One year after completion of the lengthening by 10 cm (arrows former lengthened segment)

ings of the Ilizarov apparatus contrast with the much better-looking Taylor Spatial Frame. Very precise corrections are possible with this tool.

We use ring fixators primarily on the lower leg (■ Fig. 4.21, 4.22), particularly if long segments are involved (with a high probability of secondary axial bowing), if dysplasia of the knee or ankle is present, in which case the affected joint must be included, primarily or secondarily, in the treatment. *Monolateral rigid fixators* are more comfortable. We use the Monotube particularly on the lower leg, provided the lengthened segments are not too long. These fixators can also be dynamized, which promotes callus formation. On the upper leg, rigid monolateral fixators are suitable only for the lengthening of fairly short segments, since axial varus deviations can readily occur at this level. *Monolateral fixators with angulation* have not proved effective in our experience. The angulator is usually well away from the apex of the deformity and does not correct properly since the pivot point is usually incorrectly located. Moreover, since huge forces are generated these can lead to the mechanical failure of the angulator. The Taylor Spatial Frame (see below) has superseded these systems.

*Mechanical testing* of various fixator systems has shown that the monolateral fixator systems such as »Orthofix« or



the »Wagner apparatus« are much stiffer than the Ilizarov apparatus [31]. But it is precisely the low mechanical stiffness of the Ilizarov apparatus that promotes bone formation. If monolateral fixators are to be used, it is more appropriate to use those that can readily be dynamized, for example the »Monotube«. An excessively rigid system prevents new bone formation.

Our standard device for lengthening with complex corrections is the »Taylor Spatial Frame« [10] ([www.jcharlestaylor.com](http://www.jcharlestaylor.com); ■ Fig. 4.23). The basic system consists of two rings (or part rings) and six telescopic rods with



■ Fig. 4.22. 11-year old boy with femoral hypoplasia. For the *femoral lengthening* we use a monolateral fixator with a hinge (arrow), which allows continuous axial correction

special link joints. Corrections in six motion axes are produced by modifying the length of the telescopic rods. A computer program calculates the extent and rate of lengthening of the telescopic rods required to correct the deformity. The data are forwarded, via the internet, to a central control facility where the corrections are calculated. Three different types of strategies are possible:

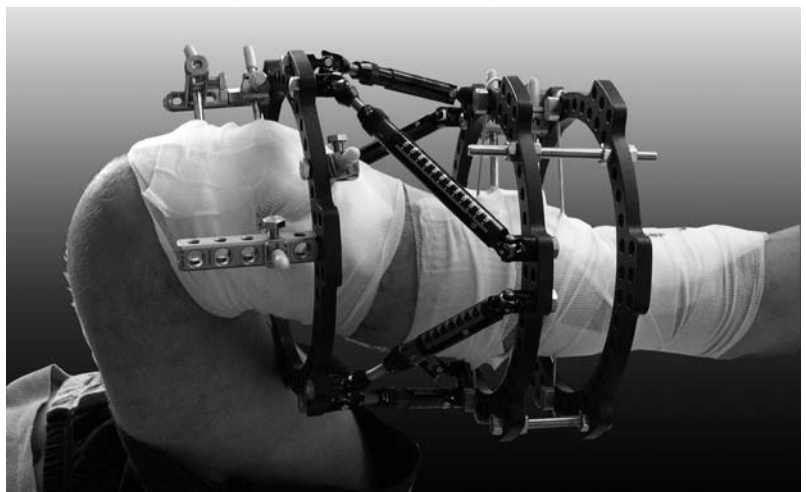
- The apparatus is fitted conventionally and then adjusted to produce the correction.
- The apparatus is fitted in the corrected position and gradually straightened during the continuous correction.
- Combined method (»crooked apparatus on crooked bone«). The pivot point of even very complex corrections can be precisely adjusted. This requires adequate imaging investigations and meticulous planning.

In order to shorten the application period of the fixator, and thus the associated handicap, we replace the frame after the desired length has been achieved and callus has started to form with a specially produced *Tomofix plate*. This plate has fixed-angle screws, does not lie completely flush against the periosteum (and does not therefore impair the circulation) and has a relatively high intrinsic elasticity (thereby promoting callus formation). It acts as a kind of »internal fixator«.

### Complications

Surgical leg lengthening and deformity correction are protracted, unpleasant treatments that are demanding for all involved and full of complications. Parents and child must be fully briefed accordingly before the start of treatment. Reputable studies have shown that every patient suffers one fairly major complication on average [7, 12, 20, 21, 32, 38]. Of course, not all complications are equally bad and some orthopaedists like to differentiate between »complications«, »problems« and »obstacles« [32]. We do not consider these euphemistic terms to be particularly useful and therefore

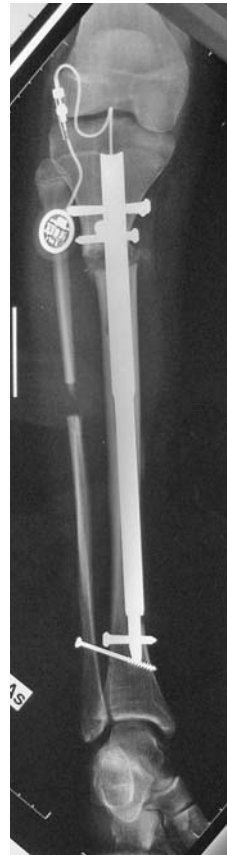
■ Fig. 4.23. Photograph of the *Taylor Spatial Frame* on the lower leg of a 15-year old boy with achondroplasia and varus deformity of the lower leg and excessively long fibula with shortening of the tibia



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talk of major and minor complications. Major complications are those that require an unscheduled operation, whereas minor complications can be solved without an operation. The principal *complications* are as follows:

- *Superficial infections* at the entry points of the Kirschner wires or screws. Such infections occur in almost all patients and can be treated by corresponding care, bathing and occasionally by antibiotic administration or a small incision. The entry points must not only be disinfected every day, it is also important that the children take regular *baths*. Soaking in water helps avoid crust formation and the retention of bacteria with consequent infection. Far from constituting an infection hazard, the bath water actually helps avoid infections. The nail entry points must also be adequately *cleaned mechanically*, since the disinfectant on its own is ineffective.
- *Deep infections*, i.e. infections that extend down to the bone can always be avoided with adequate care, and we have not observed cases of any such infection to date.
- *Movement restriction* in the adjacent joints: This can almost always be expected, particularly at knee level when the femur is being lengthened. If it occurs during lower leg lengthening there is a risk of an increasing equinus foot position. This tendency can be counteracted with physiotherapy, splints and bandages (■ Fig. 4.24). If neighboring joints are particularly at risk they must be included in the assembly (e.g. in femoral lengthening the knee must be bridged to guard against the possibility of femoral hypoplasia with aplasia of the cruciate ligaments).
- *Dislocations of the adjacent joints* : These are most serious complications of lengthening procedures. They occur especially if those joints are at risk, i.e. preexisting dysplasia of the hip, absence of cruciate ligaments in the knee joint or absence or dysplasia of the lateral malleolus in fibular deficiency. If these joints are particularly at risk they must be included in the assembly. This is usually only possible with ring fixator systems.
- *Angulations* are likewise common during the lengthening of longer segments and can be predicted on the basis of muscle forces. The angulation is always in the varus direction at the proximal femur and in the valgus direction at the distal femur and the tibia. Angulations can be avoided by the use of a ring fixator (e.g. the Taylor Spatial Frame). Otherwise corrections under anesthesia will be required, and these are always associated with a loss of length.
- *Pain* of varying degrees can be expected in all lengthening procedures. The pain can be strong particularly after approx. 10 days of lengthening, although they gradually subside thereafter and are tolerable during the consolidation phase.
- *Premature consolidation* occurs particularly if the leg has been inadequately lengthened for a certain period



■ Fig. 4.24. *Fitbone nail* in the tibia for leg-lengthening. This is a telescopic motorized medullary nail which is lengthened by remote control. A receiver is inserted under the skin at the proximal end of the tibia.

because of pain. Angulation corrections also involve a high risk of premature consolidation on the side of the pivot point of the angulation since, for geometrical reasons, the lengthening segment will be shorter here than on the other side.

- *Nerve lesions* – at least those with significant impact – are fortunately very rare provided the lengthening does not exceed 1 mm per day. Nevertheless, they can even occur at this lengthening rate [34].
- *Fractures* occur particularly after the removal of the external fixator. They can occur in the lengthened segment if the apparatus was removed prematurely. But fractures can also occur elsewhere on the lengthened leg as a result of osteoporosis, i.e. particularly if weight-bearing was inadequate while the external fixator was fitted. For this reason weight-bearing is extremely important.

Most of these complications are manageable and, with sufficient experience, avoidable in some cases. However, it is important that the parents and child realize what is in store for them so that they are not surprised when complications do occur.

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### 4.2.3 The limping child

The symmetrical gait is the most economical form of locomotion. Any asymmetry in the sequence of movements is indicative of a problem. Since walking is an extremely complex process, there are numerous ways in which the harmonious sequence of movements can be disrupted. Gait analysis is discussed in detail in ► chapter 2.1.3, as are the various types of limping. This chapter deals with the various ways in which gait is impaired and summarizes the differential diagnosis of limping in tabular form (► Table 4.10). Acute trauma has not been included, nor are tumors listed, as these can basically occur in any part of the body.

## 4.2.3 · The limping child

**Table 4.10.** Differential diagnosis of limping in children

Age group	Type of limp	Pain	Movement restriction	Tentative diagnosis	Investigations	Further details in chapter
Toddler (1–6 years)	Antalgic gait	Poss. hip, knee, ankle	Poss. hip, knee, ankle	Osteomyelitis, septic arthritis	Laboratory, x-rays, bone scan	3.2.11, 3.3.10
	Duchenne/Trendelenburg (»waddling gait«)	–	–	Hip dislocation	X-rays	3.2.4
	Antalgic gait	Poss. hip	Poss. hip	Transient synovitis of the hip	Poss. ultrasound	3.2.10
	Antalgic gait	Poss. hip	Poss. hip	Legg-Calvé-Perthes disease	X-rays (hips: AP and axial)	3.2.5
	Antalgic gait	Poss. hip, poss. knee	Poss. hip, poss. knee	Juvenile rheumatoid arthritis	X-rays, poss. ultrasound, laboratory	3.2.12, 3.3.11, 4.4.1
	Antalgic gait, clicking	Poss. knee	Poss. knee	Discoid meniscus	Poss. MRI, arthroscopy	3.3.6.6
	Paralytic gait	–	Poss. knee, poss. hip	Cerebral palsy, hemiparesis, myopathy, myelomeningocele	Neurological examination	3.2.8, 3.3.7, 3.4.10, 4.7.1
	Antalgic gait	Heel	–	Calcaneal apophysitis	–	3.4.9
Child (6–10 years)	Antalgic gait	Poss. hip	Poss. hip	Legg-Calvé-Perthes disease	X-rays (hips: AP and axial)	3.2.5
	Short-limb gait	–	–	Leg shortening	History (fracture?); Poss. investigate for hemihypertrophy syndrome	4.2.2, 4.6.1
	Antalgic/ Short-limb gait	Poss. knee, poss. hip	Poss. knee, poss. hip	Knee or hip contracture	History (trauma?); clinical findings, x-rays	3.2.15
	Antalgic/ Short-limb gait	Poss. knee, poss. hip	Poss. knee, poss. hip	Internal knee lesion	History (trauma?); clinical findings, x-rays	3.3.8
	Antalgic gait	Poss. hip, poss. knee	Poss. hip, poss. knee	Juvenile rheumatoid arthritis	X-rays, poss. ultrasound, laboratory	3.2.12, 3.3.11, 4.4.1
	Antalgic gait, pseudolocking	Knee	Poss. knee	Dislocated patella	AP, lateral and axial x-rays of the patella	3.3.5
	Paralytic gait	–	Poss. knee, poss. hip	CP, hemiparesis, myopathy	Neurological examination	3.2.8, 3.3.7, 3.4.10, 4.7.1
	Antalgic gait	Foot	–	Calcaneal apophysitis, Köhler's disease	X-rays	3.4.9
Adolescence (10–16 years)	Antalgic gait	Poss. hip, poss. knee	hip	Slipped capital femoral epiphysis	X-rays (hips: AP and axial)	3.2.6
	Antalgic gait	Poss. hip, poss. knee	Poss. hip, poss. knee	Juvenile rheumatoid arthritis	X-rays, poss. ultrasound, laboratory	3.2.12, 3.3.11, 4.4.1
	Antalgic/ Short-limb gait	Poss. knee, poss. hip	Poss. knee, poss. hip	Knee ligament or meniscus lesion	History (trauma?); clinical findings, x-rays	3.3.8
	Antalgic gait, pseudolocking	knee	Poss. knee	Dislocated patella	AP, lateral and axial x-rays of the patella	3.3.5
	Antalgic/ Short-limb gait	Poss. knee, poss. hip	Poss. knee, poss. hip	Knee or hip contracture	History (trauma?); clinical findings, x-rays	3.2.15, 3.3.13
	Antalgic gait	Foot	–	Sever's disease, Köhler's disease, stress fracture	X-rays	3.4.9

## 4.3 Infections

F. Hefti, G. Jundt

### 4.3.1 Osteomyelitis

#### ► Definition

Infection of the bone and bone marrow by bacteria

#### Classification

- **hematogenous osteomyelitis**
  - acute
    - unifocal
    - multifocal
    - special forms
      - neonatal osteomyelitis
      - spondylodiscitis
      - acute multifocal osteomyelitis
  - primary chronic
    - unifocal
    - sclerosing osteomyelitis of Garré
    - multifocal (chronic recurrent, CRMO)
  - specific (TB, BCG)
- **exogenous (secondary) osteomyelitis**
  - posttraumatic
  - postoperative

#### 4.3.1.1 Acute hematogenous osteomyelitis

##### **Etiology and pathology**

In this form of bone infection, which typically affects children and adolescents, bacteria reach the metaphysis of the long bones via the blood stream. The primary infection site usually remains unknown (nasal and oral cavities [22], skin, airways, gastrointestinal tract, urogenital system etc.). Because of the special flow conditions (slowed blood flow at the junction between the – narrow – afferent capillary loops and the – greatly dilated – efferent sinusoids before the epiphyseal plate), bacterial colonization is particularly likely to occur at this point. Moreover, vessel-associated phagocytes are lacking in these sections. This promotes bacterially induced endothelial damage with secondary thrombosis in the area of the otherwise extremely well-perfused metaphyses.

Since many bacteria possess receptors for bone matrix constituents such as collagen or bone sialoprotein, they can colonize on the bone (and also on implants) and then surround themselves with a polysaccharide-containing biofilm that makes them more resistant to attack by

antibiotics. From there, septic metastases can spread to other bones.

The focus may also remain locally restricted and lead to osteolysis or migrate across the cortex and produce a subperiosteal abscess. It can also break through into the neighboring joint and trigger a secondary septic arthritis. This occurs in infants and toddlers in approx. one third of cases [25]. Rather more plate-crossing vessels are present up until the age of 3 than in later life, and in most joints the capsule develops, at least partially, on the metaphyseal side, thus allowing infections to enter the joint directly via the periosteum.

A very wide variety of *bacteria* can trigger osteomyelitis. The typical pathogens vary according to the age of the patient. After the age of 3 or 4, staphylococcus aureus is involved in over 90% of cases, in addition to streptococci and staphylococcus epidermidis. All kinds of bacteria can be expected in younger children, e.g. enterococci, escherichia coli,  $\beta$ -hemolytic streptococci, haemophilus influenzae, anaerobes and fungi such as candida albicans [2, 7, 21, 23, 24, 32]. An increasing problem is infection with methicillin-resistant staphylococcus aureus [37].

#### **Occurrence**

The incidence varies considerable and seems to be on the decline in developed countries. A recent study from Scotland calculated an incidence of 2.9 new infections per year per 100,000 children [2], whereas the incidence a decade previously had been as high as 40 per year [7], and even then a distinct reduction compared to previous decades had been apparent. The condition affects all age groups with a frequency peak during early childhood. Boys are more often affected than girls. A recent study has shown an increased occurrence in Tennessee (USA) because of methicillin-resistant germs [37].

#### **Site**

Although acute hematogenous osteomyelitis can occur in any bone, it usually affects the metaphyses of long bones. These well-perfused areas of the long bones are typically affected, but the pelvis and spine are also particularly susceptible to the condition.

#### **Clinical features, diagnosis**

Even though the clinical features (especially in very small children) can be very variable, the symptoms are typical in most cases. The child is weak and ill, appears septic and has a high fever, although the latter can often be absent before the first birthday. The location of the pain can also pose problems since it is sometimes reported as abdominal or back pain in case of an osteomyelitis of the proximal femur. In all cases, however, the child looks ill. In older children, a clinical picture of minimal fever and fairly diffuse and unchar-



### 4.3.1 · Osteomyelitis

acteristic symptoms can likewise mislead the doctor in exceptional cases. The mobility of the adjacent joint is usually restricted. If the bone is only covered by a thin layer of soft tissue, painful swelling and inflammation may be present.

**! Any local swelling, inflammation, painfulness and restricted movement accompanied by fever should elicit the tentative diagnosis of acute hematogenous osteomyelitis. This diagnosis must continue to be maintained until it is either confirmed or ruled out.**

In order to treat any sepsis adequately, every effort should be made to identify the *triggering organism*. Bacterial screening starts (regardless of the febrile spikes) with three blood cultures taken at intervals of 30 min., which gives a 65% chance of isolating an organism. The prospects of a positive detection is increased still further if the painful site is screened for a subperiosteal abscess with *ultrasound*, followed by aspiration. An effusion of the nearest joint is also ruled out during the sonogram (particularly if e.g. the hip is affected).

*Blood cultures* and aspirates should be investigated for aerobes and anaerobes. Other *laboratory tests* include a differential blood count, C-reactive protein (CRP) and the erythrocyte sedimentation rate (ESR). The *differential blood count* and the leukocyte count are non-specific and do not show any typical changes. The *erythrocyte sedimentation reaction* is usually substantially elevated, but is a very slowly-progressing parameter. The far more sensitive and faster-progressing inflammatory parameter is the *C-reactive protein* [33]. In rare cases this can initially be normal, while the sedimentation rate is already greatly elevated.

*X-rays* are also used for monitoring the progress of the condition. The osteolytic focus itself does not provide any guide to the treatment to be initiated. The *bone scan* only has *diagnostic* significance if the test results for the local aspirates and blood cultures are negative and if the bone scan is the only way of confirming the diagnosis of acute hematogenous osteomyelitis. Since the osteomyelitis is typically located in the metaphysis, the interpretation can sometimes be difficult because of the physiologically increased uptake in the area of the growth plate. If surgical treatment is required, the bone scan should show whether any further foci requiring treatment are present. *Ultrasound* is very useful for detecting a subperiosteal abscess or an intraarticular effusion/septic arthritis. The *MRI scan* is a more sensitive diagnostic investigation than the x-ray. It is especially useful for detecting an abscess inside or outside the bone. A sequestrum can also sometimes be seen. In doubtful cases, however, a CT scan must be added, as this is more appropriate for visualizing sequestra.

### Treatment

The *antibiotic treatment* can only be effective if it is started before the formation of pus. Systemic antibiotics are unable to eliminate the bacteria either from necrotic bone or pus. Consequently, purely conservative treatment is indicated only in the early stages (i.e. before the formation of pus and sequestra). It can only be administered parenterally in an untargeted manner initially and should, wherever possible, be started only after blood cultures have been taken. The bacteriological cultures must be taken as an emergency measure in order to isolate an organism as soon as possible and prepare an antibiogram.

In children up to the age of 3 or 4, a normal sepsis treatment should be administered as a combination of aminopenicillin with clavulanic acid (=Augmentin) 220 mg/kgBW/24 hr i.v. and a cephalosporin (ceftazidime = Fortaz), 200 mg/kgBW/24 hr i.v., divided in each case into 4 single doses [26]. In older children a monotherapy targeted against staphylococci can be started as these are the bacteria most likely to be expected (e.g. aminopenicillin with clavulanic acid = Augmentin) 220 mg/kgBW/24 hr i.v., divided into 4 single doses). The dosage must be as high as possible (maximum dose: 8.8 g/day). If an organism has been found, the intravenous treatment should be continued with the highest-dose monotherapy. This does not apply to infants before their first birthday, who receive sepsis therapy from the outset. The CRP is checked on the 2nd day after the start of treatment. If the inflammatory parameters (fever, pain, CRP) have significantly regressed by this point, treatment is subsequently continued conservatively.

If this is not the case by the 5th day after the start of treatment, surgery is indicated. A bone scan is prepared preoperatively in order to establish any other additional foci.

A key requirement is the correct implementation of the *surgical treatment*, i.e. the bony abscess must be evacuated and all necrotic tissue (sequestra) must be consistently removed. This material must be examined both bacteriologically, for aerobic and anaerobic organisms, and histologically. Extensive irrigation is required at operation, although we do not insert a suction/irrigation drain. Nor do we consider the use of antibiotic-impregnated methyl methacrylate chains to be appropriate. If no further necrosis is present the acute osteomyelitis will heal of its own accord.

In acute hematogenous osteomyelitis surgery is always indicated as primary treatment (prior to the administration of antibiotics), if there is either

- an abscess (inside or outside the bone)
- a sequestrum or
- involvement of an adjacent joint

If adequate antibiotic treatment was initiated in the early stages of an osteomyelitis, the condition heals up in 80% of cases with this treatment alone. But if the bone infection has already reached a protracted stage, surgery is essential. But the only problem with this is the absence of prospective parameters for identifying a protracted stage, and which therefore has to be established on the basis of the patient's progress. Under no circumstances should one attempt to replace the operation with longer-term drug administration.

Orthopaedists have differing views about the *duration of antibiotic treatment*. A regimen of intravenous antibiotics followed by low-dose oral drugs used to be postulated. This can doubtless be explained in historical terms: On the one hand the erythrocyte sedimentation rate used to be the only inflammatory parameter available – but this is an awkward and slow parameter – and treatment was usually continued until it returned to normal. On the other hand, orthopaedists in the past often had to deal with protracted or chronic situations that could only be cured, or at least inactivated, by prolonged antibiotic treatment.

If the patient arrives for treatment *at an early stage*, the osteomyelitis is suspected and treatment initiated in good time, with subsequent healing and normalization of the inflammatory parameters (fever, pain, CRP). In this case, the parenteral antibiotic is administered until the CRP returns to normal, regardless of whether surgery was required or not. This normalization usually occurs between 5 and 14 days after the start of antibiotic administration and marks the actual completion of the treatment of the acute hematogenous osteomyelitis. Subsequent oral antibiotics are not required. After the CRP has returned to normal, the antibiotics are discontinued, the patient is discharged home and the CRP level is checked after a further 8 days. Most of the more recent studies support the principle of short-term antibiotic therapy [4, 14, 19]. If surgical clearance is required in the protracted stage, the same criteria apply.

If the *chronic stage* has already been reached, the inflammatory parameters (CRP and ESR) may have returned to normal according to both the clinical and laboratory criteria. In such cases, a combination of adequate surgery and a 6-week course of parenteral antibiotics can inactivate the condition such that no further recurrence, at least, need be expected in the subsequent years. While immobilization is unnecessary as a rule, and even rather counterproductive, a plaster splint may initially be useful for controlling severe pain. Treatment is basically functional, possibly with a passive motorized splint. The child is mobilized if at all possible (even if an infusion is in progress). If the lower extremities are affected the patient is mobilized on crutches without weight-bearing. As the pain subsides, weight-bearing can gradually be increased.

### **Follow-up controls**

After discharge, the CRP is rechecked on an outpatient basis 8 days later. If the CRP is still normal and the patient is free of symptoms and no other febrile episodes of uncertain origin have occurred, clinical follow-up checks at 3-monthly and subsequently 6-monthly intervals for up to 2 years after the onset of the illness will suffice. These are needed (particularly for the lower limbs) on the one hand to establish the consequences of stimulatory growth disorders and, on the other, to detect any recurrence in good time. If a bony osteolytic focus had been observed initially, the spontaneous filling of this focus should be confirmed after six months. If this has not occurred, it should be rechecked after a further 6 months.

### **Prognosis, postinfectious deformities**

In developed countries the mortality associated with this illness has declined over the past 40 years from 20% to almost 0% [7]. Defective healing, in the form of physeal damage with growth disorders, pseudarthroses and sequestrum formation, had been common before this time. Such residual deformities are rare nowadays, even in chronic cases of osteomyelitis (2% [7] to 3% [5]). In our own patient population we have not observed a single recurrence in the last 20 years after acute hematogenous osteomyelitis. The chronic stage of an acute hematogenous osteomyelitis with sequestrum formation and spreading to the whole shaft and surrounding tissues, represents a serious complication, not only because of local problems (instability, fracture risk, joint destruction), but also because a definitive cure is often almost impossible by this stage.

A stimulatory growth disorder can be expected after any infection in the growing skeleton. The consequences (including after trauma) depend on the age of the patient (► Chapter 4.1). Length alterations will invariably result, but these need not always be clinically significant (and even if they are, this will only apply to the lower extremities). If the premature partial or total closure of a plate occurs after chronic cases, the treatment of the defect should be based on the age and symptoms of the individual patient.

### **Special forms**

#### **Acute multifocal hematogenous osteomyelitis**

In this fortunately very rare form of the condition, acute infectious foci occur in several bones simultaneously as a result of hematogenous spread [28]. The pathogens involved are usually staphylococci. A temporary weakening of the immune system must be considered as an etiological factor as no actual immune defect has been detected in the described cases. The key symptoms are localized pain and tenderness at various sites and signs of general sepsis (fever, exhaustion, circulatory instability, elevated inflammatory parameters). The condition can occur in both small children and adolescents. A diagnostic bone

### 4.3.1 · Osteomyelitis

scan will help in identifying the inflammatory foci. MRI scans can be arranged to establish whether abscesses or sequestra have formed. Treatment consists of high-dose antibiotic administration. Foci with major accumulations of pus and/or necrosis must be surgically evacuated. Only very early and adequate treatment will be able to prevent defective healing in this form of osteomyelitis.

#### Neonatal osteomyelitis

The clinical findings in neonates differ markedly from those in older children, primarily because of the specific circulatory circumstances at the metaphyseal/epiphyseal level with much more plate-crossing vessels, the different pathogen spectrum and the immune system, which is still developing in the neonate. The situation is aggravated by the patient's inability to communicate. Fever and an elevated ESR are often absent in the neonate, which can lead to misinterpretations. Particular attention must therefore be paid to indirect signs: low mobility of the affected extremity with evasive movements («Pseudoparalysis»), sensitivity to touch, stretch position, diminished general condition, swelling and redness usually indicate that pus is already present in the joint. Because of the anatomical circumstances in the neonate, extensive destruction, penetration of pus into the joints with definitive damage to the growth plate and joint cartilage can quickly occur. The spectrum of the pathogens is much more variable in the neonate than in older children.

#### Spondylodiscitis

The diagnosis of purulent spondylitis or spondylodiscitis is often delayed. Abdominal signs and symptoms can be misinterpreted as appendicitis, and hip and thigh pain or difficulties in walking can also occur as predominant features. Localized pain on percussion and tenderness and an extended spinal posture are indicative of the condition. Further details on diagnosis and treatment can be found in ► chapter 3.1.13.

#### 4.3.1.2 (Primary) chronic osteomyelitis

##### ► Definition

Chronically progressing bacterial inflammation of the bone, which is not based on external causes and which does not appear to have undergone an acute stage.

##### Associated terms

- Brodie abscess (= metaphyseal intraosseous abscess formation without any preceding acute stage; ■ Fig. 3.333)
- Lymphoplasmacellular osteomyelitis (= classical primary chronic osteomyelitis)

- Chronic aggressive osteomyelitis/sclerosing osteomyelitis of Garré (see below)
- Chronic (recurrent) multifocal osteomyelitis (CRMO), SAPHO syndrome (see below)

#### Etiology

As with acute hematogenous osteomyelitis, the bacteria spread through the circulation to the metaphyses. But in the primary chronic form a different relationship appears to exist between the immune response and the pathogenicity of the organisms. This results in an initial local restriction of the inflammation, possibly with encapsulation of the focus without abscess formation and generalized symptoms of illness. The subsequent course of the illness appears to be characterized by immunological factors that are still not fully understood. On the one hand, the osteomyelitis can heal spontaneously without any type of late sequelae. On the other, it may lead to multifocal inflammatory metastases and a chronic course without any detectable pathogen. In fact, bacteria are detected in only around 30% of cases of chronic unifocal osteomyelitis, and their spectrum roughly corresponds to that of acute hematogenous osteomyelitis.

#### Clinical features, diagnosis

The clinical features of primary chronic osteomyelitis are highly variable and fairly non-specific. A »pseudo« or mini-trauma will often draw attention to the sub-threshold pain that had already been present. However, the onset is usually insidious, occurring over several weeks or even months. Except in cases with spontaneous healing, the symptoms usually increase steadily, but not dramatically, which often means that diagnostic investigations are initiated only at a late stage [21]. As with acute hematogenous osteomyelitis, the primary chronic form tends to be *located* in the metaphyses, particularly those in the upper ankles, followed by the metaphyses close to the knee.

The *diagnostic* process should not be restricted to laboratory tests and radiological investigations. Confirmation is obtained by a *biopsy*. Since a generalized reaction of the body to the illness is usually lacking, the diagnosis focuses primarily on local factors. If the patient is experiencing increasing musculoskeletal pain and/or swellings and no osteolytic foci are visible on the *x-ray* in either plane, symptomatic treatment of the pain is initially indicated. This consists of immobilization, possibly accompanied by the administration of anti-inflammatory drugs. If the symptoms continue to worsen during this treatment (for a maximum of 14 days), further check x-rays are indicated. If an osteolytic focus is discovered then a biopsy is indicated – as if a pathological change had already been visible initially.

A *bone scan* is required preoperatively to rule out any other foci that need to be biopsied. If there is the least suspicion of a malignant tumor on the basis of the radiographic findings, the usual tumor investigations with *CT*, *MRI* and subsequent *biopsy* should be arranged. The preoperative *laboratory tests* (differential blood count, ESR, C-reactive protein, immunoserology) supplement the diagnosis but are not specific. During the biopsy material should also be taken for *bacteriological investigation*, both in respect of aerobic and anaerobic organisms. In view of the low virulence of the pathogens involved, the bacteria are rarely detectable in culture. Sometimes the bacterial DNA can be detected by the PCR method (polymerase chain reaction).

Possible conditions to be considered in the *differential diagnosis* of osteolytic foci with and without an additional periosteal reaction include not only Langerhans cell histiocytosis, non-ossifying fibromas, enchondromas and unicameral bone cysts, but also malignant tumors (e.g. Ewing sarcoma or leukemia), which must be considered especially if there is erosion of the cortical bone and periosteal reactions.

### Treatment

**! Objective: To achieve a definitive cure for the disease as soon as possible without cosmetic or functional long-term sequelae. This requires adequate surgical treatment.**

The treatment of primary chronic osteomyelitis involves a *biopsy* combined with a radical *clearance* of the focus. The clearance may be performed in the same session only if the imaging investigations or exposure of the site rule out the presence of a possible malignant tumor. Otherwise the surgeon must await the result of the histological investigation. Any unnecessary contamination can have an adverse effect on the result of tumor treatment.

In all cases the biopsy material is forwarded for both histological and bacteriological investigation. Neither of these preparations may be fixed in formalin since this rules out the possibility of both molecular biological and other tests (e.g. tumor typing) as well as the bacteriological investigations. Consequently the material must be cooled (not frozen) and transported to the respective laboratories as soon as possible (i.e. within 30 min). An irrigation drain is unnecessary, as are locally or systemically administered antibiotics.

If no joint is affected, the site can be irrigated locally intraoperatively with an antiseptic agent (e.g. Lavasept). This agent has adverse effects on joint cartilage. If an organism is isolated, targeted postprimary intravenous, high-dose antibiotic treatment should be initiated. We administer this for 5 days, although it is not possible to establish the ideal period as the inflammatory parameters are usually normal from the outset and cannot therefore be used to monitor progression.

**! In acute hematogenous osteomyelitis the antibiotic treatment may be supplemented by an operation. In primary chronic osteomyelitis the reverse is the case.**

### Follow-up management

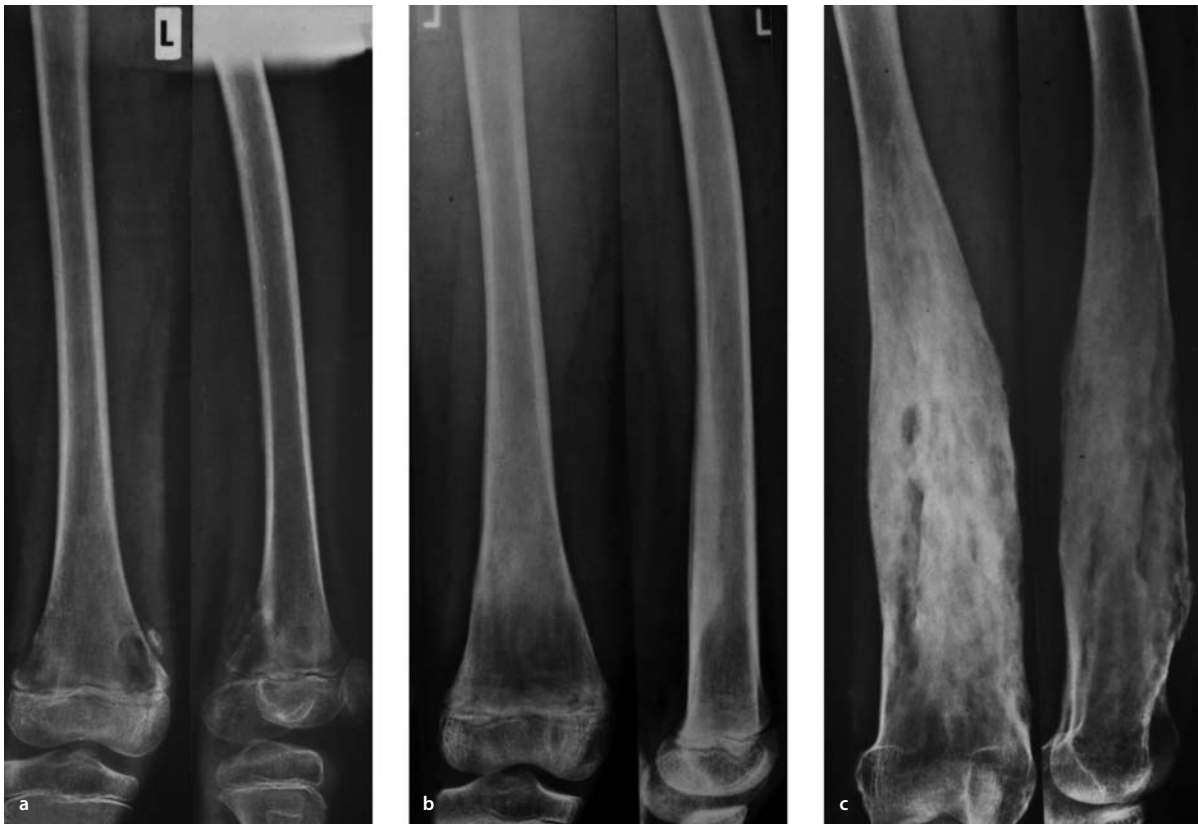
Every child will require functional follow-up management, possibly with a dynamic splint. If the lesion is located in the lower extremities, the patient is mobilized on crutches without weight-bearing on the affected side. Depending on the severity of the postoperative defect, gradually increasing weight-bearing can start within the first 5 or 8 days. If the necrotic tissue was not completely removed in the primary procedure, the bone defect will become superinfected, resulting in subsequent local irritation or abscess formation, which will then require immediate revision and radical resection of the necrosis (■ Fig. 4.25). Here, too, irrigation with Lavasept is appropriate and we do not use suction/irrigation drains or antibiotic-impregnated methyl methacrylate chains.

### Follow-up controls, postinfectious deformities

Subsequent clinical and radiological controls after 6–8 weeks are designed to ensure that the bone defect is not superinfected and gradually fills up spontaneously. If the focus has diminished in size, full weight-bearing may begin immediately depending in each case on the extent of the lesions. Further x-rays may then be required after a further 6–8 weeks, and possibly again after 6–12 months, in order to document the subsequent spontaneous closure of the defect. We do not consider follow-up bone scans to be necessary since the regeneration process can continue for a long time, which means that increased uptake may still be observed after 6–12 months and should not be confused with a recurrence. This would need to be confirmed with a leukocyte scan, although this is only indicated if the clinical findings point to a recurrence. If a large bone defect is involved and insufficient spontaneous reduction is apparent after 3 months, a cancellous bone graft may be indicated. If very large defects are present, homologous or autologous bone (cancellous bone from the iliac crest, non-vascularized fibula [30]) may be inserted.

However, if the symptoms recur and the defect continues to grow, the whole procedure will need to be repeated. If the bone defect is very large, the blood supply within the cavity may be insufficient, preventing the antibiotics from working properly. In such cases the insertion of a vascularized muscle flap (e.g. a gastrocnemius flap) may be indicated.

A *late check-up* two years after the onset of the illness serves to detect any possible *leg length discrepancies* and rule out other clinical deformities. The following *postinfectious deformities* can occur: diaphyseal deformation with inactive foci etc., stimulatory growth disorders with length alterations and direct partial or complete physeal damage (rare).



**Fig. 4.25a–c.** Course of *non-specific primary chronic osteomyelitis*. **a** 8-year old girl with spontaneous onset of pain in the left thigh near the knee but no fever. An x-ray was arranged only after 6 days of increasing pain (**a**). This shows a metaphyseal zone of lysis in the distal femur. Antibiotic treatment was initially prescribed. Since the symptoms did not regress, a (subtotal) surgical clearance was performed

after 2 weeks. No pathogen was found. The symptom-free patient was discharged after 4 weeks. **b** After 2 years the x-ray shows thickening of the distal femur. **c** After 13 years and multiple clearances the whole distal section of the femur shows clear changes indicative of chronic osteomyelitis. Although the patient is no longer experiencing any symptoms, a flare-up of the condition cannot be ruled out.

### Growth prognosis

The *growth prognosis* depends on the respective form of osteomyelitis involved. Growth disorders are usually characterized by stimulation, whereas inhibition is very rare. For more details on the consequences of growth disturbances see ► chapter 4.1.8, 4.2.1 and 4.2.2

### Special forms

#### Garré's primary sclerosing chronic osteomyelitis

In German-speaking countries the classification of primary chronic osteomyelitis is associated with the morphological findings of a plasmacellular, albuminous or sclerosing, non-suppurative osteomyelitis. All three terms are mentioned by Garré [9], who viewed them not as a separate clinical entity, but as »special forms and the sequelae of acute infectious osteomyelitis« – hence the title of his paper – and, as early as 1893, was discussing the bacterial virulence and the defensive situation of the body as a condition for the development of these clinical conditions. Some current authors mistakenly consider »sclerosing os-

teomyelitis of Garré« to be a separate clinical entity rather than just a special form of osteomyelitis.

The onset is insidious. The signs and symptoms are non-specific and tend to be progressive, but can be interspersed with asymptomatic phases. The affected sites correspond to the distribution pattern for acute osteomyelitis with a preference for the long bones. Excessive local warmth is typically, but not necessarily, present [35]. Even though it can always be assumed that bacteria trigger the disease, the relevant pathogens are rarely detected. The frequent failure to detect any bacteria in culture may be due to their low virulence. In some cases staphylococcus aureus or propionibacterium acnes may be isolated. Occasionally the PCR method (see above) will manage to detect the bacterial DNA. The x-ray shows swelling of the cortex, irregular structure and sclerosing of the medullary cavity. The periosteum appears elevated. The diagnosis is confirmed by an intralesional biopsy.

No standard treatment is available. In some cases, fenestration and opening of the medullary cavity will suffice,



and possibly the biopsy itself will serve as a stimulus to initiate regression. In other cases, the sclerotically altered bone sections will need to be completely resected in order to achieve a cure. An important requirement is to produce revascularization of the periosteum and medullary cavity. This can be achieved by various ways, e.g. by reaming the medullary cavity, by extensive longitudinal fenestration or with a vascularized muscle flap.

### Chronic (recurrent) multifocal osteomyelitis (CRMO)

This special form of chronic osteomyelitis involves the multifocal appearance of inflammatory foci almost invariably without the detection of any pathogens. Any bone can be affected. The metaphyses of the long bones are the preferred sites, but the vertebral bodies (vertebra plana), sternal ends of the clavicles, the iliosacral joint or the mandible can also be involved [4, 18]. Clinical examination reveals local pain without any major general symptoms or laboratory changes. Often the only abnormal laboratory finding is a (moderately) elevated ESR. The signs and symptoms can subside intermittently and then recur at the same site or a different location. Days, months, or even years can elapse between these episodes. The condition is occasionally accompanied by skin changes such as pustulosis palmaris et plantaris, psoriasis or acne, in association with, or even after, the manifestation of the bone pain. This clinical condition is also known as the SAPHO syndrome (Synovitis, Acne, Pustulosis, Hyperostosis, Osteomyelitis) [17].

A *bone scan* facilitates the detection of the individual foci. The diagnosis must be confirmed by a *biopsy*. The biopsy material should always be forwarded for bacteriological investigation, even though a pathogen is hardly ever detected [4]. The histological picture is completely non-specific and can range from an acute, granulocyte-rich inflammation, via chronic, lymphoplasmacellular infiltrates through to medullary cavity fibrosis and osteosclerosis with minimal inflammatory cells [16, 27]. The most important *differential diagnosis* is Langerhans cell histiocytosis (► Chapter 4.5.2). CRMO should also be distinguished from acute hematogenous multifocal osteomyelitis, which can occur acutely at several sites and trigger dramatic symptoms [28].

The *prognosis* for CRMO is good. *Treatment* consists of the administration of nonsteroidal analgesics. Antibiotics are not usually recommended as some authors believe that the condition is not triggered by bacteria [10]. In our view, however, it is not adequate to call the disease »non-bacterial osteomyelitis« [10]. Just because we are not able to cultivate the organisms with very low pathogenicity or to detect their DNA with PCR (polymerase chain reaction) does not mean that they are not the causative agents. In some cases (of exogenous osteomyelitis) we have detected bacterial activity by microcalorimetry where no other investigation was able to show the presence of germs. This method may prove helpful in the future in establishing the

true etiology of this mysterious disease. Surgical treatment is indicated only if the clinical symptoms intensify, if a fracture risk is present or if a joint is involved [3]. The lesions can heal spontaneously after several years [29].

### 4.3.1.3 Specific osteomyelitis (tuberculosis)

Infection with *Mycobacterium tuberculosis* has become rare in Central Europe since the beginning of the last century when it was still the commonest cause of death. In addition to the primarily affected site of the lung, bones and joints are typical *sites* affected by postprimary tuberculous foci. Children under 5 years are very typically affected in their *fingers* or metacarpals, with blistered swelling of the bone (*Spina ventosa*, from the Latin for a »thorn full of wind«). Another favored site is the spine [13, 34], where the tuberculosis can lead to collapsing of the vertebral bodies and pronounced gibbus formation (■ Fig. 3.118). An abscess can also form and spread along the psoas muscle and beneath the inguinal ligament. *Joint tuberculosis* typically affects one of the major joints and leads to pain, swelling, effusions, contractures and ultimately to the destruction of the joint.

A biopsy is always required to confirm the *diagnosis*. The *laboratory* findings are uncharacteristic, while direct bacteriological investigation does not identify the pathogen. *Histological examination* can reveal the tuberculous granulomas and often acid-fast bacilli as well. If the result for the direct preparation is negative, the result of culture testing must be awaited. Thanks to the introduction of new techniques, the time taken to obtain a definitive diagnosis and complete resistance testing has now been shortened to 2 to 3 weeks. More recently, the PCR method has been used successfully for pathogen detection [8].

! **The biggest diagnostic problem nowadays is that orthopaedists no longer think about the possibility of tuberculosis!**

The *treatment* consists of a specific antibiotic regimen involving a combination of rifampicin and isoniazid for 9–12 months. Tuberculous spondylitis must always be treated surgically with debridement of the affected vertebral body, if necessary backed up by an autologous bone graft.

### BCG osteomyelitis

Ever since BCG vaccination against tuberculosis was introduced, there have been occasional cases of chronic osteomyelitis caused by the attenuated pathogen in the vaccine. BCG osteomyelitis usually affects the metaphysis of the long bone nearest the vaccination site [12]. It starts within 4 years of the vaccination and can cause local pain, but no general signs of infection. The x-ray may be reminiscent of a tumor (■ Fig. 4.26). Histological investigation reveals tuberculous granulomas. After curettage the focus generally heals without any complications.



■ Fig. 4.26. 2-year old boy with BCG osteomyelitis (arrows) of the right humerus

#### 4.3.1.4 Posttraumatic and postoperative (exogenous) osteomyelitis (secondary osteomyelitis, exogenous osteomyelitis)

##### ➤ Definition

In contrast with the hematogenous types of osteomyelitis, the infection in the exogenous forms is caused directly, i.e. by external wounds on the bone.

##### Etiology and occurrence

Exogenous osteomyelitis is caused by serious open injuries, penetrating, inadequately treated, contaminated skin wounds, purulent bursitis or operations. The bacteria involved are usually *staphylococci*, although infections with any other organisms can occur. Because of their good infection defense systems children and adolescents rarely contract exogenous osteomyelitis. Even though a postoperative infection rate of 1–2% can be expected generally, osteomyelitis develops in only a fraction of cases. Deep infections can be expected in 3% of patients with open fractures. After trauma however, the risk of severe infection complications in children is much lower compared to adolescents and adults [15].

##### Clinical features, diagnosis and treatment

Clinical signs indicate the presence of an infection at an early stage. Elevated temperatures during the first 2 days following the operation or trauma are still not suspicious. However, if the fever intensifies on the 3rd day or later (possibly combined with increasing wound pain or redness), the presence of a local infection must be assumed until the opposite is proven.

Under no circumstances should the diagnosis be masked by the postoperative administration of antibiotics. The *preventive administration of antibiotics* before surgery is indicated in the following situations:

- third-degree open fractures,
- orthopaedic operations involving metal implants (bacterial traps),
- all operations lasting longer than 2 hours.

The antibiotic (a 1st generation cephalosporin, e.g. cefazolin = Keflex 1 g i.v.) is administered immediately before operation. If necessary, a repeat dose can be given after 4 hours if the operation is protracted.

! No »preventive antibiotic treatment« post-operatively.

The best *prevention* is adequate surgical treatment. This involves comprehensive debridement in severe crush injuries and open fractures and a consistent atraumatic operating technique. If the *fever* intensifies on the 3rd postoperative day, or later, we investigate the laboratory parameters. The CRP and ESR are always elevated at this time because of the operation, but a further subsequent increase confirms the infection. The clinical signs (pain, inflammation) are often sufficient to indicate the local infection. Bacteriological samples are taken from a wound swab. A *sonogram* may show a fluid accumulation, in which case *surgical revision* is always indicated. If a germ is found, appropriate *antibiotic treatment* based on the resistance-testing is administered until the infection parameters have returned to normal. If no germ is detected (as commonly happens with nosocomial infections with *Staphylococcus epidermidis* or other slow-growing bacteria), non-selective treatment is initiated with broad-spectrum antibiotics at the maximum dosage (e.g. a broad-spectrum penicillin and an aminoglycoside).

*Chronic infections associated with implants* that may not be removed (e.g. tumor prostheses), can often be successfully treated with a combination of quinolones (e.g. ciprofloxacin = Ciproxin or levofloxacin = Tavanic) and rifampicin (= Rimactane), i.e. either the infection is cured or it at least remains under control and does not spread [38]. This treatment often has to be continued for years. The purpose of the rifampicin is to prevent the formation of resistance to the quinolone.

### 4.3.2 Septic (suppurative) arthritis

#### ➤ Definition

Bacterial infection of a joint either directly or via perforation of an osteomyelitic focus.

#### Etiology, site

The etiology is almost identical to that for acute hematogenous osteomyelitis. As a result of bacteremia organisms are either transported directly to the synovial membrane or enter the joint via the metaphysis. Osteomyelitic foci located near the joint can also start producing an effusion (initially without bacteria), and cause a secondary infection of the joint at a later date via penetration or perforation. The circulation in the epiphyses differs before and after the age of three. Up until this time, the epiphyses are supplied by plate-crossing vessels. Subsequently, however, the epiphysis and metaphysis are supplied by their own vascular systems that are largely independent of each other. Consequently, metaphyseal infections in children up three years of age can more readily enter the joint via the transepiphyseal vessels than is the case in older children.

The *bacterial distribution* roughly matches that of osteomyelitis with the same pattern of age-dependency. In an investigation of 58 cases, staphylococcus aureus was found in 43% of cases, coagulase-negative streptococci in 10%, streptococcus pneumoniae and salmonellae in 5%, and haemophilus influenzae and group B streptococci in 3% [36]. Although any joint can be affected in principle, the major joints of the lower extremities are involved in 90% of cases (the hip in over 50 percent of cases, knee and ankle) [36].

#### Growth prognosis

An infection that migrates through the growth plates can subsequently lead to infection-induced epiphyseal separations and the possibility of physeal damage. Furthermore, any infection located in a joint can, in the long term, lead to direct or indirect, irreversible damage to the cartilage. In severe cases, avascular necrosis of the femoral head can occur. A certain remodeling of the joint may take place, with reconstruction of the cartilage with hyaline and fibrous replacement cartilage, although this process can never be accurately predicted in the individual case. The residual mobility after the acute phase has subsided plays a key role in the regeneration.

❗ **In any joint infection that persists for longer than 4 days [1], the possibility of direct joint damage and growth disorders with corresponding consequences for the physeal area must be borne in mind.**

Scarred structures can cause additional problems and, particularly in the hip, possibly resulting in increased decentering and secondary dislocation.

#### Clinical features

In infants, the septic arthritis can start without any fever, although the child looks ill and has a septic appearance. Fever is more common as a key sign in older children, in addition to the joint effusion, but is not necessarily present. Roughly 10–15% of all cases of septic arthritis start without any fever. The clinical picture is primarily characterized by the spontaneous onset of painful movement restriction of the affected joint. If the lower limbs are involved, a spontaneous limp is a sign of joint impairment. The history can sometimes be misleading as the spontaneity of the onset of the illness is overlaid with genuine or apparent trauma. The signs and symptoms always intensify over time. Confirmation of the diagnosis is particularly difficult in infants. The child no longer moves the affected extremity spontaneously and resists attempts by the parents to move the limb.

#### Diagnosis and treatment

❗ **Bacteria and pus in the joint must be removed mechanically as drug treatment on its own is inadequate in a case of suppurative arthritis.**

In all feverish patients with swelling and pain in the vicinity of a joint, the effusion must be located and *aspirated as an emergency procedure*. The aspiration serves as both a diagnostic and therapeutic measure. In most cases an effusion can be diagnosed clinically if it occurs in the knee, ankle or elbow. In doubtful cases – as invariable applies in the hip – the effusion will need to be diagnosed with the aid of *ultrasound*. Only when the effusion has been diagnosed and the preparations have been made for aspiration under general anesthesia are all the other investigations that also apply in a suspected case of acute hematogenous osteomyelitis arranged: *Laboratory*: Three blood cultures at intervals of 30 minutes, ESR, C-reactive protein, differential blood count. An *x-ray* is arranged to rule out any osteolytic foci near the joint. Only if the aspirated fluid is clear does the surgeon await the results of bacteriology testing before initiating further treatment.

However, if the aspirate is cloudy, or even purulent, the actual local *treatment* is initiated in the same anesthetic session, i.e. *arthroscopic lavage* [6]. The aspirated pus is investigated for anaerobic and aerobic organisms. We no longer perform an open arthrotomy and insert an irrigation drain, since this only irrigates a track inside the joint rather than the whole of the joint. During arthroscopic irrigation, the joint is irrigated liberally throughout – depending on the size of the joint – with 500–1000 ml of fluid. Only if the arthroscopic assessment of the joint reveals severe destruction of the cartilage, with cartilage residues and fragments floating freely in the joint, and these cannot be removed arthroscopically, do we consider arthrotomy with extensive joint debridement to be indicated. The same is true for cases with concomitant osteo-

### 4.3.2 · Septic (suppurative) arthritis

myelitis of the adjacent bone. However, we never delay the closure of the wound [11].

This local treatment is accompanied by *intravenous antibiotic therapy*, either with targeted drugs after resistance testing or in an untargeted manner with a combination of two antibiotics that cover a broad spectrum. On the second day after the start of treatment, the patient initially remains fasted, the CRP is repeated and the situation is clinically re-assessed and, if necessary, a sonogram arranged. If joint mobility continues to be significantly restricted – and if a residual or recurrent effusion is confirmed clinically or by ultrasound – the *arthroscopic lavage* under anesthesia is repeated. In stubborn cases this process should be repeated after a further two days until joint mobility has returned to normal, pain is no longer present and the CRP has also returned to normal levels. This is usually the case after 14–20 days. Normalization of the inflammatory parameters – as with the treatment of acute hematogenous osteomyelitis – then signifies the conclusion of the antibiotic treatment.

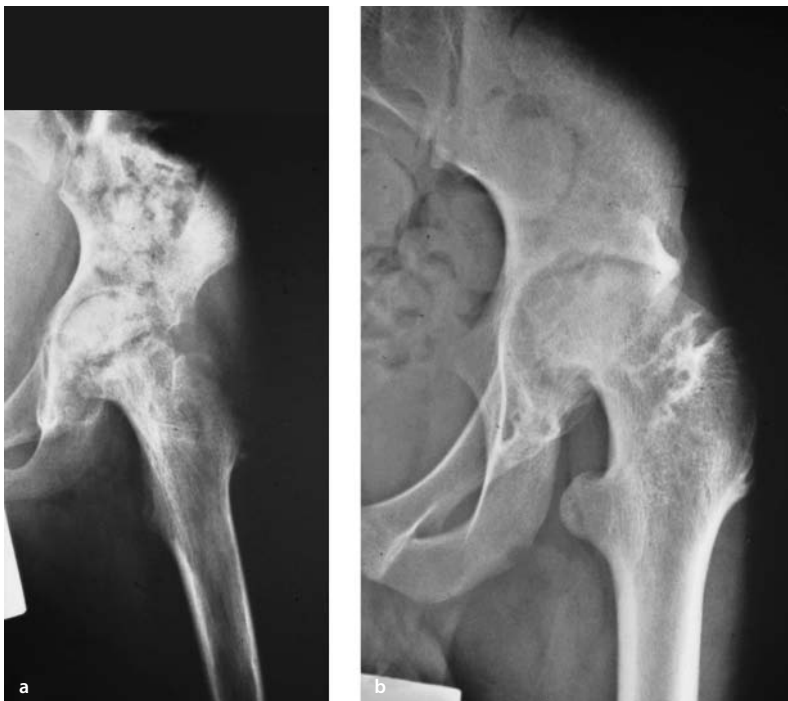
#### **Follow-up management, follow-up controls**

Follow-up management is essentially functional, ideally with the aid of a dynamic splint. Otherwise, the spontaneous mobility of the patient should be assisted passively by the physiotherapist with adequate analgesia. The patient is mobilized in the immediate postoperative period while the infusion is still in situ. Weight-bearing is permitted according to the level of pain. A further CRP check is arranged on an outpatient basis eight days after

the discontinuation of the antibiotic treatment. If the result is normal, if the child is subjectively free of symptoms and the affected joint is freely mobile, another functional check-up is arranged after a further 4–6 weeks. If, at this second check-up, joint mobility continues to remain normal and no other symptoms are present, the patient may resume sports activities. Subsequent clinical controls at 3- or 6-monthly intervals for two years serve, on the one hand, to document the continuing free mobility of the joint and, on the other, to rule out any incipient growth disorders. If the patient is free of symptoms at the end of this period, the treatment can be considered as concluded.

#### **Postinfectious deformities**

Postinfectious deformities usually pose complex and difficult therapeutic problems. The widespread destruction of a joint is often a tragedy for a child. But even if very severe contractures are present, stiffening of a joint should not be accepted too soon in a child. With aggressive, consistent and long-term mobilization and exercise therapy, it is often possible to restore function in substantially destroyed joints thanks to the considerable remodeling potential possessed by children. This process will require multiple hydraulic mobilization procedures under anesthesia, intensive postoperative exercise therapy under epidural anesthesia, followed by more long-term physical therapy (■ Fig. 4.27). If avascular necrosis of the epiphysis has occurred, insertion of a vascularized autologous bone graft can be helpful [31].



■ Fig. 4.27a. b. Situation after *suppurative coxitis* in a 10-year old boy. a Complete destruction of the joint cartilage and incipient avascular necrosis of the femoral head. b At the age of 16 years, after several years of intensive exercise therapy, there is moderately pronounced deformation of the femoral head, but a joint space of largely normal width. In clinical respects there was normal mobility (only the rotation and abduction were restricted), and the patient is now free of symptoms



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## 4.4 Juvenile rheumatoid arthritis

» *When a rheumatic child comes for a medical check, always think of the eyes and neck...* «

### Definition

Juvenile rheumatoid arthritis is an inflammatory condition that occurs during childhood or adolescence and affects one or more joints, although it can also affect other organ systems (particularly the eyes). It tends to affect the major joints rather than the smaller joints of the hands and feet as with the primary chronic adult form. Atlantoaxial subluxation can be a concomitant problem. The course of the disease is very variable and the prognosis is good (particularly if only a small number of joints are involved) in 80% of cases.

— Synonym: Still's disease

### Historical background, occurrence

The systemic form of the disease was described by G. F. Still in 1897 [20]. The incidence of juvenile rheumatoid arthritis is approx. 3–5 new cases per 100,000 children under 15 years of age [1, 13]. A major study on chronic juvenile rheumatoid arthritis in Germany calculated an incidence of 6.6 and a prevalence of 14.8 per 100,000 individuals under 16 years of age [22]. Substantial geographical differences exist, with the illness occurring more frequently in northern countries. The male to female ratio is 1:2.5.

### Etiology

Immunological, genetic, climatic, infectious and psychological factors have been discussed as *etiological factors*. Some children with juvenile rheumatoid arthritis, particularly the severe forms, show *anomalies of the immune system*, e.g. antinuclear antibodies or hypogammaglobulinemia. Autoantibodies, abnormal antigen-antibody complexes and other anomalies detectable in the laboratory also occur [11]. *Genetic components* likewise play an important role. So although there is a North-South differential in the frequency of the disease (which is associated with climate), the condition is also widespread in those hot countries with a predominance of Anglo-Saxons (New Zealand, Australia). The illness is particularly common in the UK with its damp, cold climate. *Microorganisms* such as Chlamydia trachomatis, Yersinia enterocolitica and Mycoplasma fermentans have also been discussed as the possible cause of juvenile rheumatoid arthritis. A bacteria-specific, synovial cellular immune response has been observed [14, 18], although these findings have not yet been corroborated. *Psychological factors* also appear to play a role in the manifestation of the disease, as children have often been reported as being in a particularly stress-

ful situation prior to its onset. Children with juvenile rheumatoid arthritis also tend to be rather reserved and seem to have difficulty in expressing their problems and conflicts.

### Pathogenesis

The illness starts with a synovitis. The joint mucosa becomes edematous and hypervascularized, and an effusion that is moderately rich in leukocytes (particularly lymphocytes) forms. Over time the synovial cells proliferate, causing the synovial membrane to thicken and form nodules and protuberances and, in some cases, cysts. At a later stage fibrinoid degeneration occurs with granulomatous changes of the hypertrophied synovial membrane. As the condition progresses, the subchondral bone starts to erode at the margins and the cartilage is damaged. Pannus spreads from the edge of the cartilage across its surface and destroys the hyaline cartilage. Cysts form in the subchondral bone. The whole joint becomes hypervascularized. A similar sequence of events can also unfold in the area of the tendon sheaths. Subcutaneous rheumatic nodules also occur occasionally.

### Classification

Juvenile rheumatoid arthritis can occur in the following forms:

#### Classification of juvenile rheumatoid arthritis

- A. Juvenile chronic arthritis
  1. *Systemic form* (Still's disease)
  2. *Polyarticular form*: more than four joints affected, antinuclear factors in 40 %, asymptomatic iridocyclitis
  3. *Oligo- (pauci-)articular forms*
    - *Type I*: Commonest form, often antinuclear factors, often iridocyclitis
    - *Type II*: HLA B 27-associated form, predominantly in boys, later possibly ankylosing spondylitis (Bechterew disease) (► Chapter 3.1.12.3)
- B. Rheumatoid factor-positive juvenile rheumatoid arthritis: Initial manifestation after the age of 10, small joints affected as in chronic rheumatoid arthritis in adults.
- C. Juvenile psoriasis-associated arthritis.

### Clinical features

The *systemic form* of the illness (approx. 30 % of cases) starts with fever, rash and polyarticular arthritis. The heart, liver, spleen and lymph nodes may be affected. Finally, iridocyclitis also develops. Laboratory findings include anemia, leukocytosis and an elevated ESR. Antinuclear factor and rheumatoid factors are usually nega-

tive. The *prognosis* for this form is poor, with severe joint destruction occurring in 40 % of cases, and even death in 1–2 %. In the *polyarticular form* five or more joints are affected. The disease can start acutely or insidiously with symmetrical arthritis, and the upper and lower limbs can be affected. These patients have usually already reached adolescence, i.e. are older than 10, by the onset of the disease. The serum rheumatoid factor is negative in one subgroup and positive in another. The *prognosis* is more favorable in those with a negative serum rheumatoid factor. The polyarticular form of the disease lasts longer than the oligoarticular form and the associated physical handicap is greater and the prognosis worse. *Only approx. 25 % of cases are cured.* The *oligoarticular form* is the most common (approx. 45 % of cases) and, by definition, fewer than five joints are affected. *Type I:* The mildest form only involves arthritis with no eye involvement, and the test for HLA B 27 is negative. Iridocyclitis is often observed in the more severe form and, in 10 % of cases, can lead to blindness. This form has the best *prognosis* for a cure. *Type II:* In this group the test for HLA B 27 is positive. This form can progress to *ankylosing spondylitis* and the *prognosis* is less favorable. Abnormal radiological findings in the iliosacral joints only appear after the age of 20. Patients with the oligoarticular form are usually younger than 10 years when the symptoms first appear. Overall, a remission can be expected in the oligoarticular form in 60 % of cases. In *rheumatoid factor-positive juvenile rheumatoid arthritis* girls are more frequently affected than boys, the initial joint symptoms occur in the hands and feet, the IgM rheumatoid factor is positive, and the disease often progresses to the adult form of *rheumatoid arthritis*. *Juvenile psoriasis-associated arthritis* sometimes manifests itself initially in the joints before the skin changes occur, which can aggravate the diagnosis. The course of the disease in children is similar to that in adults.

The *key symptom* in juvenile rheumatoid arthritis is the inflammatory synovitis. The patients complain of moderate pain or, rarely, intense pain. Morning stiffness or stiffness after inactivity is frequently reported. The affected joints swell up and shows signs of inflammation. Chronic joint effusions appear at a later stage. In juvenile rheumatoid arthritis it is the joint stiffness that predominates, in contrast with rheumatoid arthritis in adults, where instability is the main problem. The systemic form is also accompanied by fever and exanthema. The macular rash is salmon-colored and affects the central parts of the trunk, but rapidly disappears again.

**! Ocular changes occur in 20 % of cases in the oligoarticular form, but tends to be fairly rare in the systemic or polyarticular forms.**

*Amyloidosis* can occur if there is severe systemic involvement which, though rare, constitutes the commonest

cause of death. The course of the disease is characterized by phases of deterioration and remission.

### Radiographic findings

The soft tissue swelling, in particular, is observed in the early stages, shortly followed by periarticular osteoporosis. These are non-specific changes. As the disease progresses, the affected joint space appears narrowed as a result of destruction of the hyaline cartilage. In contrast with mechanically induced *osteoarthritis* there is no subchondral sclerosis. On the contrary, the bone is less radiopaque as a result of the osteoporosis (■ Fig. 3.336). Only a relatively slight osteophytic reaction takes place. The osteoarthritis of the *hip* (■ Fig. 3.243) is concentric and associated with acetabular protrusion [8, 9]. By contrast, arthritic cysts are observed in the epiphyses. As the disease progresses, a fibrous or bony ankylosis develops. At the *spinal* level, changes are primarily apparent in the cervical spine with, in particular, erosion of the dens with subsequent atlantoaxial subluxation. *Additional imaging investigations* are rarely useful. Bone scans, CT scans and MRI scans do not provide any additional information that would have therapeutic consequences. A sonogram may be appropriate for detecting any joint effusion, particularly in the hip.

### Differential diagnosis

Joint effusions occur in a range of diseases, e.g. *hemophilia* or *suppurative arthritis*, but also in other rare conditions such as *enthesopathic arthritis*, *leukemia*, *systemic lupus erythematoses* and *rheumatic fever*. A traumatic cause must also be ruled out. As tumor-like lesions, *pigmented villonodular synovitis* and *synovial chondromatosis* can also produce chronic effusions.

### Prognosis, complications

Since, as already mentioned, *ocular complications* can occur, particularly in the oligoarticular form [17], children with juvenile rheumatoid arthritis must always be referred to an *ophthalmologist*. Growth disorders not infrequently occur as a result of the disease [8], locally in the form of growth stimulation or, more commonly, as premature physal closure. However, systematic retarded growth can also occur, particularly as a result of corticosteroid treatment and loss of appetite [23]. Another important complication is the *involvement of the cervical spine* in 60 % of cases [9]. Although very few patients develop an atlantoaxial subluxation with neurological problems (hyperreflexia and clonus), particular attention should be paid to this problem. *The cervical spine must always be x-rayed before any intubation.* Another serious complication is the formation of *avascular necrosis of the femoral head* [14, 18]. Although rare, this can lead to a dramatic deterioration in hip function.

### Treatment

The proper treatment of these often severely handicapped children requires the collaboration of various specialists: pediatricians (possibly including pediatric rheumatologists), pediatric orthopaedists, physiotherapists, social workers and ophthalmologists.

### Conservative treatment

*Non-steroidal anti-inflammatory drugs* are the most important therapeutic agents. Aspirin is very effective at a dosage of 60–100 mg/kg BW/day. Since this involves very high doses, naproxen can be more pleasant to administer, since it only requires a dosage of 10–15 mg/kg BW/day. Possible side effects include abdominal symptoms or tinnitus. Gastric or intestinal ulcers are extremely rare in children. The treatment must continue for several months even if a remission occurs after a short time. We generally administer this treatment for three months initially. If a remission occurs during this time we try withdrawing the treatment. If the disease recurs, the treatment must be resumed. More severe forms must be treated with *corticosteroids*. These are used particularly if pericarditis, myocarditis or iritis are present or if severe joint changes fail to respond to other drugs. In the most serious cases, *cytostatic agents* (methotrexate) are administered. The conservative treatment is always accompanied by *physiotherapy*, which is primarily aimed at preventing joint contractures. *Splints* can sometimes be used to extend contracted joints.

### Surgical treatment

The following surgical options are available:

- (Arthroscopic) joint lavage with (hydraulic) mobilization under general or epidural anesthesia,
- Synovectomy,
- Lengthening of contracted muscles and tendons («soft tissue release»),
- Joint extension with an external ring fixator (Ilizarov apparatus),
- Arthrodesis,
- Joint prostheses.

*Arthroscopic lavage* and *hydraulic mobilization* of a joint can be very useful in juvenile rheumatoid arthritis. It can occasionally improve joint mobility and the flushing out of the cartilage breakdown products and inflammatory cells can also improve joint symptoms for a prolonged period. On the other hand, this operation does not produce any long-term effect and thus does not improve the prognosis for a joint. Nor has the procedure of *synovectomy* lived up to expectations in juvenile rheumatic arthritis [7, 14], since it cannot halt the deterioration in joint function and the pain relief is usually only temporary [13].

*Soft tissue release* can be very useful, particularly if there are severe flexion contractures in the hip. Lengthen-

ing of the hip flexors can produce a permanent improvement in joint function. Although some of the correction may subsequently be lost in the initial stages, part of the correction can generally be preserved in the long term [12, 22].

The most successful method for knee contractures has proved to be continuous *extension with the Ilizarov apparatus* or the *Taylor Spatial Frame* [4]. We have gained some experience with this technique in our own hospital (■ Fig. 3.347 and 3.349). In a case of severe flexion contracture, the knee can be straightened with the Ilizarov apparatus within a few weeks. Although this does not improve joint mobility, the extended position is much better in functional respects than a flexion position, as the amount of energy expended when walking with flexed knees is very substantial. The drawbacks of the extended position with restricted flexion become apparent when sitting in cramped conditions or when difficulties are encountered during cycling.

*Arthrodesis* is rarely performed these days and is only useful in ankle if severe joint destruction is present [3, 6]. Otherwise, prostheses tend to be inserted in all the other major joints. *Joint prostheses* are used worldwide, and very successfully, for idiopathic arthritis of the knee or hip, particularly in patients with degenerative arthritis aged 60 years and over. Whereas orthopaedists used to be very cautious about implanting artificial joints in younger patients, they now insert them for a wider range of conditions (■ Fig. 3.244).

In previous years patients with badly destroyed joints had a greatly reduced quality of life. The implantation of artificial joints gives them freedom from pain and improved mobility. Although experience over very long periods is still lacking, »long-term results« are available to date for follow-up periods of 10, and in some cases 20 years. But if a prosthesis is implanted in someone under 20 years of age, then one has to think about periods of 50 years and more. What is certain is that a prosthesis implanted now will not last for 50 years. On the other hand, it is no longer acceptable to let a young patient continue to suffer with stiff joints only to finally insert a prosthesis at the age of 60. Loosened prostheses can be replaced, and if no further replacement is possible after multiple revisions, an arthrodesis can still be performed or, if the hip is involved, a Girdlestone procedure. The outcome would still be acceptable to anyone who is reliant on a wheelchair.

We therefore consider that a total knee or hip replacement is indicated for patients with severe joint destruction even if they are under 20 years old. Substantial corresponding experience has been accumulated worldwide for patients with juvenile rheumatoid arthritis [1, 2, 10, 15, 16, 21]. The loosening rates for the prostheses are higher than in older patients with degenerative arthritis, but slightly better than those for other young patient

populations. This finding is attributable to the physical activity of the patients: Young rheumatic patients are more active than old ones, but less mobile than healthy young people. A 10-year survival rate for the prostheses of approx. 80 % can currently be expected, with no major differences between hip and knee implants. Problems affecting the *cervical spine* may occasionally require surgical intervention. In particular, major instability will need to be secured by atlantoaxial screw fixation [5] (► Chapter 3.1.13).

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## 4.5 Tumors

### 4.5.1 Basic aspects of tumor diagnosis

F. Hefti, G. Jundt

#### Definition

A tumor is a proliferation of autonomously growing cells. While tumor-like lesions may resemble tumors in clinical and radiological respects, no autonomous cell proliferation takes place. *Benign tumors* grow autonomously, but their cells are not atypical, nor do they infiltrate or metastasize. Individual tumors behave *aggressively at the local level* and tend (particularly after inadequate treatment) to recur. These were formerly known as »semi-malignant tumors« and are currently described by the World Health Organization as »intermediate«. This term also covers those tumors that very rarely metastasize at all, but whose biological behavior cannot be derived from their histological picture, e.g. the giant cell tumor of bone or infantile fibrosarcoma.

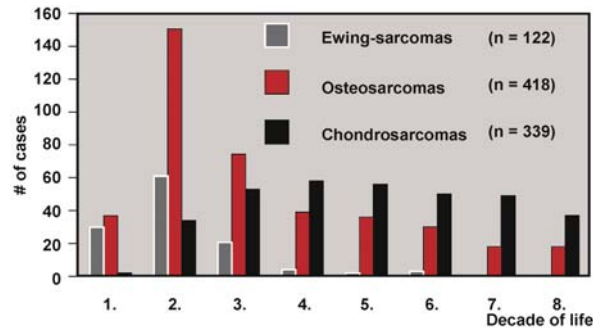
*Malignant tumors* are subdivided into low-grade and high-grade tumors. Low-grade malignant tumors tend to grow slowly and metastasize at a late stage, whereas high-grade malignant tumors grow rapidly, their cells show little differentiation and are highly polymorphic. Their growth is invasive, destructive and infiltrative. Metastasis occurs at an early stage. Typical low-grade malignant tumors are the classical chondrosarcoma and the parosteal osteosarcoma. High-grade malignant tumors are the conventional osteosarcoma and Ewing sarcoma.

#### 4.5.1.1 Clinical considerations

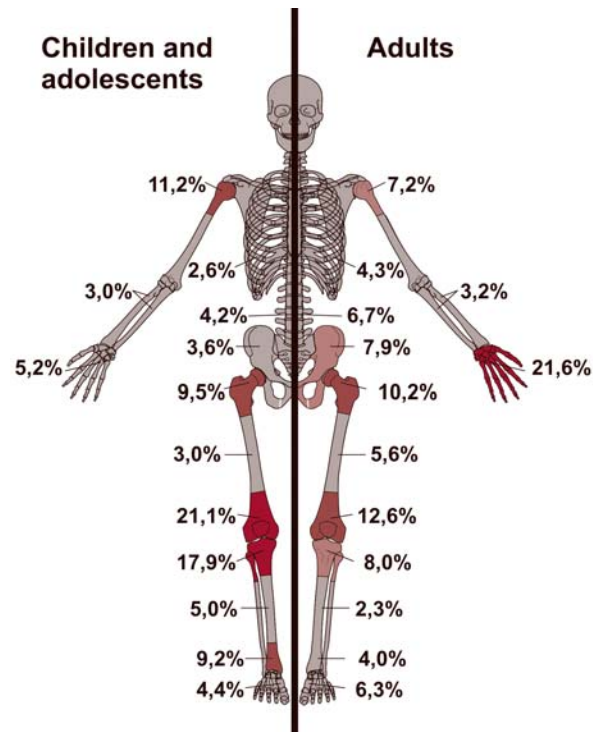
*Bone tumors* are rare. Their malignant variants represent only approx. 1% of all malignant lesions. But it is precisely the high-grade malignant types that typically occur in children and adolescents (■ Fig. 4.28). A general practitioner will encounter one such tumor in one patient in every 10,000 or so [3], while the frequency will be slightly higher for a pediatrician or general orthopaedist. Nevertheless, a bone tumor remains a rare event.

❗ **It is precisely because malignant bone tumors are so rare that the consulted doctor often fails to consider the possibility of such a diagnosis. Thus, it takes around nine weeks, on average, to diagnose an osteosarcoma, four months for a Ewing sarcoma and five months for a chondrosarcoma [16].**

The history duration depends greatly on the rate of tumor growth and the tumor site (■ Fig. 4.29). The more the tumor is covered by soft tissues, the longer it takes to reach a



■ Fig. 4.28. Age distribution for 879 primary solid malignant bone tumors of the postcranial skeleton registered by the Basel Bone Tumor Reference Center. The Ewing sarcoma and osteosarcoma, in particular, typically occur during the first two decades of life



■ Fig. 4.29. Site of 3,436 primary solid bone tumors in children and adolescents (left) and adults (right) as registered by the Basel Bone Tumor Reference Center

diagnosis. In our experience, a history of three months for osteosarcomas of the femur and of 6–8 months for Ewing sarcomas of the pelvis is very typical.

Much more common than malignant bone tumors are the *benign variants*. Very few of these lesions cause any symptoms, and most benign tumors are discovered as chance findings when x-rays are recorded for other reasons or in the event of a pathological fracture. A correct assessment of the findings and their significance is very



important in such cases. *Soft tissue tumors* in children and adolescents are even rarer than bone tumors. In this age group they occur particularly in association with systemic illnesses (e.g. neurofibromatosis) or as congenital hamartomas (e.g. congenital fibromatosis). The locally aggressive desmoid tumor is not infrequently seen in adolescence, whereas the main malignant tumor observed at this age is a rhabdomyosarcoma.

### History

The history plays a key role not only in the detection of a tumor, but also in the evaluation of its activity. Pain triggered by tumors is not usually clearly load-related, although such load-related pain can occur if the tumors interfere with the mechanics of the bone or even lead to a loss of stability. But usually the typical tumor pain is caused by tissue displacement and a feeling of tension. This pain is perceived to a much greater extent when there is nothing else to distract the patient, i.e. particularly at night. Cell growth is also more pronounced during the night than the day, since growth hormone is primarily secreted at night [1]. But this pain pattern is also typical of infections.

**! Unilateral pain that is not clearly load-related should always raise the suspicion of a tumor or inflammation. This also applies to nocturnal pain in the spine.**

Nocturnal pain in the legs, particularly in the knee area, is very common in small children between the ages of three and eight. These are described as »*growing pains*« (► Chapter 3.3.3.1). Fortunately, it is not particularly difficult to differentiate between these pain sensations: growing pains usually occur (alternately) on both sides, which is never the case with painful tumors (■ Table 4.11).

### Examination findings

The *clinical examination* is very important, particularly for soft tissue tumors, although it can also often provide valuable information for the diagnosis of bone tumors. We assess any protuberance by palpation according to the following *criteria*:

- *Consistency*: hard, soft, firmly elastic,
- *Mobility* of the skin,
- *Mobility* on the underlying structure (e.g. on the bone),
- *Tenderness*,
- *Redness*,
- *Excessive warmth*.

Hard painless tumors close to a joint are usually *osteochondromas*. The overlying skin is invariably very mobile, whereas the tumor itself is firmly fused with the bone. Redness is only observed over mechanically exposed sites. Firmly elastic, painless soft tissue nodules are most likely to be *ganglia* or *cysts* (a typical lesion in children is the popliteal cyst). Soft protuberances in the subcutaneous tissues that are highly mobile over the underlying structures are typical of a lipoma. Fairly rough, poorly demarcated areas of hard tissue and protuberances are indicative of a *fibromatosis* or *desmoid*. Painful, moderately hard protuberances are highly suspicious of a *malignant tumor*.

### Laboratory investigations

The most important differential diagnosis to be considered in relation to bone tumors is always an infection (*osteomyelitis*, *septic arthritis*). Infections can also cause nocturnal pain, swellings, redness and protuberances. Laboratory investigations (differentiated white cell count, erythrocyte sedimentation rate, CRP) can often help in establishing the correct diagnosis, although it should be borne in

■ **Table 4.11.** Pain characteristics of tumors or tumor-like lesions during childhood and adolescence (malignant tumors are shown in red colour)

Tumors that produce no pain	Tumors that produce no pain or only mechanical pain	Tumors that produce nocturnal pain	Tumors that produce severe nocturnal pain
<b>Bone tumors and tumor-like lesions</b>			
Non-ossifying bone fibroma	Osteochondroma	Osteblastoma	Osteoid osteoma, <b>osteosarcoma</b>
Enchondroma		Chondroblastoma, hemangioma, giant cell tumor	
Simple bone cyst		Aneurysmal bone cyst, <b>Ewing sarcoma</b> , <b>chondrosarcoma</b>	
<b>Soft tissue tumors and tumor-like lesions</b>			
Fibroma, lipoma	Desmoid, ganglia, cysts	Hemangioma and other vascular tumors, <b>sarcomas</b>	

#### 4.5.1 · Basic aspects of tumor diagnosis

mind that the laboratory results can still be normal in the earliest stages of an acute inflammation and that a case of primary chronic osteomyelitis often shows no changes in the laboratory test results at all (► Chapter 4.3.1.2). The inflammatory parameters are generally negative in the case of malignant bone tumors (except for Ewing sarcomas), and any changes tend to occur at a late stage. The *alkaline phosphatase* level is a factor to consider in the tumor diagnosis since it is generally elevated in osteosarcomas. It serves as an indicator of osteoblast activity. The serum level of alkaline phosphatase is also a good indicator for the response of the tumor to chemotherapy [4].

##### 4.5.1.2 Imaging procedures

❗ **In any patient with unilateral musculoskeletal pain that is not load related, plain x-rays of the affected area should be recorded in two planes.**

The *conventional x-ray* is very characteristic for most bone tumors and provides important information for the subsequent course of action (■ Fig. 4.30). Under no circumstances is any other (more expensive) investigation indicated as the *primary imaging method* in cases of sus-

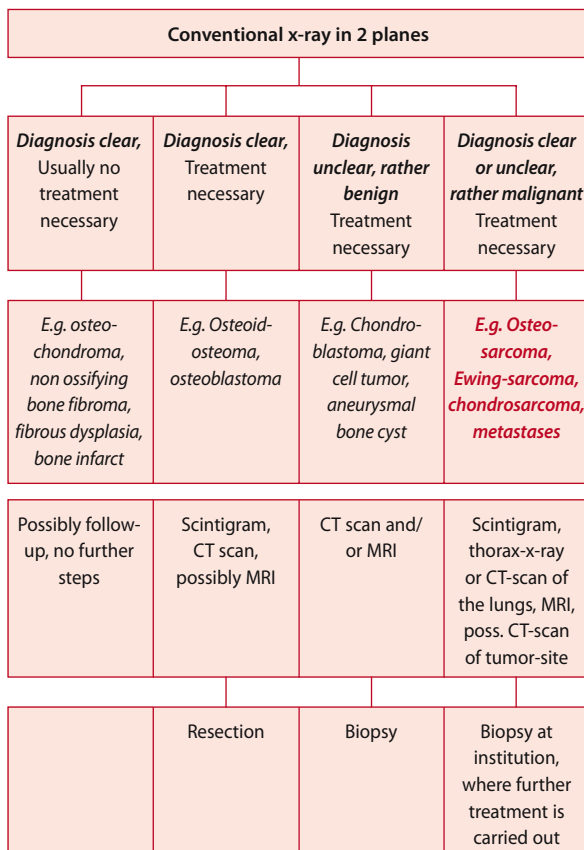
pected bone tumor, with the possible exception of cases involving a reasonable suspicion of a soft tissue tumor (MRI), but only if doubts exist about the benign or malignant status of the process. A suitable (and inexpensive) primary imaging investigation for *soft tissue processes* is a *sonogram*, since it can differentiate effectively between fluid-containing tissues (e.g. ganglia, bursae, hematomas) and solid tissues.

##### Conventional x-ray

The conventional x-ray shows very characteristic changes for any tumor, some of which are induced by the tumor itself and some of which represent the reaction of the bone to the tumor. Although few tumors can be diagnosed with complete certainty on the basis of conventional general x-rays, more extensive imaging procedures can only strengthen or weaken a suspicion. While a simple x-ray may not be able to produce a clear diagnosis in the case of tumors, nor is any other imaging method capable of providing a one-hundred percent reliable diagnosis.

The very *site* of a lesion within bone (■ Table 4.12) can provide valuable information about the expected diagnosis. Thus, for example, chondroblastomas almost invariably affect the epiphyses, while osteosarcomas are usually located in the metaphyses, and the rare adamantinomas are predominantly found in the diaphyses. The following tumors are *not primarily located in the epiphyses* [6]: *Ewing sarcoma, osteochondroma, simple bone cyst, non-ossifying bone fibroma, aneurysmal bone cyst*. Giant cell tumors, which are frequently located in the epiphysis or metaphysis, also do not occur at purely epiphyseal level if the growth plates are open [8].

Lodwick and Wilson [9] investigated the phenomenology of solitary bone tumors and related their radiographic morphology to the *biological behavior and patho-*



■ Fig. 4.30. Diagnostic-therapeutic algorithm based on the conventional x-ray

■ Table 4.12. Typical sites of tumors within the long bones (malignant tumors are shown in red)

Site	Tumor
Epiphysis	Chondroblastoma, <b>clear cell chondrosarcoma</b>
Metaphysis	Osteochondroma, non-ossifying bone fibroma, juvenile bone cyst, osteoblastoma, giant cell tumor (usually with epiphyseal involvement), aneurysmal bone cyst, <b>osteosarcoma, chondrosarcoma</b>
Diaphysis	Fibrous dysplasia, osteofibrous dysplasia, <b>Ewing sarcoma, adamantinoma</b>
Secondarily in diaphysis	Osteochondroma, non-ossifying bone fibroma, juvenile bone cyst

logical anatomy. Since their classification already provides valuable information about the aggressive nature to be expected, without any knowledge of the histology, it will be described briefly below. Tumors and tumor-like lesions always involve bone formation and resorption. If bone breakdown predominates, osteolysis results, whereas excessive bone formation results in osteosclerosis. The turnover processes differ depending on whether cancellous or cortical bone is involved.

The above statements indicate that the site is very important for the appearance of the tumor on the x-ray. While the degree of loading influences the reaction to tumor growth, the appearance on the x-ray is most strongly affected by the rate of tumor growth.

**Destruction pattern in compact and cancellous bone according to Lodwick and Wilson [9]**

The classification system involves three basic patterns of bone destruction:

- I: geographic (map-like), primarily involving the cancellous bone,
- II: mixed forms (geographic and moth-eaten/permeative),
- III: moth-eaten lesion, in compact and cancellous bone, or permeative destruction in the compact bone only.

Various grades are differentiated according to the reaction of the compact bone and the penetration of the cortex in each case (Table 4.13, Fig. 4.31). In the case of slow growth, the surrounding healthy bone reacts by

the formation of new stabilizing bone (sclerosis, increased thickness). In the case of faster growth the bone does not have time to react with new bone formation, and osteolysis only is visible on the x-ray.

**Periosteal reactions**

Tumors can produce widely differing periosteal reactions (Table 4.14, Fig. 4.32) that involve the formation of new bone. But these are not visible on the x-ray until they

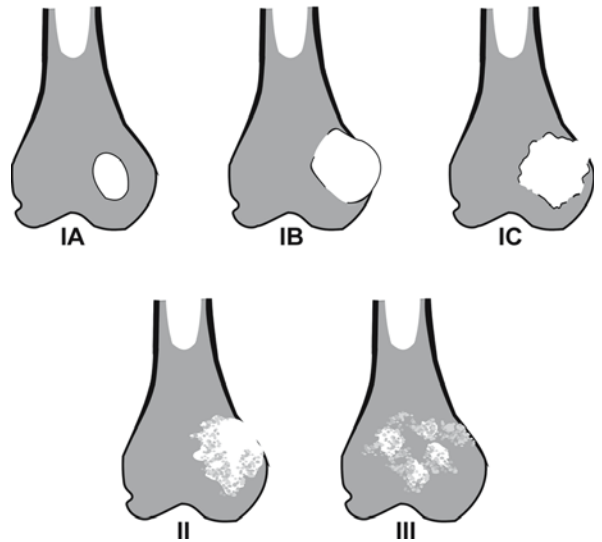


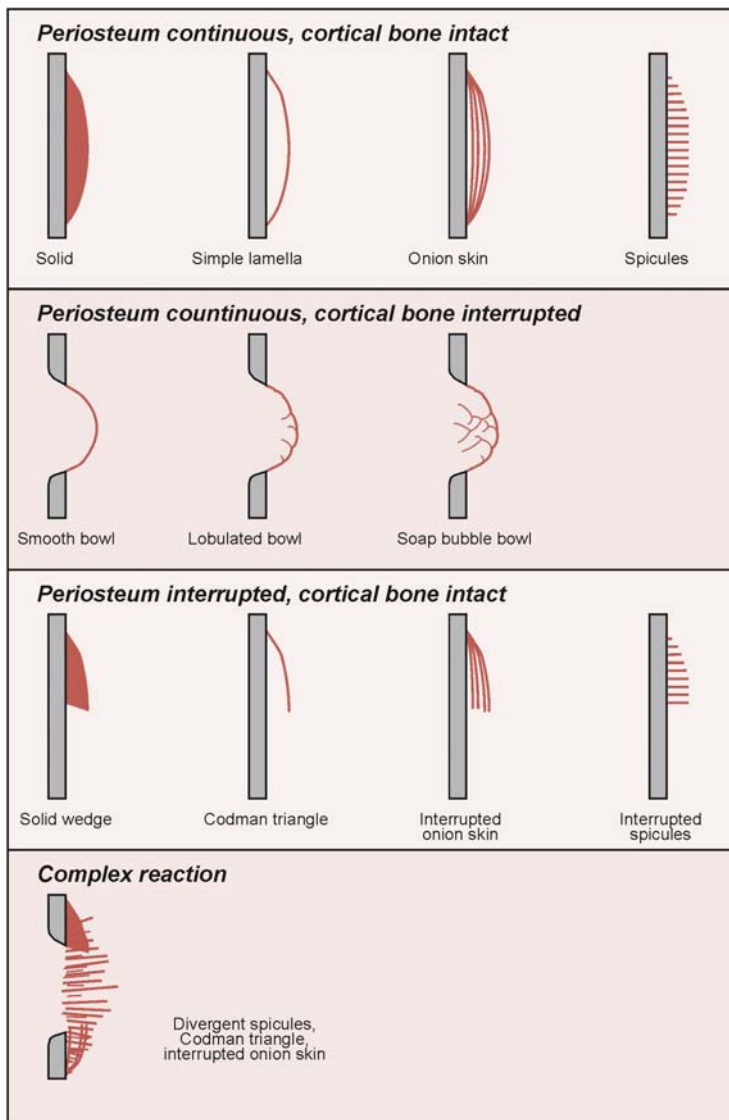
Fig. 4.31. Destruction pattern in bone on the x-ray according to Lodwick. See Table 4.13 for details of the classification

Table 4.13. Radiological grading of bone tumors based on the reaction of the compact bone and the penetration of the cortex

Type (grade)	Destruction	Contours	Compact bone penetration	Sclerosis	Growth	Periosteal reaction	Typical examples
IA	Geographic	Sharply-defined	No	Yes	Slow	None	Enchondroma, non-ossifying bone fibroma, osteoid osteoma
IB	Geographic	Ragged, irregular	No, poss. partially	Mostly yes	Slow	Solid	Giant cell tumor, chondroblastoma, juvenile bone cyst, osteoblastoma, chondromyxoid fibroma, aneurysmal bone cyst
IC	Geographic	Poorly-defined, reef-like	Yes	Possible	Slow	Solid	Chondrosarcoma, aneurysmal bone cyst
II	Mixed geographic and moth-eaten/permeative	Poorly-defined	Yes	Mostly no	Intermediate	Bowl-shaped	Osteosarcoma, fibrosarcoma, chondrosarcoma
III	Moth-eaten/permeative	Poorly-defined	Yes	Mostly no	Fast	Radial, onion-skin-like, complex	Ewing sarcoma, osteosarcoma

■ **Table 4.14.** Types of periosteal reaction

Periosteum	Cortical bone	Appearance	Typical lesions
Continuous	Intact	Solid single lamella onion skin, spicules (radial)	Chronic osteomyelitis, Langerhans cell histiocytosis, osteoid osteoma, Chronic osteomyelitis, Langerhans cell histiocytosis Acute osteomyelitis, <i>Ewing sarcoma, (osteosarcoma)</i>
Continuous	Destroyed	Single bowl lobulated bowl ragged bowl	Aneurysmal bone cyst, enchondroma, chondroblastoma, Chondromyxoid fibroma, fibrous dysplasia, giant cell tumor Chondrosarcoma, plasmacytoma, metastases
Interrupted	Intact	wedge-shaped  Codman triangle, interrupted onion skin, radial	Aneurysmal bone cyst, giant cell tumor, chondromyxoid fibroma Aneurysmal bone cyst, <i>osteosarcoma, Ewing sarcoma, chondrosarcoma</i>
Interrupted	Destroyed	Combinations of Codman triangle, interrupted onion skin, divergent rays	Osteosarcoma



■ **Fig. 4.32.** Periosteal reaction on the x-ray. See Table 4.14 for details of the classification. (According to Freyschmidt and Ostertag [3])

mineralize, a process that takes 10–20 days and is age-related (the older the patient the longer the process). The morphology is determined by the aggressivity and duration of the underlying process. The periosteal reaction can either be continuous or intermittent, with or without cortical destruction.

#### Matrix mineralization

Some tumors form a matrix, a cell-free intercellular ground substance that *mineralizes*, i.e. introduces calcium salts. Typical *matrix-forming tumors* are:

- osteoblastoma, osteoid osteoma, osteosarcoma (matrix = bone ground substance or osteoid),
- osteochondroma, enchondroma, chondromyxoid fibroma, chondrosarcoma, (matrix = cartilaginous ground substance),
- desmoplastic fibroma, fibrosarcoma (matrix = collagen fibers),
- fibrous dysplasia (matrix = mixed: osteoid and collagen fibers).

The *osteosarcoma* can also produce a *mixed matrix* (osteoid, chondroid or collagen fibers). Depending on the prevailing matrix type the osteosarcoma can be described as osteoblastic, chondroblastic, fibroblastic, etc.

The following tumors and tumor-like lesions *do not form a matrix*: simple bone cyst, aneurysmal bone cyst, giant cell tumor, Ewing sarcoma.

The pattern of radiologically visible *matrix calcification* can provide an indication of the underlying, matrix-forming tumor type. Depending on the degree of mineralization, the osteoid shows a cloud-like or solid, or even ivory-like appearance on the conventional x-ray. Benign lesions such as the *osteoblastoma* can also show a peripheral, seam-like zone of brightening, since the tumor matrix in these immature proliferating areas has not usually yet mineralized.

The extent of mineralization associated with *osteosarcomas* varies and can range from usually ill-defined, cloud-like areas of condensation to highly sclerosed, ivory-like zones. Slow-growing tumors can cause trabecula-like areas of calcification. In *fibrous dysplasia* the formation of irregular metaplastic fibrous bone is accompanied by collagen fiber production. Characteristically, the mineralization in such cases leads to a relatively uniform increase in density and produces the ground-glass opacity on the x-ray that is typical of fibrous dysplasia.

The *cartilaginous ground substance* can also mineralize directly. Such calcifications may have a stippled or fluffy appearance. If enchondral ossification occurs at the periphery of cartilaginous tumors, which often appear to have a lobular structure on histological examination, then this non-tumorous bone calcifies, producing radiologically visible ring- or arc-shaped condensation

patterns. These are typical of *enchondromas*, *osteochondromas* and *chondrosarcomas*. Necrotic areas (*bone infarcts*) can calcify and ossify secondarily. The reactive formation of new bone that mineralizes can occur in varying degrees in almost all lesions and may obscure the basic picture.

#### Bone scan and positron emission tomography (PET)

The technetium 99 bone scan is a relatively non-specific investigation that enables the blood flow of a process, and thus bone turnover activity, to be evaluated. Active processes show greatly increased uptake, whereas older, »burnt-out« processes show little uptake. Particularly high levels of uptake are observed for bone-forming tumors such as osteoid osteoma, osteoblastoma and osteosarcoma. A case of *osteomyelitis* can be differentiated from a tumor (e.g. Ewing sarcoma) by adding gallium 67 as a »tracer« or by means of antigranulocyte immunoscintigraphy. The bone scan is less suitable for evaluating the extent of a tumor, since the activity uptake extends beyond the tumor margins.

**! The bone scan is the simplest and most cost-effective method for detecting bone metastases (including skip metastases), and should be implemented if there is any suspicion of a malignant tumor or to rule out multiple lesions.**

Another valuable investigation is *positron emission tomography (PET)*, which appears to be a particularly sensitive method for locating metastases.

#### Ultrasound

Ultrasound is primarily suitable for the primary evaluation of *soft tissue processes*, especially for distinguishing between fluid-containing and solid tumors. It can therefore be used as an inexpensive primary screening method if a soft tissue tumor is suspected before deciding whether an expensive MRI scan is indicated. Ultrasound is also suitable for the initial scanning for metastases in the abdomen.

#### CT scan

Despite the popularity of the MRI scan and its outstanding discriminatory power, particularly for soft tissues, computed tomography has not lost any of its importance. The CT scan is better for distinguishing intraosseous processes (particularly in cancellous bone). The extent of a tumor, particularly in the vicinity of a joint, in the pelvic area and the spine, can be determined very precisely by recording transverse slices. In critical cases however (particularly in establishing whether the tumor has penetrated into the joint or not), the inability to record sagittal and frontal slices is a drawback. Computer-assisted reconstruction in one of these planes is only helpful if the resolution (which depends on the slice thickness) is good



enough. In appropriate cases, targeted thin-slice CT scans are indicated.

- ❗ **The CT scan is particularly suitable for the evaluation of the intraosseous spread of the tumor in cancellous bone and is indicated for all tumors near a joint that must be removed surgically (particularly outside the lesion). It is also superior to conventional x-rays or MRIs in the search for metastases in the lung. If abdominal metastases are suspected on the basis of the sonogram, a spiral CT with contrast medium will provide further information. The best information about the existence and localization of metastases is obtained with the combination of positron emission tomography (PET) and the CT scan (PET/CT). The data from the two examinations are digitally combined to produce an exact anatomical assignment of the findings.**

### Magnetic resonance imaging (MRI)

Magnetic resonance imaging has greatly improved the diagnostic options for evaluating bone tumors. The *main advantages compared to the CT scan* are as follows:

- better tissue characterization: better evaluation of the tumor matrix,
- clearer definition of the tumor in the soft tissues and bone marrow,
- any spatial slicing possible (sagittal, horizontal and coronal slicing).

The tissue characterization is achieved primarily by the differing weighting of the MRI images. ▶ Chapter 2.1.4.1 discusses the characterization of MRI images by the repetition time (TR) and echo time (TE) and special techniques such as fat suppression and the effect of contrast medium.

- ❗ **In view of its superior performance in tissue differentiation, the ability to evaluate the spread of tumors in the soft tissues and bone marrow and their relationship with the major nerves and vessels, an MRI scan is essential nowadays before the surgical resection of any malignant bone and soft tissue tumor. But: Far too many MRI scans are ordered these days. Because it is not possible to identify a benign lesion solely on the basis of the history, clinical findings and a conventional x-ray or sonogram, this certainly does not mean that an expensive MRI scan is always indicated, particularly since it may not reveal the diagnosis in any case. In such cases it would be more appropriate to send the patient, or at least the images, to a colleague with more experience in the diagnosis and treatment of bone tumors.**

### Angiogram

Although the (non-invasive) MRI is very good for showing the relationship between the tumor and the major vessels, an angiogram is occasionally indicated since it is better at showing the course of the vessels and the lesion-related topography of the vessels. Angiography is indicated particularly in cases of vessel bridging or preoperative embolization. Vascular imaging can be performed conventionally, as MR angiography (digital subtraction angiography; DSA) or, for outstanding quality images, as spiral CT (▶ Chapter 2.1.4.1).

#### 4.5.1.3 Biopsy

A biopsy must be arranged for all lesions that cannot be clearly diagnosed on the basis of imaging investigations or if malignancy is suspected. The following *options* are available:

- fine needle biopsy,
- percutaneous biopsy (e.g. with a trephine),
- open biopsy.

The *fine needle biopsy* only allows a tiny cylinder to be removed, which is not sufficient for confirming the diagnosis of an unclear tumor, which often shows a heterogeneous structure. *For bone tumors, CT-guided fine needle biopsy is recommended only in very experienced centers* [10, 13]. All too often the material is unusable, or conditions are misdiagnosed. But fine needle biopsy can, at least, usually indicate whether a metastasis or primary tumor is involved. It is also important to mark the entry site so that the contaminated biopsy channel can subsequently be removed with the tumor.

A *trephine biopsy* can be performed under image intensifier or CT control. A standard trephine (or core trephine) used for removing broken screws is not suitable for this purpose since it transports the drilled bone inwards and crushes the withdrawn cylinder. In addition, the biopsy material is thermally damaged as a result of the build-up of heat, rendering it almost impossible to evaluate. More appropriate instruments are *special trephines* that transport the bone fragments outwards and that incorporate a special device for ejecting the cylinder. In cortical bone the heat build-up can be so excessive that the excised material becomes completely necrotic despite the use of the special trephine. On the other hand, a good result can usually be achieved when such a trephine is used in cancellous bone. In most cases, however, an *open biopsy* is indicated. Examination of a frozen section may reveal whether representative lesional tissue was biopsied, rather than the diagnostically unproductive marginal zone. But in any case, frozen sections should only be evaluated by a pathologist with considerable experience in bone tumor diagnosis.

This frozen section and/or biopsy diagnosis should never be implemented without any knowledge of the radiological morphology. The surgeon must therefore provide the pathologist at least with general x-rays in two planes. A histological bone tumor diagnosis without knowledge of the x-ray findings may amount to malpractice, especially when cartilaginous tumors are being investigated. Since bone tumors are usually mineralized, a reliable histological assessment is often possible only after decalcification several days after the biopsy. Moreover, diagnostic »rush jobs« for a restlessly waiting surgeon and anesthetist can result in mistakes. Since, in any case, high-grade malignant tumors are precisely the type that do not require resection primarily, there is no immediate need for an intraoperative diagnosis.

On the other hand, if expert evaluators are available, *excision biopsy* is permissible for certain benign, but locally aggressive, tumors (occasionally even for small low-grade malignant tumors). This particularly applies if the reconstruction does not pose any special problems or if the patient will not have to cope with any major drawbacks.

*Needle biopsy* is a simpler procedure for *soft tissue tumors* since no bone resistance has to be overcome. But here, too, there is a considerable risk that insufficient material is collected or that the sample is not representative. A *sentinel node biopsy* is indicated for malignant soft tissue tumors that metastasize into the lymph nodes (these include, in particular, synovial sarcomas and epithelioid-cell sarcomas). In this procedure a radioactive marker is injected into the tumor and is then transported, via the lymphatic drainage system, into the first node, or »sentinel node«, of the primary lymph node station. This is then removed and examined by frozen section.

**! If malignant tumors are present, the biopsy procedure may result in contamination of the biopsy channel with malignant cells [11–12]. During tumor resection, therefore, the biopsy channel must always be removed together with the tumor en bloc. Accordingly, the procedure for the resection and reconstruction must already be known at the time of biopsy. For this reason, if a malignant tumor is suspected, the biopsy should always be performed in a hospital where the potentially necessary treatment can also be provided.**

### Technique of open biopsy

The biopsy should always be performed in such a way that the biopsy channel can be resected together with the tumor en bloc. For this reason, the biopsy should *never be performed via the traditional access routes* (which are usually crossed by vessels and nerves), but always 1–2 cm away, through the muscle proceeding directly to the bone.

Thus, the subsequent resection can use a standard access route and include the resection of the adjacent biopsy channel. Vessels and nerves are not contaminated by the biopsy. Within the bone, the *active areas* in particular must be biopsied. These tend to be located at the periphery of the tumor, where the most aggressive areas of osteolysis are visible on the x-ray. *Hohmann retractors* should never be used to expose the bone. These are placed around the bone and are especially useful for retracting the muscles. However, since the tip of the Hohmann retractor is rotated around the bone, tumor cells can be transported behind the bone [5], possibly promoting the further spread of the tumor.

A wedge-shaped sample of the tumor measuring at least 2 cm<sup>3</sup> (or larger if possible) should be taken from a representative site (i.e. the most active site according to the imaging investigations). At the end of the operation, any *Redon drain* must be inserted very carefully so as to avoid contamination of any new compartments. The drain should be pushed through the skin at a maximum distance of 10 mm from the end of, and aligned with, the incision. The *skin* should be *sutured* not with transverse interrupted sutures, but with an intracutaneous continuous suture. The biopsied tissue should always be *unfixed* and just slightly cooled (never frozen) as soon as possible (as for a frozen section) and forwarded to a competent pathologist by prior arrangement. Fixation in formalin can interfere with, or even make impossible, important diagnostic investigations (e.g. RNA-based PCR investigations; see below).

### Summary of the seven deadly sins associated with biopsy

- access via the classical access route
- collection of insufficient and unrepresentative tumor sections
- use of Hohmann retractors
- Redon drain far away from the incision
- skin sutured with interrupted sutures
- fixation of the excised material in formalin
- biopsy of malignant tumors not in the same hospital used for the treatment

### Special aspects of the biopsy from the standpoint of the pathologist

A bone biopsy is indicated if a clear diagnosis cannot be obtained by conventional radiological and clinical means. This unclear situation requires consultation, before the biopsy, between the clinician / radiologist and the pathologist to discuss the procedure to be adopted. If possible, this consultation process, including a discussion of the radiographic findings, should clarify the following questions:

#### 4.5.1 · Basic aspects of tumor diagnosis

##### — What can be expected?

The history, clinical and radiological findings provide an indication of the type of lesion involved (*tumor, tumor-like lesion, inflammation, metabolic bone changes or a (post-) traumatic situation*).

##### — Are additional investigations required?

The differential diagnostic ranking will produce various options for substantiating or ruling out clinical conditions by means of additional investigations. An undifferentiated sarcoma, for example, can be identified as an osteosarcoma if enzyme histochemical tests are positive for alkaline phosphatase. *However, this test can only be performed on unfixed tissue that is shock-frozen.* The same applies to molecular biological investigations and the detection of the translocation that is typical of Ewing sarcoma. Microbiological investigations should be arranged if osteomyelitis is suspected.

##### — Can the additional investigations and/or likely required therapeutic procedures be implemented on site?

The answer to this question is of crucial importance to the subsequent outcome. If the doctor lacks the diagnostic and therapeutic experience, irreparable mistakes that impair the prognosis can be made even at the biopsy stage. Consequently, the decision as to whether the patient can subsequently be treated on site or will need to be transferred to a specialist hospital must be made *before* the biopsy.

#### Remarks on the biopsy procedure

If the differential diagnostic alternatives are clear and the patient can be treated on site, the orthopaedist and pathologist must agree on the timing of the biopsy. The surgeon should collect a sufficiently large tissue sample – approx. 0.5 cm thick and the size of a stamp – that is oriented from the periphery to the center of the tumor. If it is not macroscopically obvious, the peripheral end of the tumor should be *marked* (suture, ink). The pathologist must possess precise knowledge, on the basis of the x-ray, of the biopsy site. The tissue should be well cooled (but not frozen) as soon as possible (ideally under frozen section conditions) and forwarded for further investigations. Imprint cytology can be used to prepare unfixed biopsy material and samples shock-frozen for additional investigations (see above). A *frozen section diagnosis* is then required only if it involves therapeutic consequences. The frozen section evaluation can be helpful in ensuring that representative tumor tissue has been collected so that further material can be biopsied in the same session if necessary.

#### 4.5.1.4 Pathological anatomy, histology

Even the macroscopic appearance can provide an indication of the expected histological findings. As a rule, all

of the biopsy material should be examined histologically (unless it is required for any other special investigations), otherwise there is a risk that diagnostically crucial findings, which are often only present in small sections in bone tumors with a heterogeneous structure, may be overlooked. In addition to matrix formation (osteoid, chondro-osteoid, hyaline cartilaginous or myxoid cartilage matrix), the cellular composition of the lesion in particular should be examined, and the pathologist will need to establish, whether any matrix is formed from tumor cells or whether e.g. reactive, though occasionally very immature, pseudosarcomatous new bone formation is involved. It should always be borne in mind that giant cells occur in numerous lesions and can frequently confuse the diagnostician.

Pseudocystic, blood-filled cavities are also not necessarily synonymous with the diagnosis of an aneurysmal bone cyst, but can also occur as secondary phenomena as constituents of other lesions. The possibility of callus-like new bone formation with superimposed microfractures should also be considered. For these reasons, the tentative histological diagnosis should always be checked against the conventional x-ray. If any discrepancies arise between the radiological and the histological diagnosis, and if these are not satisfactorily resolved in the interdisciplinary discussion, even including one with experienced specialists, a further biopsy should be performed, possibly in a center with corresponding diagnostic and therapeutic experience.

#### 4.5.1.5 Tumor staging

##### **Bone tumors**

Once the diagnosis has been confirmed, the overall situation must be evaluated so that the appropriate therapeutic steps can be initiated. The usual staging system for tumors, i.e. the TNM system, is not suitable for bone tumors because, on the one hand, the regional lymph nodes are not involved (since they are rarely affected) and, on the other, the compartment is a very important factor to be considered in the assessment. For these reasons Enneking [2] has introduced a separate staging system for bone tumors that takes account of the following parameters:

- the histological *differentiation grade* (G),
- the *anatomical situation* of the tumor (T) (i.e. whether intra- or extracompartmental),
- the *metastases* (M).

In respect of the *histological differentiation grade* (G), G0 refers to a *benign tumor*, G1 to a *highly differentiated (low-grade) malignant tumor*, and G2 to a *poorly differentiated, high-grade malignant tumor*. As regards the *anatomical situation* (Site: T) we distinguish between T1, *intracompartmental tumors*, and T2, *extracompartmental tumors*. In principle, a bone tumor becomes extracompartmental

as soon as it breaks out of the bone into the surrounding soft tissues. *Metastases* are either *not detectable* (M0) or have been *confirmed* by imaging investigations (M1). Accordingly, *benign tumors* can be divided into three stages (■ Table 4.15) and *malignant tumors* into six stages (■ Table 4.16). Staging of the tumor enables the orthopaedist to decide on the appropriate treatment (► Chapter 4.5.5).

### Soft tissue tumors

Like bone tumors, soft tissue tumors must also be staged. Apart from the histological *differentiation grade* (G), the *anatomical situation* of the tumor (T) – i.e. as well as the compartment the tumor size is crucial here – and the presence of *metastases*, the regional lymph nodes should also be included in the staging process (N). ■ Table 4.17

■ Table 4.15. Staging of benign bone tumors. (After [2])

Stage	Histological differentiation (Grade = G)	Anatomical situation (Site = T)	Metastases (= M)	Radiological type	Clinical course
1	G0	T0	M0	I A	Latent, static, self-healing
2	G0	T0	M0	I B	Active, expansive, causes bone to swell
3	G0	T1–2	M0	I C	Active, expansive, can break through the cortex

■ Table 4.16. Staging of the malignant bone tumors. (After [2])

Stage	Histological differentiation (Grade = G)	Anatomical situation (Site = T)	Metastases (= M)
I A	G1	T1	M0
I B	G1	T2	M0
II A	G2	T1	M0
II B	G2	T2	M0
III A	G1–2	T1	M1
III B	G1–2	T2	M1

■ Table 4.17. Staging of malignant soft tissue tumors according to the UICC Staging System

Stage	Histological differentiation (Grade = G)	Anatomical situation (Site = T)	Lymph nodes (Nodes = N)	Metastases (= M)
IA	G1 (differentiated)	T1a/b ( $\leq 5$ cm)	N0 (none)	M0 (none)
	G2 (moderate)	T1a/b ( $\leq 5$ cm)	N0 (none)	M0 (none)
IB	G1 (differentiated)	T2a ( $> 5$ cm)	N0 (none)	M0 (none)
	G2 (moderate)	T2a ( $> 5$ cm)	N0 (none)	M0 (none)
IIA	G1 (differentiated)	T2b ( $> 5$ cm)	N0 (none)	M0 (none)
	G2 (moderate)	T2b ( $> 5$ cm)	N0 (none)	M0 (none)
IIB	G3 (dedifferentiated)	T1a/b ( $\leq 5$ cm)	N0 (none)	M0 (none)
	G4 (dedifferentiated)	T1a/b ( $\leq 5$ cm)	N0 (none)	M0 (none)
IIC	G3 (dedifferentiated)	T2a ( $> 5$ cm)	N0 (none)	M0 (none)
	G4 (dedifferentiated)	T2a ( $> 5$ cm)	N0 (none)	M0 (none)
III	G3 (dedifferentiated)	T2b ( $> 5$ cm)	N0 (none)	M0 (none)
	G4 (dedifferentiated)	T2b ( $> 5$ cm)	N0 (none)	M0 (none)
IV	G1–4	T1–2	N1 (present)	M0 (none)
	G1–4	T1–2	N0/1 ( $\pm$ )	M1 (present)

**Table 4.18.** Tumor staging in the UICC system

Stage	Size and anatomical situation of the tumor
T1a	Tumor diameter ≤5 cm, no infiltration of the fascia
T1b	Tumor diameter ≤5 cm, with infiltration of the fascia
T2a	Tumor diameter >5 cm, no infiltration of the fascia
T2b	Tumor diameter >5 cm, with infiltration of the fascia

shows the *staging system* of the Standardization Committee of the *International Union Against Cancer* (formerly the »Union Internationale contre le Cancer« UICC) [14, 15]. Since it includes tumor size it is slightly more precise than that of the *American Joint Committee on Cancer Staging (AJCC)*, which is also commonly used. The classification of the *anatomical situation* of the tumor (T) in the UICC system is shown in **Table 4.18**.

Staging is a valuable aid in establishing the therapeutic course of action.

#### 4.5.1.6 Basic aspects on the treatment of musculoskeletal tumors

The various diagnostic measures should be established only in close consultation with a specialist center. The responsibility for planning and implementing the biopsy (incl. fine needle biopsy) must always rest with the person who is also responsible for the definitive surgical treatment. Part of the correct biopsy procedure includes the appropriate planning of the potential access route as this must also be taken into consideration at the time of resection.

Many mistakes are also made in relation to benign tumors and tumor-like lesions, e.g. unnecessary biopsies and treatments for non-ossifying bone fibromas, enchondromas, bone infarcts etc. The recurrence rate for active tumors (e.g. giant cell tumors) varies according to whether the treatment was administered in a specialist center or not, from less than 10% to as high as 50%.

Malignant tumors should always be treated in a specialist center with a multidisciplinary team (including, in addition to an orthopaedist specialized in tumors, an oncologist, a pathologist with experience in sarcoma diagnosis, a musculoskeletal radiologist and a radio-oncologist) [7].

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## 4.5.2 Benign bone tumors and tumor-like lesions

F. Hefti, G. Jundt

### ➤ Definition

Benign bone tumors occur predominantly during the first two decades of life. Of the total of 2,194 primary benign bone tumors registered since 1972 by the Basel Bone Tumor Reference Center, 43.3% (949 cases) occurred in this age group [25]. Benign bone tumors are much more common than would be suggested by the number diagnosed since they usually remain clinically asymptomatic. In accordance with the proposal of the WHO bone tumors are described according to the tissue type that they most resemble in terms of their composition [50]. The description of the findings is essentially based on our own experience and on numerous monographs [10, 33, 51, 57], which also provide a comprehensive overview of the relevant literature.



### 4.5.2.1 Bone-forming tumors

#### Osteoid osteoma

##### ➤ Definition

Benign tumor, usually about the size of a grain of rice (<1 cm), that forms substantial amounts of osteoid. It is painful and characterized by a nidus.

##### Occurrence, site

Relatively *common* tumor (approx. 11% of all benign bone tumors), particularly during *adolescence*. The male:female ratio is 2:1. The tumor is typically located in the diaphyseal or metaphyseal/diaphyseal cortex of long bones. Multicentric or multifocal osteoid osteomas are very rare [49]. Their occurrence at the same site in identical twins has also been reported [28].

##### Clinical features, diagnosis

An osteoid osteoma is a painful tumor characterized by a highly typical nocturnal pain. The pain is often diffuse and cannot be located accurately by the patient, but responds very well to the administration of salicylates.

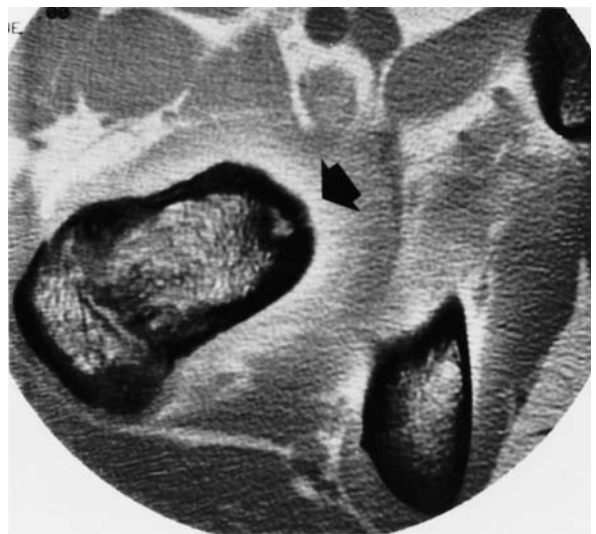
- The x-ray is very characteristic. An osteolytic zone the size of a grain of rice, the »nidus«, can be seen in the center of a cortical sclerotic area (■ Fig. 4.33, see also ■ Fig. 3.401, ► Chapter 3.4.9). A *bone scan* may reveal the »double-density sign« that is highly characteristic of the osteoid osteoma. This involves a highly active central zone surrounded by another zone of less active uptake. This finding is almost diagnostic. The nidus is also well visualized on a *CT scan* (■ Fig. 4.34).
- *Histologically* the nidus consists of loose, highly vascularized connective tissue with irregularly distributed, unmineralized trabeculae of woven bone with greatly proliferating active osteoblasts without atypia. The nidus is surrounded by a broad zone of sclerosed, cortical bone tissue.
- *Differential diagnosis*: The most difficult task is to differentiate the tumor from a *stress fracture* (► Chapter 3.3.3.7). A stress fracture, particularly of the tibia, also results in hyperostosis, and the fracture appears as a fine line of osteolysis. However, the symptoms are related to exertion and respond minimally, if at all, to salicylates. A corresponding history is also usually present. A *sclerosing periosteitis* can also resemble an osteoid osteoma, although the condensation in these cases is purely periosteal and not oriented toward the medullary cavity. An intracortical *osteomyelitis* may show similar pictures.

##### Prognosis, treatment

While the tumor may heal spontaneously after 2–3 years, this is not always the case [24]. During growth, the tumor may stimulate the growth plate, resulting in hypertrophy of the affected extremity.



■ Fig. 4.33. X-ray of the proximal femur in a 13-year old boy with *osteoid osteoma*. The cortical bone is thickened on both the medial and lateral sides (arrows)



■ Fig. 4.34. CT scan of a 12-year old boy with an *osteoid osteoma* in the femoral neck and a clearly visible nidus (arrow)

- *Treatment*: Curettage (intralesional resection) of the nidus is sufficient. CT-guided resection with a core trephine is usually recommended. Ablation should also be performed with a thermal probe, radioactive substances or alcohol [44].

## Osteoblastoma

### Definition

Benign osteoid-forming tumor (>1 cm). It is usually located in the cancellous bone, but can also occur periosteally and, apart from its size, cannot be distinguished histologically from an osteoid osteoma. It may show a tendency toward progressive growth.

### Occurrence, site

The osteoblastoma is rarer than the osteoid osteoma (approx. 3% of all benign bone tumors). Here too the male:female ratio is 2:1. Osteoblastomas occur particularly during adolescence. Around half of osteoblastomas are located in the spinal area, mostly at lumbar level [40] and predominantly in the pedicles. But they can also occur in any other bone, particularly the cancellous bone in the metaphyses, and more rarely the epiphyses.

### Clinical features, diagnosis

While an osteoblastoma also causes pain, this is less intense and less characteristic than for osteoid osteoma. In the spine osteoblastomas can produce antalgic scoliosis. In such cases, the osteoblastoma is usually located in the pedicle at the apex on the convex side of the scoliosis [40].

- The x-ray is less characteristic than for osteoid osteoma. The tumor can vary in size (and can grow as large as 10 cm [10]), is usually configured geographically and is predominantly lytic with slight floccular condensations. It may show irregular sclerosis, but usually only if it has been present for a long time. An AP x-ray of the spine initially shows widening of the pedicle. At a later stage, the normally oval pedicle may also appear eroded. The suspicion of an osteoblastoma can be strengthened by a *bone scan*, which may show a localized area of very high uptake (»hot spot«). The osteoblastoma is also well visualized on a *CT scan*.
- *Histology*: Osteoblastomas are rich in cells and consist of irregularly arranged fibrovascular tissue containing interconnected fibrous trabeculae of woven bone. The osteoblasts show active nuclei and occasionally typical mitoses, but no atypia. Bleeding and osteoclastic giant cells are always present.
- *Differential diagnosis*: Differentiating between an *osteoblastoma* and an *osteoid osteoma* is not very relevant as the latter are simply larger, histologically identical tumors (»bigger brother of the osteoid osteoma«). The osteoid osteoma is located in cortical bone and shows much greater perilesional new bone formation than the osteoblastoma. Large osteoblastomas can be confused with an *osteosarcoma*, although the latter always contains atypical cells and infiltrates at its periphery into the local bone. Associated purely cellular areas

should arouse suspicions of an osteosarcoma. Large osteoblastomas in the metaphyseal area of long bones can also be confused with giant cell tumors or, in the spine, with an *aneurysmal bone cyst*.

### Treatment, prognosis

Curettage is normally sufficient if performed carefully (with burring of the tumor cavity). It is unrealistic to perform anything more than curettage in the spine since an en-bloc resection would require excessive tissue destruction, and the use of phenol or similar substances in the immediate vicinity of the spinal cord is, in any case, out of the question. Consequently, recurrences can occur. However, recurrences are attributable to intralesional treatment since they are not observed after resections. To this extent, it is not appropriate to define a special aggressive form of osteoblastoma (»aggressive« or »malignant« osteoblastoma).

## 4.5.2.2 Cartilage-forming tumors

### Osteochondroma (cartilaginous exostosis)

The multilocular form, osteochondromatosis (also often incorrectly termed »hereditary multiple cartilaginous exostosis«) is discussed in ► Chapter 4.6.2.31.

### Definition

Since osteochondromas show characteristic cytogenetic aberrations in 8q22–24.1, i.e. also the site of the EXT1 gene which is mutated in osteochondromatosis, osteochondroma is probably also a genuine tumor rather than a hamartoma.

### Occurrence, site

Osteochondroma is one of the most common bone tumors (male:female = 2:1), but is rarely observed before the age of eight. No new lesions appear after the completion of growth. Most osteochondromas are located in the metaphyses of the long bones, primarily in the vicinity of the knee, and rarely in the diaphyses. The following metaphyses are affected in order of greatly decreasing frequency: distal femur, proximal tibia, proximal humerus, distal tibia, and proximal femur. Flat bones with apophyses (pelvis, scapula) can also be affected. Osteochondromas do not occur in the epiphyses.

### Clinical features, diagnosis

Osteochondromas are not primarily painful. Only if they are mechanically disruptive can irritation occur as a result of rubbing against muscles and tendons and thereby produce mild symptoms. The lesions can also appear unsightly and occasionally restrict joint mobility. On the scapula they are usually located on the anterior side and cause the shoulder blade to protrude. Osteochondromas

can also produce significant axial deformities and length discrepancies.

— **Radiologically**, a distinction can be made between broad-based sessile osteochondromas and pedunculated osteochondromas. The sessile forms involve cortical lesions that cover a wide area. Like the pedunculated versions, their base projects into the adjacent cancellous bone. The pedunculated forms develop finger-like projections and often possess a cauliflower-shaped tip that generally points towards the center of the diaphysis. The tumors are always sharply defined on x-rays by a thin layer of cortex (■ Fig. 4.35).

The tip of the tumors is formed by a cap of cartilage of varying width. Apart from the non-ossifying bone fibroma, the osteochondroma is the only tumor that can be diagnosed with complete reliability on the basis of conventional x-rays in two planes. The cartilaginous area of the tumor can occasionally show areas of calcification, although these are often not visible on x-rays. Further imaging investigations are usually unnecessary. CT scans are only required if the tumors are located in very specific sites (on the



■ Fig. 4.35. 11-year old boy with a broad-based *osteochondroma* on the medial aspect of the distal femur that has resulted in a growth disturbance (varus deformity)

scapula or the spine). The thickness of the cartilaginous cap can be determined by MRI (another example of osteochondromas is shown in ■ Fig. 3.524, ► Chapter 3.5.8).

- **Histologically**, the surface of osteochondromas consists of hyaline cartilage, which is usually a few millimeters thick and rarely wider than 2 cm. The cellularity of the cartilage decreases towards the surface. Fatty marrow, and occasionally blood-forming marrow as well, can be seen between the cancellous bone trabeculae.
- **Differential diagnosis**: The most difficult task in the case of large osteochondromas is to establish whether *malignant change* (»epiexostotic chondrosarcoma«) is present. The cap is usually wider than 3 cm in these cases, and any change in its size after completion of growth should raise suspicions of malignancy. Another important differential diagnosis is *periosteal osteosarcoma*. Here too, however, the history (pain), x-ray findings and enlargement of the tumor after completion of growth should point to the correct diagnosis. If *multiple lesions* are present, a hereditary disorder is involved (► Chapter 4.6.2.31). When such multiple lesions point towards the joints the condition is known as *metachondromatosis* (► Chapter 4.6.2.31). If »exostoses« are observed at the epiphysis, these are not osteochondromas, but probably a sign of *dysplasia epiphysealis hemimelica (Trevor's disease)* (► Chapter 4.6.2.31).

### Treatment, prognosis

If they do not cause any problems osteochondromas do not need to be removed. Resection is indicated only if pain or restricted movement is present. Patients occasionally request their removal for cosmetic reasons, but this operation should be performed only after completion of growth, since there is always a risk of recurrence before this time. Large lesions and lesions near the trunk (particularly on the pelvis and proximal femur) should be removed even if they do not cause any symptoms since they pose the greatest risk of developing a sarcoma. While no reliable information about the risk of malignant change is available in the literature, this risk is probably less than 1% for solitary osteochondromas.

### Enchondroma

#### ► Definition

Benign intraosseous tumor consisting of well differentiated cartilaginous tissue.

- **Synonym**: *Chondroma* (also includes periosteal and extraskeletal tumors)

### Occurrence, site

The enchondroma is a relatively common tumor. However, the diagnosis is rarely made before the end of growth.

Both sexes are affected with equal frequency. Two thirds of enchondromas are located in the small long bones of the *hand* [10]. They are much rarer in other bones, although the foot is a relatively typical site.

### Clinical features

The tumor usually remains asymptomatic and does not cause any pain. On the hand, a thickening of the affected phalanx is an indication of its presence. At all other sites, the enchondroma is usually discovered by chance. Pain may be a sign of malignant change.

— **Radiographic findings:** A relatively sharply defined osteolytic area without any significant marginal sclerosis is apparent. Slight, arch-shaped *erosions of the cortex* are not necessarily indicative of malignancy, in contrast with penetration through the cortex. A typical feature are the stippled *calcifications within the tumor* (■ Fig. 4.36; also ■ Fig. 3.522, ► Chapter 3.5.8). Enchondromas are relatively small tumors that grow very slowly. Rapid enlargement with purely osteolytic areas next to calcified areas may also be an indication of progression to malignancy.

If doubt exists about the diagnosis in a situation requiring treatment, an MRI scan is recommended. The cartilaginous tissue has a highly characteristic signal pattern, with relatively intense signals on the T2-weighted image and even more intense signals

on the proton-weighted image. The signals with T1-weighting are less intense than those for the fatty tissues (► Chapter 2.1.4.1). On the basis of this information, the cartilaginous character of the tumor can be diagnosed with a high degree of accuracy. Since enchondromas only show slight vascularization, they generally show low peripheral enhancement after the administration of contrast medium (gadolinium).

- **The histology** of the enchondroma varies according to its location. Enchondromas of the large long bones and trunk are usually lobular, consist of hyaline cartilage and possess a relatively low cell content. The cellularity is always much higher in the small tubular bones of the hands and feet and is not a sign of malignancy at this site. However, any incorporation of residual local cancellous or cortical bone fragments or soft tissue infiltration in long bones should raise suspicions of a sarcoma.
- **Differential diagnosis:** Enchondromas with small calcifications cannot be reliably differentiated from other osteolytic processes on the basis of a plain x-ray. An MRI scan will usually clarify the situation. Incipient *progression to a chondrosarcoma* must be detected in good time. Signs of such progression are provided not only by the radiographic findings, e.g. rapid growth and penetration of the cortex, but also by the clinical features.



■ Fig. 4.36. Enchondroma in the femoral shaft of a 20-year old female patient with typical stippled calcifications

### Treatment, prognosis

Provided an enchondroma remains asymptomatic, no treatment is required. Troublesome enchondromas on the hand can be removed by curettage and the resulting gap filled with a cancellous bone graft. Large enchondromas (>7 cm) in the long bones should also be carefully curetted out and burr drilled [7]. If signs of malignancy are present we proceed as follows: If there is a low probability that the tumor shows malignant change, we perform a complete curettage without prior biopsy and remove the whole tumor mass. Suspicious sites should be examined histologically, separately if possible, for subsequent correlation with the x-ray findings. If the suspicion of a chondrosarcoma is confirmed, we either perform a wide resection with bridging or, in case of a grade 1 sarcoma, wait and monitor the situation closely depending on the individual situation.

If there is a strong suspicion that a chondrosarcoma is involved, we proceed as described for a chondrosarcoma (► Chapter 4.5.3.2). The risk of malignant change is not reliably known for enchondroma, but is certainly higher than for an osteochondroma but lower than for enchondromatosis (Ollier's disease). The risk of malignant change for enchondromas with a diameter of >7 cm has been estimated as 5% [7]. However, such tumors are rarely observed before the age of 30.



### Periosteal chondroma

This is a variant of the chondroma that can develop beneath the periosteum at the insertions of tendons and ligaments. Periosteal chondromas are very rare, usually form in adolescence and occur slightly more frequently in boys than in girls. They are usually detected on clinical examination as they produce a palpable protuberance over the bone. The increased periosteal tension can also occasionally cause pain.

- **Radiological investigation** often reveals a bowl-shaped indentation of the cortex with distinct sclerosis. The MRI scan usually clarifies the situation, since the periosteally located tumor shows the signal pattern of cartilaginous tissue (▶ Chapter 4.5.5). Macroscopically the tumor appears glazed, white and lobed.
- **Histological examination** reveals a typical cartilaginous tumor that is more cellular than an enchondroma. Consequently, it can be difficult to differentiate between a periosteal and a juxtacortical chondrosarcoma. The tumor should be removed by marginal resection.

### Chondroblastoma

#### ➤ Definition

Benign cartilage-forming tumor that occurs in childhood or adolescence and that is usually located only in the epiphyses.

#### Occurrence, site

This tumor is fairly rare and occurs predominantly during the second decade of life. The male:female distribution is 2:1. A chondroblastoma is almost always located in the *epiphyses* [21] and rarely penetrates the epiphyseal plate.

#### Clinical features

Chondroblastomas generally cause *pain* which, although not particularly intense, tends to be almost constantly present. Chondroblastomas in the epiphyses near the knees can cause palpable swellings if they are off-center.

- **Radiographic findings:** These are usually exclusively epiphyseal, circular or oval osteolytic tumors with slight marginal sclerosis (■ Fig. 4.37, also ■ Fig. 3.246, ▶ Chapter 3.2.13 and ■ Fig. 3.338, ▶ Chapter 3.3.12). As a rule, the tumor is not very large, but can extend into the metaphysis. Calcifications are fairly rare. In view of its high cell content and abortive matrix formation, the tumor's signal pattern on the MRI scan is not wholly typical of cartilaginous tissue (▶ Chapter 2.1.4.1).
- **Histologically** the tumor consists of chondroblasts with oval nuclei embedded in a small amount of,



■ Fig. 4.37. AP x-ray and MRI scan of a large *chondroblastoma* in the medial femoral condyle of a 15-year-old boy

usually chondro-osteoid, matrix. The cells usually possess a wide, and occasionally slightly eosinophilic, cytoplasm with clearly discernible cell boundaries that often look as if they have been drawn in with a fine pencil. Giant cells are always present.

- For this reason, the most important differential diagnosis is a giant cell tumor, which does not usually occur before the age of 18 and which contains far more giant cells with more numerous nuclei (>20/sectional plane). This tumor is also relatively clearly demarcated and located in the epiphyses and metaphyses, although it only extends towards the epiphyses after completion of growth, remaining mainly in the metaphyses while the epiphyseal plates are still open [30]. Other important differential diagnoses are the *clear cell chondrosarcoma*, which also tends to be located in the epiphyses [27], and the *classical chondrosarcoma*. Both tumors tend to occur in adulthood and grow faster and more expansively than the chondroblastoma, i.e. usually without marginal sclerosis. Another important possibility to consider in the differential diagnosis of chondroblastoma of the *femoral head* is *Legg-Calvé-Perthes disease* (▶ Chapter 3.2.5). Since the collapsing of the femoral head is not a feature of chondroblastoma, the distinction is readily possible, but mix-ups still occur.

#### Prognosis, treatment

Although a chondroblastoma is a benign tumor, metastases have been reported in isolated cases [52]. Growth disorders can occur if the chondroblastoma penetrates through the epiphyseal plate. Because of its epiphyseal location, a wide or en-bloc resection of this locally expansively growing tumor is not usually possible. Accordingly, a complete curettage with burring of the tumor wall is indicated, a procedure that reduces the recurrence rate to below 10% (in contrast with rates of up to 50% after a standard curettage).



### Chondromyxoid fibroma

This rare tumor occurs mainly in males between the ages of 10 and 30.

- **Site:** The tumor always develops eccentrically in the *metaphysis* and is almost exclusively located in the lower extremity, particularly in the proximal tibia.
- **Clinical features:** Since the tumor grows very slowly in parallel with the longitudinal axis of the affected bone, symptoms such as pain or swellings are rare.
- **Radiographic findings:** An oval osteolytic area surrounded by sclerosing is visible in the vicinity of the metaphysis. A multilocular tumor with septum-like internal demarcations develops at a later stage. The MRI scan shows a signal pattern that is typical of cartilage tissue (▶ Chapter 2.1.4.1).
- **Histologically** the cartilaginous-myxomatous tumor has a lobular structure subdivided by fine, connective tissue-like, vessel-containing septa with compacted tumor cells at the lobular margins and in the vicinity of the septa, and always including giant cells.
- The most important differential diagnosis is *chondrosarcoma*. The polycyclic appearance of the tumor is highly characteristic. As long as the tumor appears to be unilocular, confusion with a giant cell tumor is possible, although the metaphyseal location argues against this possibility.
- **Treatment:** Since the tumor is usually located in the metaphyses, an en-bloc resection is usually possible without major problems. Nor does bridging pose any difficulties. A wide, or at least marginal, resection offers the best guarantee of freedom from recurrences. While an intralesional curettage is possible in exceptional cases, it must be performed very carefully in view of the risk of recurrence. Isolated cases of malignant change have been reported [10].

#### 4.5.2.3 Connective tissue tumors (benign fibrous histiocytoma, desmoplastic fibroma)

##### Benign fibrous histiocytoma

Very rare fibrohistiocytic tumor in the epiphyses and diaphyses of long bones, although it can also occur in the ribs, the pelvis and the clavicle. When the metaphysis is involved, the diaphysis or metaphysis is also invariably affected. This very rare tumor can occur at almost any age and is always painful.

- **Radiologically** the tumor appears lobulated, mainly well demarcated and often partially surrounded by a sclerotic border. The bone scan usually shows increased activity since this tumor grows faster than a fibrous metaphyseal defect [19].
- **Histologically** the structure of the tumor is reminiscent of a non-ossifying bone fibroma (NOF, see chapter on tumorlike lesions below) and indistinguishable from

the latter under the microscope, although it is more active and larger. The diagnosis should therefore be made only in the knowledge of the x-ray findings or the location, which is atypical of a NOF (e.g. pelvis). Since the tumor can recur if the *curettage* is not performed correctly, a complete curettage with burring of the tumor walls is required.

##### Desmoplastic fibroma

This is an intramedullary tumor that corresponds histologically to an aggressive fibromatosis of the soft tissues (▶ Chapter 4.5.4.1). It shows aggressive local growth, can grow very large and frequently recurs after curettage. The tumor does not affect one sex more than the other, usually occurs between the ages of 10 and 30 [10] and has been observed in all long bones, the mandible and the spine [3].

- **Clinically** this slow growing tumor causes few symptoms. Pain occurs occasionally, and the diagnosis is often made only after a pathological fracture.
- **Radiologically**, a geographically configured area of osteolysis, which can cause the bone to swell up considerably, surrounded by a thin sclerotic border. The tumor has a polycyclic margin and may be trabeculated by residual bone projections.
- **Radiologically**, a low-grade malignant fibrosarcoma, in particular, can be confused with a desmoplastic fibroma, which itself does not show any major cell polymorphism or nuclear hyperchromasia on histology. Some lesions, like aggressive fibromatosis, will show chromosome aberrations (trisomies 8 and 20) [6]. There are also microscopic similarities with fibrous dysplasia and the highly differentiated central osteosarcoma.
- **Differential diagnosis:** The tumor must be differentiated radiologically from a *benign fibrous histiocytoma*, a well differentiated *fibrosarcoma* and a *chondromyxoid fibroma*.
- Curettage is not sufficient as *treatment*, since it leads to recurrence in over 50% of cases. The minimum requirement, therefore, is a *marginal*, or preferably a *wide en-bloc resection*. Large tumors requiring corresponding defect bridging are usually involved (▶ Chapter 4.5.5). Malignant change has not been described.

#### 4.5.2.4 Vascular tumors

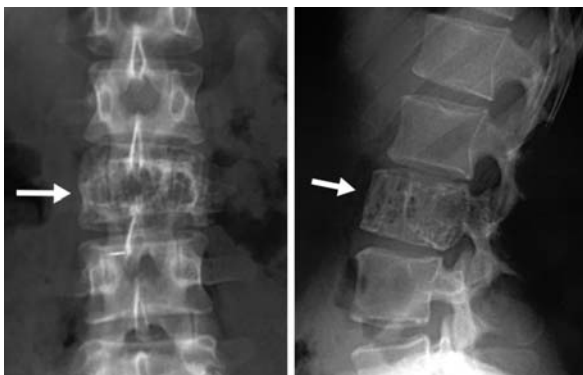
##### Hemangioma

Intraosseous hemangiomas consist of accumulations of differently structured (capillary/cavernous) blood vessels. They are very common in the spine, but extremely rare in the long bones. Hemangiomas in the spine invariably remain asymptomatic, and they are almost always painless when they occur in the long bones.

- *Radiologically*, hemangiomas appear as osteolytic processes, often surrounded by a sclerotic border, with areas of intralesional, straggly-striated condensation. They can also be very variable in terms of size and appearance (■ Fig. 4.38, 4.39).
- *Histologically*, most hemangiomas consist of cavernous blood spaces lined with normal-looking, occasionally rather prominent and »epithelioid«, endothelium. Mixed forms with capillary sections can occur.
- *Differential diagnosis*: Diagnosing a hemangioma solely on the basis of imaging investigations is very difficult and usually requires a biopsy or resection. Given its variable appearance, *most osteolytic processes* (including *metastases*) must be included in the differential diagnosis.
- *Treatment*: Asymptomatic hemangiomas do *not necessarily require treatment*. Large and/or symptomatic hemangiomas can be resected, although they can recur if the resection is insufficient. Radiotherapy or embolization should be considered for very large progressive hemangiomas.



■ Fig. 4.38. Conventional tomogram of a 19-year old female patient with *hemangioma* in the area of the lateral tibial condyle



■ Fig. 4.39. AP and lateral x-rays of the lumbar spine of a 17-year old girl with a *hemangioma* in vertebral body L3.

#### 4.5.2.5 Nerve tissue tumors

##### *Schwannoma and neurofibroma*

While neurofibromas in bone almost always occur in connection with neurofibromatosis (► Chapter 4.6.6.2), intraosseous schwannomas (neurilemmomas, schwannomas) are observed, in rare cases, as solitary tumors, particularly in the mandible or sacrum. They can occur in the periosteum, where they lead to a lenticular excavation of the cortex. Round, osteolytic foci can also be observed centrally. Schwannomas can cause persistent, diffuse pain. They grow slowly. The recurrence rate after removal is low.

#### 4.5.2.6 Giant cell tumor

##### *Giant cell tumor*

##### 🔴 Definition

A locally aggressive tumor that occurs in the metaphysis in the area of the (former) epiphyseal plate and spreads towards the epiphysis. It contains accumulations of macrophages and large amounts of uniformly distributed osteoclastic giant cells with numerous nuclei, intermixed with mononuclear ovoid cells (forming the actual tumor component). In view of its aggressive nature and unpredictable course, the giant cell tumor is classified by the WHO as an »intermediate grade« tumor between benign and malignant tumors [50].

##### Occurrence

The tumor is relatively common in adulthood, usually occurring between the ages of 20 and 40. Females are slightly more frequently than males. The tumor also occurs, in rare instances, during adolescence while the epiphyseal plates are still open. In the USA the giant cell tumor accounts for 5% of all bone tumors, compared to 20% in China [16].

##### Site, pathogenesis

The typical site of the tumor is *epiphyseal or epiphyseal/metaphyseal* area. If the epiphyseal plates are still open, it usually remains limited to the metaphysis [30]. By far the most commonly affected sites are the *distal femur* and the *proximal tibia*. Less frequently, the tumor occurs in the proximal femur, distal tibia, proximal humerus and distal radius. While it can form in a vertebral body, the most commonly affected site in the spine is the sacrum [31]. In isolated cases the lesion is also found in flat bones, e.g. the pelvis or base of the skull. The tumor is predominantly located in bones with the most active growth plates.

##### Clinical features

Giant cell tumors are *painful* and can lead to swollen joints and movement restrictions.

- **Radiologically**, giant cell tumors are eccentrically positioned osteolytic lesions without partitions. They show epiphyseal / metaphyseal spread and are usually well demarcated. An actual sclerotic border is normally absent, as is a periosteal reaction. The tumors can extend to the joint subchondrally and occasionally possess a bulging neocortex. Not infrequently, they penetrate the compact bone. During adulthood, the tumors can also spread to the diaphysis [53] (Examples of giant cell tumors can also be found in ► Chapter 3.3.12, ■ Fig. 3.339, 3.343). The bone scan usually shows a relatively pronounced uptake. The *MRI* signal of the tumor is low in all weighted images.
- **Histology**: Although the accumulations of giant cells, which are usually distributed uniformly in the lesion, are diagnostic pointers, the actual proliferating tumor cell component consists of the intervening misshapen or spindle-shaped, oval mononuclear cells. These probably originate from osteoblast precursor cells that have lost the ability to mature into osteoblasts, but with the common property of secreting osteoclast-attracting factors, e.g. RANKL (receptor activator of nuclear factor kappa-ligand). This would explain the high content of giant cells and also the abortive formation of new osteoid and – usually occurring to a lesser extent – bone that is observed in almost half of all giant cell tumors.
- **Differential diagnosis**: The giant cell tumor must be differentiated from a *chondroblastoma*, which is also osteolytic and involves the epiphysis. During childhood and adolescence, however, the topography

usually provides a clear diagnostic indication. The chondroblastoma is almost invariably confined to the epiphysis and possesses a sclerotic border, while the giant cell tumor is always located in the metaphysis as well. If a metaphyseal-epiphyseal location and more aggressive radiological findings are present, the possibility of an *osteosarcoma* must also be considered, although the latter additionally shows permeating bone destruction, focal, cloudy areas of matrix calcification and a periosteal reaction. If pop-corn like matrix-mineralizations are absent, *chondrosarcomas* can be confused with giant cell tumors in the advanced stage, but show a signal pattern that is typical of cartilage on the *MRI* scan.

*Aneurysmal bone cysts* may also have a similar appearance, although fluid levels will be present on the *MRI* scan. However, aneurysmal bone cysts can occur secondarily to giant cell tumors.

#### **Treatment, prognosis**

The tumor can behave in widely differing ways and cannot be predicted on the basis of radiological, histological, or flow cytophotometric criteria [47]. It can grow in a weakly or strongly expansive manner or even behave aggressively. It can turn into a sarcoma without transformation and metastasize (primary malignant giant cell tumor: giant cell tumor in addition to a sarcoma component) [55] or transform itself into a malignant tumor following several recurrences and radiotherapy (secondary malignant giant cell tumor, giant cell tumor component no longer usually detectable), which occurs in 1% of cases [2, 20].



■ **Fig. 4.40a–c.** 5-year old girl with large aneurysmal bone cyst in the proximal radius. **a** X-ray of the proximal upper arm. **b, c** Coronal and sagittal *MRI* sections. Note the levels formed within the fluid accumulations

The degree of differentiation, local aggressiveness and risk of malignant change do not correlate with each other. A standard intralesional *curettage* is associated with a high risk of recurrence of up to 70%, while an en-bloc resection can lead to major defects and difficult reconstruction problems as a result of the proximity of these tumors to joint. For these reasons, the use of necrotizing agents is recommended (phenol, methyl methacrylate, liquid nitrogen).

Even these substances, however, are not without their own problems when used in large cavities near joints. The recurrence rate is primarily dependent on the quality of the curettage. We perform the curettage through a large incision, burr drill the whole cavity with a *special drill* that can drill »round corners«, remove the septa and then examine the whole tumor cavity with the *arthroscope*. The recurrence rate after this procedure is low (less than 10%).

We usually manage to dispense with the use of phenol or methyl methacrylate and thus avoid damaging the joint cartilage. Leaving a large plug of methyl methacrylate in place can cause problems in adolescents as the plug, particularly in the distal femur, tends to migrate towards the joint and can cause damage at a later date. We therefore fill the cavity with autologous and/or homologous cancellous bone, sometimes using an *allograft* (■ Fig. 3.339). On the distal radius, replacement with an allograft or the proximal fibula may be possible.

#### 4.5.2.7 Tumor-like lesions (pseudocysts, non-ossifying bone fibroma, fibrous dysplasia, histiocytosis, infarct)

##### Simple bone cyst

###### ➤ Definition

Pseudocystic, osteolytic lesion occurring predominantly during childhood and adolescence, filled with serous fluid and located centrally in the metaphyses of long bones. Cause unknown, probably atrophic-degenerative, not an actual tumor.

— **Synonyms:** Juvenile bone cyst, unicameral bone cyst, solitary bone cyst

##### Occurrence, site

The simple bone cyst is common during childhood. After the non-ossifying bone fibroma and the osteochondroma, it ranks third among all primary bone lesions (male: female ratio = 2:1). The lesion almost always occurs between the ages of 5 and 15 and heals on completion of growth. The most susceptible site is the proximal humerus, where almost 50% of all simple bone cysts are located. The proximal femur is involved in approx. 25% of cases. The remaining approx. 25% are distributed evenly over the whole skeleton.

##### Clinical features

We distinguish between inactive (latent) and active cysts. The *inactive cysts* migrate towards the diaphysis as the bone grows longer. The *active cysts* are immediately adjacent to, but never cross, the epiphyseal plate. They hardly cause any signs or symptoms as they are not palpable beneath the large soft tissue masses over the proximal humerus or proximal femur. Not infrequently, however, they can lead to a pathological fracture, at which point the diagnosis is then confirmed.

- **Radiographic findings:** In the metaphysis (usually immediately adjacent to the epiphyseal plate) there is an area of unilocular or multilocular osteolysis, often with septations, that causes the bone to swell slightly and symmetrically. The cyst wall may be paper-thin and fragile. Isolated bone fragments can occasionally be seen in the cyst and are the result of a fracture of intralesional bony septa (»fallen fragment«; for examples of radiographic findings of simple bone cysts see ■ Fig. 3.253, ► Chapter 3.2.13, and ■ Fig. 3.521, ► Chapter 3.5.8).
- Since, in *histological* terms, the wall is not lined with epithelium, this tumor should, correctly-speaking, be described as a »pseudocyst«. The tissue consists of thin, slightly collagenized and vascularized connective tissue septa, which show slight new bone formation and contain a few giant cells.
- **Differential diagnosis:** It can sometimes be difficult to distinguish between the simple bone cyst and an *aneurysmal bone cyst (ABC)*, and active cysts in particular can appear similar. However, the ABC almost always produces an eccentric swelling of the bone. The MRI scan does not always clarify matters, since fluid levels can occur with both lesions. *Fibrous dysplasia* can also lead to similar swellings in the metaphyseal area. In such cases, the ground-glass opacity on the x-ray serves as a crucial pointer to the diagnosis. Moreover, fibrous tumors can show relatively high signal levels (resembling fluid) on the MRI scan. A *Langerhans cell histiocytosis*, *non-ossifying bone fibroma* and *enchondroma* can likewise occasionally be confused with a simple bone cyst.

##### Treatment, prognosis

Fractures usually heal without any problems at all. Not infrequently, the cyst disappears following a fracture. Recurrences are common after curettage and cancellous bone grafting for active cysts near growth plates. Cortisone administration [42, 48] seems to be more successful, although recurrences are still possible, particularly with polycystic lesions close to the epiphyseal plate. The cyst is aspirated using two large cannulas. Under image intensifier control, the cortex is pierced and the cyst visualized using contrast medium; 50–100 mg of methylprednisolone acetate are then injected. The injection of bone mar-



row also appears promising [32]. For cysts in the proximal femur we reinforce the bone using Prévot nails in order to avoid any fractures, which would otherwise be difficult to stabilize at this site. The cysts usually heal up after this nailing procedure.

### **Aneurysmal bone cyst (ABC)**

#### **Definition**

A lesion that grows expansively and eccentrically, with unknown, and probably non-uniform, etiology. Since aneurysmal bone cysts can also develop from other benign or malignant bone tumors, very careful histological examination and correlation with the radiographic findings is always essential. In addition to the usual pseudocystic form, a solid variant of this lesion also exists [35].

#### **Occurrence, site, pathogenesis**

Aneurysmal bone cysts are fairly rare, occurring only half as frequently as the giant cell tumor for example. Both sexes are affected with roughly equal frequency. They tend to occur between the ages of 10 and 20, and are hardly observed at all after the age of 30. The cysts are most often found in the metaphyses of long bones, particularly in the knee area and the spine, usually in the area of the vertebral arches or processes. At this site the aneurysmal bone cyst is, after osteoblastoma, the most common benign lesion that occurs during adolescence. It can affect several vertebrae. In the long bones, the lesion is never located primarily in the epiphysis, although it spreads into the epiphysis by breaking through the epiphyseal plate [10]. The ABC shows a characteristic translocation  $t(16;17)(q22;p13)$  that has been detected in classic ABCs, as well as in solid and extraosseous variants [11, 41]. The translocation  $t(16;17)(q22;p13)$  has been shown to lead to upregulation of the USP6 protease, which is probably involved in the invasion [36, 37]. However, the absence of this translocation in secondary ABCs suggests that the etiology of secondary (reactive) lesions differs from that of genuine lesions [38].

#### **Clinical features**

Aneurysmal bone cysts spread rapidly and can therefore be painful. The lesions are usually located eccentrically and tend to break out of the bone and expand. This leads to palpable swellings and protuberances.

— **Radiographic findings:** The ABC is an eccentrically located, metaphyseal osteolytic lesion with minimal marginal sclerosing or septum formation. The bone is blown up like a balloon and is often paper-thin, resembling a soap bubble (■ Fig. 4.40, also ■ Fig. 3.121, ► Chapter 3.1.14, and ■ Fig. 3.339 and 3.340, ► Chapter 3.12). The cortex is frequently replaced by an indented neocortex. Usually, however, it shows a distinct border, though this may occasionally just

be vaguely discernible, or even completely extinguished, on a conventional x-ray. In contrast with malignant tumors, the lesion is sharply defined in relation to the medullary cavity. Periosteal reactions are also occasionally observed. The MRI scan shows typical fluid levels, which are signal-intensive in the T2-weighted images. Fairly large, more solid areas are lacking in aneurysmal bone cysts. If these are observed, the possibility of a teleangiectatic osteosarcoma should be considered in the differential diagnosis.

- The macroscopic and *microscopic picture* is characterized by large, blood-filled, septated cavernous cavities, whose walls are lined with flattened pseudo-endothelial covering cells and giant cells. The septa are of differing thicknesses and contain loosely-arranged spindle-shaped fibroblasts with typical mitoses, macrophages, lymphoid infiltrates, capillary proliferations, collagenization and the formation of immature fibrous bone. In the solid variant, the cavities fade completely into the background, leaving the impression of a compact tumor.
- **Differential diagnosis:** In view of the location and radiographic findings, an ABC can be confused with a *giant cell tumor*, *simple bone cyst*, *chondromyxoid fibroma*, but also with a *teleangiectatic osteosarcoma*. The solid variant must be differentiated primarily from a conventional intramedullary osteosarcoma. Aneurysmal bone cysts are also frequently a secondary component of other tumors (e.g. giant cell tumors), producing mixed pictures [9, 54].

#### **Treatment, prognosis**

Aneurysmal bone cysts usually grow expansively and can reach a considerable size, although individual cases of spontaneous healing have been described [46]. As a rule, however, surgical treatment is required. Freedom from recurrence can be achieved reliably only by at least a marginal, or preferably wide, resection. After curettage there is a high risk of recurrence, but this can be reduced through the use of necrotizing agents (phenol, methyl methacrylate, liquid nitrogen). Since aneurysmal bone cysts rarely extend into the epiphysis, en-bloc resection is usually preferable.

If, in the event of a recurrence, resection of the complete tumor is not possible, embolization of the supplying blood vessels may produce a cure [60]. A biopsy should always be taken before the resection since aneurysmal bone cysts can develop from another tumor. It is perfectly possible for a malignant tumor to be concealed beneath the picture of an aneurysmal bone cyst, and even then rarely be differentiated from an ABC at a later stage. At any rate, the therapeutic consequences would be minimal if a wide resection is performed, whereas a curettage would be the wrong treatment in such cases.



### Non-ossifying bone fibroma (NOF)

#### ➤ Definition

Metaphyseal lesion consisting of fibroblasts, histiocytes, giant cells and scanty lymphocytic infiltrates, and occurring at the insertions of tendons or ligaments in the epiphyseal plate. They sclerose or disappear after completion of growth.

- **Synonyms:** Histiocytic fibroma, fibroma, fibrous cortical defect, fibrous xanthoma, histiocytic xanthogranuloma

#### Occurrence

The non-ossifying bone fibroma is extremely common, particularly in the area of the *metaphyses near the knee*. In fact, it is the *most common lesion in bone*, and is observed in 20–30% of all children between the ages of 4 and 10 years [10]. Both sexes are affected with equal frequency. On completion of growth, the fibromas either disappear or remain visible as sclerosed zones.

#### Site, pathogenesis

The non-ossifying bone fibromas are always located in the *metaphyses*, especially those near the knee, but also in the distal tibial metaphysis. Their etiology is unclear, although traumatic factors and overloading of the insertion sites of tendons and ligaments, combined with a local malfunction, probably play a role [43]. The lesions subsequently migrate in the direction of the diaphysis during the course of growth until they are remodeled by normal bone turnover processes. They are always located eccentrically near the cortex.

#### Clinical features, diagnosis

Non-ossifying bone fibromas are completely *asymptomatic*. They are usually discovered by chance on an x-ray. In very rare instances, the cortex may cave in, resulting in lesion-induced pain. Exceptionally, very large fibromas can cause the bone to swell up and lead to a palpable thickening.

- The *radiographic findings* are so characteristic that the diagnosis can be reliably confirmed on the basis of plain x-rays, provided the tumor does not exceed a certain size. The x-ray reveals polycyclic, grape-shaped, relatively well-defined, defects surrounded by a clear sclerotic border (■ Fig. 4.41 and ■ Fig. 3.337, ► Chapter 3.3.12). The defect is aligned lengthwise with the bone and is often centered over the cortex (fibrous cortical defect). But it frequently also affects the medullary cavity (non-ossifying bone fibroma), when it is always located off-center. The cortex may bulge outward slightly. Provided the focus is small and takes up less than two-thirds of the bone width, no further investigation is required.

- *Histologically* the lesion consists of fibroblasts without atypia arranged in intertwining bundles and forming moderate amounts of collagen. Scattered among these cells are histiocytes, foam cells and a few giant cells.
- *Differential diagnosis:* If the foci are small there is usually no doubt about the diagnosis. However, confusion with a *desmoplastic fibroma* is possible, and a *chondromyxoid fibroma* can produce a similar picture.

#### Treatment, prognosis

An asymptomatic non-ossifying bone fibroma *does not require any treatment*. It disappears either spontaneously or leaves a residual ossified scar. Only very large, potentially destabilizing, fibromas will require, at most, curettage and filling with cancellous bone. Biopsy is required only very rarely in cases where the presence of pain cannot be fully explained.

### Monostotic fibrous dysplasia

#### ➤ Definition

Benign, fibrous bone-forming lesion without cuboid osteoblast seams. A distinction is made between polyostotic (► Chapter 4.6.2.31) and monostotic forms.

#### Occurrence, site, pathogenesis

Monostotic fibrous dysplasia is commoner than supposed, since many cases progress without symptoms. It is also commoner than polyostotic fibrous dysplasia, which accounts for 20% of cases [14], and has a prevalence of 2.6



■ Fig. 4.41. X-rays of a non-ossifying bone fibroma of the proximal fibula of an 11-year old boy

per million inhabitants. A McCune-Albright syndrome exists only in approx. 5% of patients with fibrous dysplasia (polyostotic form and McCune-Albright syndrome; ► Chapter 4.6.2.31). Females are slightly more frequently affected than males. The disease usually manifests itself during adolescence. Most commonly, the monostotic form affects the *jaw* and *proximal femur*, and occurs rather less frequently in the tibia, humerus, ribs, radius and iliac crest. The condition is thought to be caused by a mutation in a gene that codes for a membrane-bound signal protein (GS- $\alpha$ ). The manifestation of the clinical picture (McCune-Albright syndrome, polyostotic or monostotic fibrous dysplasia) depends on the time at which the mutation occurs [1].

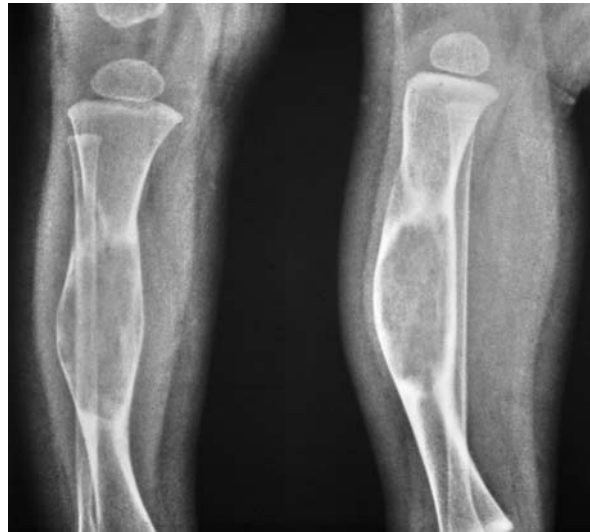
### Clinical features

The monostotic forms often remain asymptomatic. If the affected bone is covered only by a thin layer of soft tissue, a bulge may be palpable. Bowing or axial deviation of the bone may also be visible (■ Fig. 4.42). A very typical finding is bowing of the proximal femur in the shape of a shepherd's crook (■ Fig. 3.245, ► Chapter 3.2.13). Pain occurs only if fractures are present, or occasionally during periods of overexertion.

- The *radiological appearance* on the plain x-ray, and also on CT images, is very typical. The bone appears distended and the cortex thinner than normal. In the medullary cavity there is a large osteolytic area interwoven with bone trabeculae (under magnification), producing a characteristic ground-glass opacity. Occasionally pronounced sclerosis is visible around the focus. On the *MRI scans* the tissue signal is low in all weightings.
- *Histologically* the picture is dominated by irregularly shaped fibrous trabeculae embedded in a moderately cell-rich fibrous stroma. The trabeculae show flattened cells on the surface rather than cuboid osteoblasts.
- *Differential diagnosis*: Since the x-ray findings are usually very typical, the diagnosis poses no problems. In the initial stages (particularly in relation to the proximal femur or humerus), the condition may be confused with a *simple bone cyst*, in which the cortex is also thinner than normal and the bone is distended by osteolysis. However, the frosted-glass opacity and bowing are both absent. A fracture may be apparent. On the lower leg monostotic fibrous dysplasia can be confused with an *osteofibrous dysplasia* (see below), although the latter almost always affects the tibia alone and shows osteolytic-sclerotic changes in the cortical bone.

### Treatment, prognosis

Provided no major deformation is present, *surgical treatment is not usually required*. Nor does a biopsy need to be



■ Fig. 4.42. Monostotic fibrous dysplasia in the area of the right tibia of a 2-year old boy

taken if the diagnosis is clear. An operation may be appropriate if pronounced bowing is present, which particularly occurs in the proximal femur and when very large foci are involved. Since it can be difficult to obtain sufficient autologous cancellous bone to fill the gap, homologous cancellous bone or hydroxyapatite can also be used. Reinforcement with an intramedullary load-bearing implant, e.g. Prévot nails can also be useful. For the proximal femur, an intertrochanteric valgus osteotomy and stabilization with a gamma-nail is appropriate. In children with an open epiphyseal plate we use a telescopic gamma-nail developed by ourselves specifically for this purpose. The sleeve and the nail are inserted from the greater trochanter and the nail can be transfixed at the distal epiphysis with a screw. The telescopic gamma-nail is also useful in osteogenesis imperfecta.

The progression of the condition is usually halted on completion of growth. *Malignant change* can occur. In a sample of approx. 1,000 patients with fibrous dysplasia, sarcomas were observed in 28, half of whom had already received radiotherapy [45].

### Osteofibrous dysplasia (according to Campanacci)

#### ► Definition

Congenital, probably hamartomatous, predominantly intracortical lesion consisting of osteofibrous tissue, almost invariably located in the tibia, rarely in the fibula and with typical anterior bowing that progresses during growth. The condition was described in 1976 by Campanacci [8].

- *Synonyms*: Congenital fibrous defect of the tibia, Campanacci's disease, ossifying fibroma

**Occurrence, site**

The lesion is rarer than fibrous dysplasia and does not show any GS- $\alpha$  mutation. The male sex is more frequently affected. The disease usually manifests itself within the first five years of life and occurs almost exclusively in the *tibia*, and only rarely in the fibula. It tends to start in mid-shaft and then spread distally or proximally.

**Clinical features**

Osteofibrous dysplasia does not cause any pain, but does produce visible anterior bowing of the tibia. The surface of the tibia often appears uneven with bumps. Not infrequently, a pathological – possibly incomplete – fracture can occur. Recovery can sometimes be problematic, and progression can occur after fractures or surgical procedures.

- On the x-ray alternating areas of osteolysis next to sclerosis are visible in the cortical bone. The foci are located not in the medullary cavity of the bone but in the cortex, which gradually bends and may show microfractures. The picture is also characterized by remodeling processes and callus formation (■ Fig. 3.341, ► Chapter 3.3.12).
- *Histology*: In contrast with fibrous dysplasia, the immature bone trabeculae, which are likewise embedded in a fibrous stroma, are occupied by cuboid osteoblasts. The lesions are structured in zones with a centrally dominating fibroblast section, while the width of the trabeculae and their maturation into lamellar bone increases towards the periphery.
- The most important differential diagnosis is an *adamantinoma*, a low-grade malignant tumor that almost always occurs in the tibia and typically shows intralesional epithelial cell islands (► Chapter 4.5.3.6) [26]. While the radiological appearance is similar, the adamantinoma is always located in the medullary cavity, in contrast with osteofibrous dysplasia. Both lesions can also occur next to each other [4]. A possible connection between an adamantinoma and osteofibrous dysplasia has been discussed [56], but has not been proven to date. The disease must also be differentiated from *fibrous dysplasia*, which is located in the medullary cavity. Apart from a slight narrowing, the latter does not show any cortical alterations and also shows a much more uniform radiological picture, with the typical frosted-glass opacity.

**Treatment, prognosis**

The course of the disease varies considerably. Some lesions stop spreading even before puberty, while others continue expanding until growth is complete. Operations should be avoided during the first 10 years of life, as these tend to promote the spread of the lesions. A high

recurrence rate is observed after curettage procedures. Fractures should be treated conservatively. Surgery (if possible only after the completion of growth) is indicated if the bone is greatly weakened, or if substantial bowing or pseudarthrosis are present. If the x-rays raise doubts about the possibility of an adamantinoma (intramedullary involvement!), a representative biopsy must be taken. As a rule, osteofibrous dysplasia does not cause any major problems in adulthood.

**Langerhans cell histiocytosis****Definition**

Clonal, possibly neoplastic, proliferation of Langerhans cells with activation of lymphocytes, eosinophils, macrophages, multinuclear giant cells and Langerhans cells. Langerhans cell histiocytosis also occurs in connection with Hand-Schüller-Christian disease and Abt-Letterer-Siwe disease.

- *Synonyms*: Histiocytosis X, eosinophilic granuloma

**Occurrence, site**

Langerhans cell histiocytosis is a rare condition that occurs primarily in the first two decades of life. Boys are twice as frequently affected as girls. The disease can be found in all bones. It is especially common in the mandible and skull, but can also affect all long bones, the ribs, the spine and the flat bones [22].

**Classification**

*Langerhans cell histiocytosis* occurs in the following forms:

- monostotic form,
- polyostotic form,
- polyostotic form with visceral involvement,
- Hand-Schüller-Christian disease: Combination of eosinophilic granulomas, diabetes insipidus and exophthalmos,
- Abt-Letterer-Siwe disease: Malignant (fatal) form of Langerhans cell histiocytosis.

**Etiology**

This condition probably involves a dysfunction of the immune system [22]. Changes in the thymus also appear to play a certain role, as do genetic aspects [18, 58]. Recent studies have shown a clonal proliferation of the Langerhans cells, which suggests that it may be a neoplastic process with a high degree of variability in its biological behavior [59].

**Clinical features**

The signs and symptoms differ considerably depending on the site involved. In addition to benignly progressing forms with solitary and multiple bone foci, highly malignant, potentially fatal, forms can also occur. Visceral involvement is a key factor, particularly in small children.

#### 4.5.2 · Benign bone tumors and tumor-like lesions

Among our own patients we have observed 93 mono-stotic and 24 polyostotic forms, 5 cases with visceral involvement, 7 patients with Hand-Schüller-Christian disease and one with Abt-Letterer-Siwe disease [22]. The older the child at the first appearance of the disease and the less soft tissue involvement, the better the prognosis [5, 15, 17].

— The *radiographic findings* are highly variable (■ Fig. 4.43). If Langerhans cell histiocytosis is suspected, a bone scan should always be arranged in order to establish whether several foci are present. An MRI scan should also form part of the investigations and typically shows low signal intensity in T1-weighted images and high signal intensity in T2-weighted images. However, no imaging procedure can confirm the diagnosis with complete certainty. Also very typical are foci in the area of the vertebral bodies, which result in collapsing of the vertebral bodies and a clinical picture of *vertebra plana* (■ Fig. 3.122 and 3.123, ► Chapter 3.1.14).

However, neurological lesions are extremely rare despite this impressive collapsing process since they do not produce kyphosing and the lesion itself is soft, and not solid, and does not therefore press against the spinal cord. A typical feature in a patient with extensive involvement is a »map-like skull«, particularly in Hand-Schüller-Christian disease (■ Fig. 4.44).

— *Histology*: The crucial finding under a light microscope is the appearance of Langerhans cells, which can even be detected with HE staining thanks to their typical morphology (well-defined, slightly eosinophilic cytoplasm, medium-sized, indented or notched, occasionally gyri-form nuclei). The Langerhans cells can be interspersed in granulomatous nests with differing quantities of eosinophils, in some cases in clusters. Immunohistochemical investigations (positive reactions with antibodies against CD1a and S100 protein) are recommended to confirm the diagnosis [22]. Electron microscopic examination reveals the Birbeck granules that are typically seen in Langerhans cells.

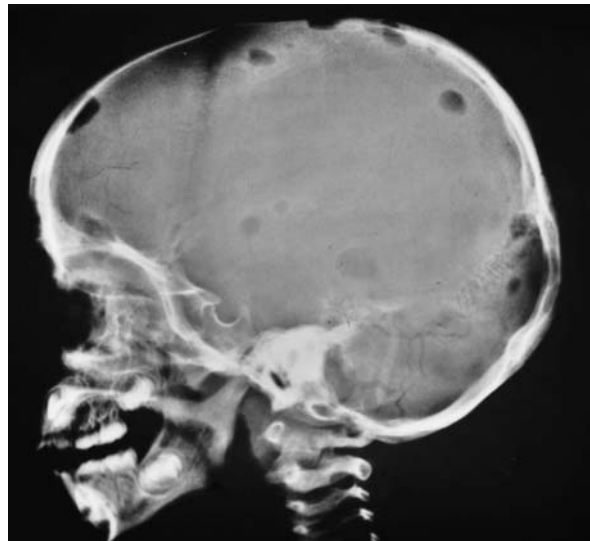
— *Differential diagnosis*: Many other tumors, including malignant ones (e.g. *Ewing sarcoma*), produce similar images. The polyostotic involvement may indicate the presence of a Langerhans cell histiocytosis, but the diagnosis can be confirmed only with an – ideally open – biopsy.

#### **Treatment, prognosis**

Most cases of osseous Langerhans cell histiocytosis heal spontaneously after the biopsy [15]. They are rarely progressive and an intralesional curettage is always sufficient in the initial stages. Stabilization is required only if bone strength is jeopardized. If the vertebral bodies are affected we stabilize the spine with a corset. Although



■ Fig. 4.43. Axial x-ray of the proximal humerus of a 15-year old boy with *Langerhans cell histiocytosis*. Note the pronounced periosteal reaction and the erosion of the cortical bone, a finding that can resemble a malignant bone tumor



■ Fig. 4.44. Skull x-ray of a 2-year old boy with *polyostotic Langerhans cell histiocytosis*, so-called »map-like skull«

radiotherapy is also recommended by some authors, we do not consider it necessary if bone is exclusively involved. Chemotherapy is indicated in cases of visceral involvement and is also a possibility with polyostotic involvement. It is not necessary, however, if just 1–3 foci are present. Bisphosphonates have also been used recently in such cases [29].



In the overwhelming majority of cases the course of Langerhans cell histiocytosis is very benign, although problems can occur at a later stage and are more common in patients with multifocal involvement. For this reasons, follow-ups should continue, in some cases for more than 10 years.

### Bone infarcts

Intraosseous necroses in the metaphyseal (more rarely the diaphyseal) section of long bones. They occur particularly in the elderly, frequently without any clinical signs and symptoms. They can produce calcification at a later stage and are sometimes difficult to differentiate from enchondromas.

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### 4.5.3 · Malignant bone tumors

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### 4.5.3 Malignant bone tumors

*F. Hefti, G. Jundt*

#### 4.5.3.1 Bone matrix-forming tumors (osteosarcomas)

##### Conventional osteosarcoma

##### ► Definition

High-grade malignant tumor with direct formation of bone ground substance (osteoid) by the tumor cells. The tumor occurs between the ages of 15 and 25 and is – apart from the plasmacytoma, which is classed as a hematological condition – the most common primary malignant bone tumor.

##### Occurrence

The osteosarcoma is the most common solid malignant bone tumor. The annual incidence is approx. 4–5 cases per million inhabitants (WHO 2000), 60% of which occur before the age of 25. ■ Fig. 4.28 shows the age distribution among our own registered patients. Studies have shown that the age at onset has been rising in recent decades [21, 46]. Males are 1.5- times more frequently affected than females.

##### Site

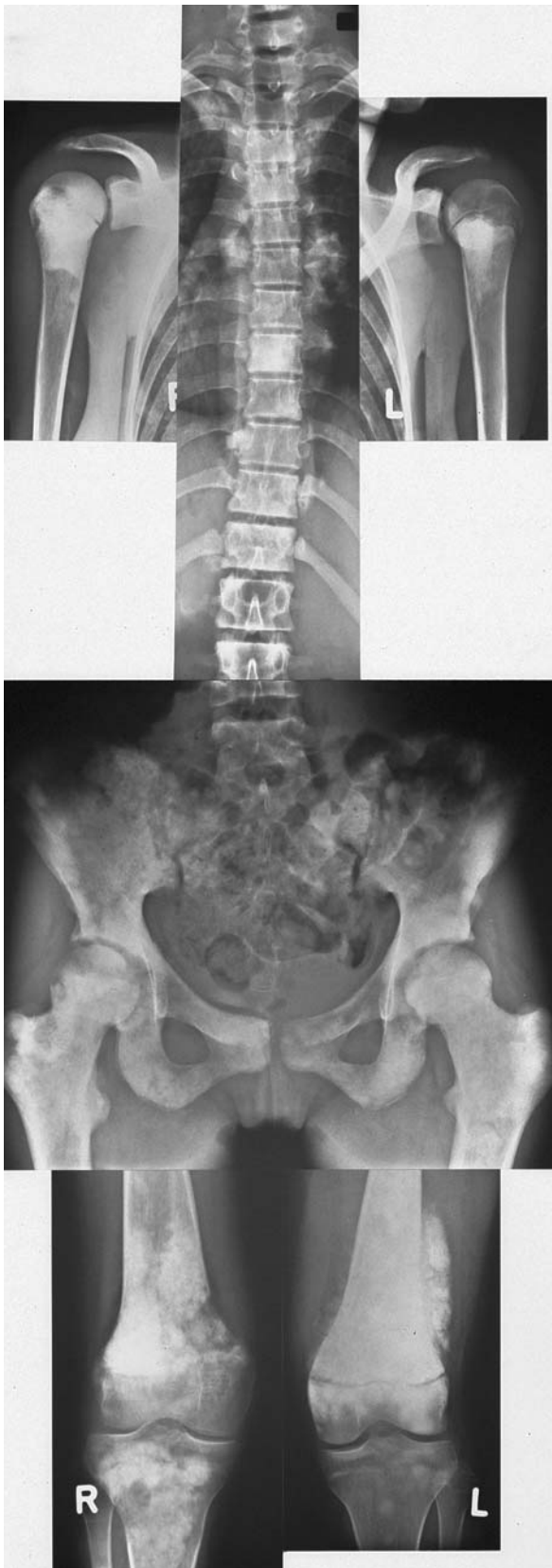
While an osteosarcoma can basically occur in any bone and in any part of the bone, it usually affects the metaphyses of long bones. By far the most commonly affected site is the knee region (distal femoral metaphysis, proximal tibial metaphysis), followed by the proximal humeral, proximal femoral, proximal fibular and distal tibial metaphyses. Occasionally, the tumor can start in the diaphysis, while epiphyseal osteosarcomas are rare. In isolated cases, an osteosarcoma can also develop extraskeletally in the soft tissues [3].

##### Etiology

Almost all osteosarcomas show complex (numerical and structural) clonal chromosome aberrations. They also occur in connection with genetic conditions e.g. retinoblastoma syndrome (mutations in the tumor-suppressor *RB* gene), Li-Fraumeni syndrome (mutations in the *TP53* gene), Rothmund-Thomson syndrome and Werner syndrome (very young patients with unusual tumor sites, e.g. patella) [19, 33, 40, 43; WHO 2000]. Another indication that genetic factors are involved in the development of osteosarcomas is the fact that the tumor can occur concurrently in various sites (multifocal) in certain patients (■ Fig. 4.45).

##### Clinical features, diagnosis

The signs and symptoms are non-specific in the initial stages. Diffuse pain occurs, which may initially be load-related, and subsequently worsens – particularly at night.



■ Fig. 4.45. X-rays of a 14-year old girl with *multifocal osteosarcomas*, which have occurred almost simultaneously in almost all the major bones. The girl died a few months later

With the passage of time, and depending on the site and soft tissue covering in each case, a painful bulge becomes palpable. A Swedish study has shown that the average period from the start of symptoms until the diagnosis of osteosarcoma is confirmed is nine weeks (much shorter than the 16 weeks for Ewing sarcoma or the 20 weeks for chondrosarcoma) [51]. The pain usually becomes very intense after 4–5 weeks. The patient's general health is still unimpaired at this point. Laboratory tests often show an elevated alkaline phosphatase level.

#### Radiographic findings, biopsy

A *plain x-ray* shows the typical picture of a high-grade malignant tumor, i.e. one that penetrates the cortex, destroys the bone, is ill-defined and with irregular cloud-like or solid areas of matrix mineralization within the medullary cavity and in the soft tissues. The rapid growth and destruction of bone produces a spicular or onion-skin-like periosteal reaction, which becomes broader as it nears the tumor and often stops abruptly at tumor level (Codman triangle) as the periosteal new bone formation is destroyed by the rapidly growing tumor (■ Fig. 4.46a). Alternating osteolytic and sclerotic areas are visible in the tumor itself, as well as cloud-like areas of bone matrix calcification. If the tumor also forms cartilage, stippled, annular or arched calcifications can occur. If the tumor is located in the metaphyseal part of the bone, the findings even on the conventional x-ray are so characteristic as to leave almost no doubts about the diagnosis. The extent of the sclerosis can be highly variable however.

The *bone scan* shows massive uptake. The *MRI scan* is particularly suitable for visualizing the soft tissue component, which is usually larger than suggested by the overall picture (■ Fig. 4.46b). The intraosseous extent of the tumor, which is often underestimated on conventional x-rays, can also be better evaluated on the MRI. The MRI scan should always include the whole bone as well as adjacent joints. »Skip metastases« are characteristic of an osteosarcoma. These are tumor islands with no connection with the main tumor and located proximally in the same bone. The bone scan will also indicate the presence of such skip metastases.

Other imaging investigations for an osteosarcoma include a *chest x-ray*, a *chest CT scan* and an *ultrasound* or *CT scan* of the *abdomen* in order to establish whether metastases are already present. The diagnosis is confirmed definitively on the basis of an open *biopsy* (▶ Chapter 4.5.1). This procedure should ideally be performed in the same hospital in which the definitive treatment is provided, so that the surgeon can ensure that the incision does not interfere with the subsequent resection and reconstruction. The biopsy sample must be representative in respect of the differentiation of the tumor, and the specimen should be forwarded unfixed (i.e. under frozen section conditions) to a competent pathologist.



■ Fig. 4.46a, b. Osteosarcoma on the distal femur of a 9-year old girl: a AP and lateral x-rays, b Frontal and sagittal MRI scans. Note the large soft tissue component and the crossing of the epiphyseal plate by the tumor

### Histology, classification

The diagnosis is essentially confirmed by the qualitative detection of direct osteoid formation by atypical tumor cells, although osteoid is sometimes present only in scant quantities. No confirmatory immunohistochemical or molecular biological markers for the diagnosis of an osteosarcoma have been found to date. The overwhelming majority (approx. 85%) of all osteosarcomas are centrally located (intramedullary), high-grade malignant grade III tumors. The high-grade malignant surface osteosarcoma, which by definition extends into the medullary cavity only to a minimal extent, is even rarer (approx. 1%) than teleangiectatic osteosarcoma (4%), which appears as a

purely osteolytic tumor on the x-ray, or small-cell osteosarcoma (1.5%), which can be confused with lymphomas or Ewing sarcoma. In view of the differing therapeutic consequences in each case, such tumors should always be ruled out by immunohistochemical or molecular biological tests if osteoid production cannot be detected with certainty.

Depending on the histology results in each case, a distinction can be made between osteoblastic, chondroblastic or fibroblastic osteosarcomas. The cartilage formation, in particular, should not lead to confusion with a chondrosarcoma, which does not actually show any direct osteoid formation by tumor cells but, at best, enchondral ossification of the tumor cartilage. Certain special forms, e.g. the low-grade malignant central osteosarcoma or the parosteal and some forms of periosteal osteosarcoma, behave less aggressively and can therefore be treated differently.

### Differential diagnosis

Diagnosing a conventional osteosarcoma is not difficult as a rule. It is important to differentiate it from other malignant tumors (*chondrosarcoma, malignant fibrous histiocytoma*). Occasionally the extraosseous parts can be confused with *periarticular calcification* or *myositis ossificans*, which can sometimes appear dramatic enough to make the distinction difficult. However, periarticular calcifications do not ossify from the center outwards like the osteosarcoma, but in the opposite direction. They are also more sharply defined towards the edges. Nevertheless, there have been reports of patients with periarticular calcification undergoing amputation after this had been misdiagnosed as an osteosarcoma. During histological examination it can sometimes be difficult to differentiate an osteosarcoma from an osteoblastoma, giant cell tumors, an aneurysmal bone cyst, a chondrosarcoma and callus tissue.

### Treatment, prognosis

! Osteosarcoma treatment should ideally be the preserve of experienced tumor centers where all the relevant specialists are accustomed to working together.

The treatment must proceed according to a protocol that has been proven in prospective, randomized studies (► Chapter 4.5.5). In our case we follow the COSS protocol (*Cooperative OsteoSarcoma Study*), which stipulates the following treatment:

- high-dose chemotherapy for approx. 3 months (4 cycles),
- wide resection of the tumor with a margin of healthy tissue,
- continuation of the chemotherapy for a total of 9 months [4].

Where the response to chemotherapy was good, a ten-year survival rate of 73.4% has been achieved with the latest COSS protocol since the study was initiated [4]. Similar treatment protocols exist in most West European countries and in North America [2, 4, 13, 26, 30]. Recently, these study groups merged to form the European and American Osteosarcoma Study Group (EURAMOS) and draw up a common protocol so as to increase the store of common experience and achieve better results in a shorter time thanks to the higher pooled case numbers. The practice of initiating the chemotherapy even *before* the resection (neoadjuvant chemotherapy) has become established almost everywhere. This enables the response of the tumor to the drug treatment to be assessed before it is resected. A good result signifies that over 90% of the tumor is necrotic. The histologically verified response to this preoperative chemotherapy is (assuming an adequate tumor resection is performed) the strongest prognostic factor. A recent study failed to provide any evidence that TP53 gene-mutations can predict for the development of metastases [53].

In one study in the Cooperative OsteoSarcoma Study (COSS) series, the survival rates were 73% with a good response and 54% with a poor response [4]. Good indicators for the response to the chemotherapy are the thallium-201 bone scan [28], and the reduction in the alkaline phosphatase level. The presence of metastases or »skip metastases« [26] tends to be a negative prognostic factor. On the other hand, the former assumption that a chondroblastic or telangiectatic osteosarcoma has a poorer prognosis has not been confirmed [31]. Naturally, the initial *tumor size* and *site* (whether close to the trunk or located more peripherally) also play a key role in the prognosis [4]. Female patients appear to have slightly better prospects of a cure than males. Of course, tumor recurrence is a poor prognostic factor. Until recently the intra-arterial administration of the chemotherapeutic agents has not managed to produce any improvement in the results since the contact time is evidently too short. A more recent report challenges this finding [53].

To sum up: the following factors are *prognostically favorable* (in decreasing order of importance):

- good response to chemotherapy,
- adequate resection,
- tumor smaller than 15 cm,
- tumor located more peripherally,
- no metastases,
- no »skip metastases«,
- female patient,
- age at onset of under 30 years.

The *tumor resection* also crucially affects the prognosis. In the past, a radical, i.e. extracompartmental resection

(usually amputation) used to be required. Nowadays, a limb-preserving wide resection is considered to be sufficient [1, 17, 39]. To date we have had to perform an amputation only in approx. 5% of patients. Preservation of the extremity was not possible in these cases because the tumor had penetrated into nerves. The principles of tumor resection and bridging are discussed in ► Chapter 4.5.5. Pulmonary metastases accompanying osteosarcoma are removed surgically. Even multiple metastases in both lungs are resected, repeatedly if necessary. A five-year survival rate of approx. 30% is possible if metastases are resected aggressively [26].

#### **Low-grade malignant central osteosarcoma**

This is an extremely rare bone-forming tumor that usually occurs between the ages of 10 and 30, can affect any bone, but mostly the femur. The tumor grows very slowly and breaks out of the bone only at a late stage. Radiologically, the picture resembles that of the *classical osteosarcoma*, but the x-ray shows a less aggressive pattern (■ Fig. 3.527, ► Chapter 3.5.7).

— *Histologically* the tumor resembles a *parosteal osteosarcoma*, but can also be confused with an *osteoblastoma* or *fibrous dysplasia*. Atypia is fairly rare and must occasionally be actively sought. A wide resection is required since recurrences regularly occur after an *intralesional resection*. The prognosis is good, although metastases can occur. Primary chemotherapy or radiotherapy is not required.

#### **Small-cell osteosarcoma**

The small-cell osteosarcoma can be confused histologically with a *Ewing sarcoma* or *malignant lymphoma*. It is very rare, but has an age and sex distribution pattern similar to that for the *classical osteosarcoma*. The prognosis tends to be slightly worse since the chemotherapy is less effective [32].

#### **Periosteal osteosarcoma**

A rare low- to intermediate-grade malignant tumor, predominantly occurring between the ages of 10 and 20. The x-ray shows a fusiform elevation of the periosteum with erosion of the cortex and ossifying spicules. However, the tumor does not penetrate into the medulla to any great extent and is usually located in the diaphysis. Histological examination reveals a chondroblastic osteosarcoma, usually of intermediate malignancy. The appropriate treatment is wide resection of the tumor. If this is performed in a correct and timely manner, the prognosis is good. Chemotherapy is not particularly effective and should be reserved for high-grade tumors. Metastases occur in approx. 15% of cases. In a recent European multicenter study the 10-year survival rate was 83% [18].



### Parosteal osteosarcoma

Low-grade malignant tumor that grows on the bone surface but that can become highly malignant as a result of dedifferentiation.

- **Occurrence:** Rare, accounting for around 4% of all osteosarcomas (much rarer than the classical osteosarcoma, but some three times more common than periosteal osteosarcoma), peaks during the second and, particularly, the third decades, and affects both sexes with equal frequency.
- **Site:** Most commonly in the metaphysis near the knee [9], in around 50% of cases on the posterior side of the distal femur.
- **Clinical features:** Since the tumor grows slowly it does not cause much pain, but can produce a palpable bulge that the patient finds disturbing.
- The x-ray is very typical, showing a parosteal ossifying tumor mass on top of the bone. The more advanced the tumor, the more radiopaque the ossifications (■ Fig. 4.47). Since it starts to grow at the periphery, a characteristic gap of brightness that is readily visible on the x-ray or CT scan develops between the cortex and the overlying developing mushroom-head-like osteosarcoma (■ Fig. 4.47c). By definition, no more than 25% of the medullary cavity should be affected [34].
- **Differential diagnosis:** While a parosteal osteosarcoma is not difficult to diagnose it must be differentiated from a *high-grade malignant surface osteosarcomas*, which must be treated as for a classical osteosarcoma [49]. Nor is it always easy to differentiate it from *osteochondromas* or secondary (epiexostotic) *chondrosarcomas*.
- **Histologically** the tumor consists of usually parallel trabeculae formed by a moderately cell-rich, collagenized stroma. Osteoblast seams are lacking. The spindly connective tissue cells show – occasionally only slight – atypia. At the periphery the cell content increases markedly, and the tumor can also infiltrate into the adjacent skeletal muscles. Around half of the tumors show cartilaginous differentiation, which can occasionally imitate the cartilaginous cap of an osteochondroma. Cytogenetically, parosteal osteosarcomas are characterized by surplus ring chromosomes.
- The appropriate *treatment* is wide resection. The tumor always recurs after intralesional resection. If the tumor persists for a long time, or if there are multiple recurrences, the tumor can ultimately dedifferentiate, producing the equivalent of a grade 3 sarcoma and with a poor prognosis. On the other hand, if the tumor is resected with a margin of healthy tissue, the prospects of a cure are good, since early metastases almost never occur (resection and bridging in ► Chapter 4.5.5). No other measures are required.



■ Fig. 4.47a–c. Lateral (a) and AP (b) x-rays and CT scan (c) of an 18-year old girl with a *parosteal osteosarcoma*

### 4.5.3.2 Cartilage-forming tumors (chondrosarcomas)

#### Classical central chondrosarcoma

##### ► Definition

Low- to medium-grade malignant (rarely high-grade malignant) tumor with differentiated hyaline cartilage and that develops centrally in the medullary cavity.

— **Synonym:** Central chondrosarcoma

##### Occurrence

After the osteosarcoma, the chondrosarcoma is the second most common solid malignant bone tumor. While it also occurs in adolescents in less than 5% of cases [48], the age peak is between the 5th and 7th decades (■ Fig. 4.28). Males are slightly more frequently affected than females.

##### Site

The chondrosarcoma is usually located in the metaphyses of the long bones, but can also occur in the diaphyses or epiphyses. The tumor also tends to affect the pelvis and scapula.



### Clinical features

The diagnosis of a chondrosarcoma is usually made only at a relatively late stage. According to a British study, the average period between the onset of symptoms and diagnosis is 50 weeks [15]. This slow-growing tumor does not cause severe pain, although dull, diffuse continuous symptoms may be present.

- Radiographic findings: On the *plain x-ray* the tumor appears osteolytic and shows marginal sclerosis of varying severity depending on the rate of growth. The tumor usually erodes or penetrates the cortex (see ■ Fig. 3.254, ▶ Chapter 3.2.13 for an example). A highly typical feature of the tumor, though not necessarily present, are circular or arched (popcorn-like) and stippled calcifications. On the *MRI scan*, the tumor signal is low in T1-weighted images, more intensive in T2-weighted images and particularly strong in proton-weighted images. On the *bone scan* the uptake is not usually very pronounced. Frequently, the *CT scan* is better than MRI for revealing the intraosseous spread of the tumor.
- *Differential diagnosis*: With children and adolescents, the most important task in the differential diagnosis is to separate the chondrosarcoma from a *chondroblastic osteosarcoma*. This is usually possible just on the basis of the x-ray findings. The histological differentiation from an *enchondroma* can be very difficult. Here too, the radiological picture can often provide crucial guidance.

### Histology

Depending on the malignancy grade in each case, histological examination reveals numerous double nuclei and slightly to highly atypical chondroblast tumor cells with dispersed chromatin, visible nucleoli or hyperchromatic giant cell nuclei in a hyaline cartilaginous matrix, some of which shows myxoid quality. Around 60% are Grade 1 tumors, just over 35% G2 and less than 5% G3 sarcomas. Secondary ossification of the cartilaginous tumor matrix can occur. Until the contrary is proven, intraosseous cartilage-forming malignant tumors in adolescents should be considered as chondroblastic osteosarcomas, particularly if these are located in the metaphyses and the x-ray also shows cloud-like calcification patterns.

### Treatment, prognosis

The *treatment* of a classical chondrosarcoma is purely surgical, taking the form of a wide resection. However, in borderline cases where it cannot always be reliably established whether a malignant tumor is already present, an intralesional curettage is permissible provided that the patient is closely monitored with regular subsequent check-ups (resection and bridging are discussed in ▶ Chapter 4.5.5). Chondrosarcomas do not respond to chemotherapy and only minimally to radiotherapy, al-

though the latter (e.g. proton therapy [47]) or treatment with heavy ions [41] may be indicated in exceptional cases, for example if a chondrosarcoma in the sacrum or spine cannot be resected with a margin of healthy tissue and recurs, particularly if neurological lesions occur.

! The prognosis for classical, usually low-grade malignant, chondrosarcoma is good. If an adequate resection is implemented in good time, the survival rate is over 90%, since the tumor grows slowly and only metastasizes at a late stage.

In around 10% of cases however the tumor can dedifferentiate, usually after several recurrences, and transform into a high-grade sarcoma.

### Peripheral (epiexostotic) chondrosarcoma

This form usually develops from an underlying osteochondroma, is rarer than a central chondrosarcoma and shows slightly slower growth than the latter. While this tumor also occurs particularly during middle age, it can occur as early as the second decade, but has never been observed before the onset of puberty.

- They are *sited* wherever osteochondromas occur, but show a predilection for the proximal femur and pelvis (particularly the ilium).
- The primary *clinical* features are an increase in height after the completion of growth and the occurrence of pain. Whether patients with exostosis disease are at increased risk (5-25% according to WHO 2002) is a matter of dispute, although secondary chondrosarcomas do appear to affect the flat bones more frequently and behave more aggressively in patients with exostosis disease.
- *X-rays* show a cauliflower-like tumor with a broad cartilaginous cap (usually >3 cm), whose stem blends with the adjacent bone and whose periphery is characterized by multiple ossifications and calcifications (■ Fig. 4.48). The MRI shows the typical features of the cartilaginous ground substance.
- *Histology*: In contrast with the centripetal, column-like arrangement of increased numbers of cells found in osteochondroma, the epiexostotic chondrosarcoma shows increased cells with nodular proliferating nests, particularly at the edges of the cartilaginous cap, atypia and usually coarse infiltration of the adjacent muscles and underlying medullary cavity.
- *Treatment*: The treatment of choice is wide resection. Note that, since the tumor can extend well into the shaft, continuity should be broken during the resection, entailing a corresponding need for bridging.
- After an adequate resection, the *prognosis* is good since peripheral chondrosarcomas metastasize even slower than central ones. Nevertheless, dedifferentiation is also possible with this tumor, in which case the prognosis is much worse. If malignant progression is

### 4.5.3 · Malignant bone tumors

suspected, even in the absence of symptoms, a wide resection should be recommended to the patient. The prognosis of dedifferentiated chondrosarcoma is extremely poor and adjuvant chemotherapy has little influence on the outcome [10].

#### **Clear cell chondrosarcoma**

Rare tumor with very low-grade malignancy, frequently located in the epiphyses. The tumor has not hitherto been observed during adolescence, and all our patients are over 20 years old. The prognosis is comparable to that for the classical low-grade chondrosarcoma [23]. The treatment consists of a wide resection.

#### **Mesenchymal chondrosarcoma**

This is a rare tumor that is more likely to occur in the 3rd rather than the 2nd decade of life, although we have also observed cases in small children. The tumor can occur in any bone, but also as a primary tumor in the soft tissues (up to 30% of cases). Radiologically this is an osteolytic lesion that can penetrate into the cortex.



■ Fig. 4.48. AP x-ray of the proximal femur in a 20-year old young man with an *epixostotic chondrosarcoma*

- *Histological* examination reveals a tumor with a biphasic structure containing rotund tumor cells in addition to well differentiated sections with nodular hyaline cartilage. The picture is in part reminiscent of a *Ewing sarcoma*, or a *hemangiopericytoma*. *This tumor is more malignant than a classical chondrosarcoma.*
- *Treatment* must involve a wide resection.
- The *prognosis* is worse than for other types of chondrosarcoma. As with Ewing sarcoma, chemotherapy appears promising.

#### **Periosteal chondrosarcoma**

This rare tumor occurs between the ages of 10 and 50. It originates from the cortical bone, develops along the bone, and is of low malignant potential, and therefore has a better prognosis than the classical chondrosarcoma.

- Nevertheless, the *treatment* of resection should ideally include a margin of healthy tissue.
- In the differential diagnosis, the tumor must be differentiated primarily from a *periosteal chondroma* and a periosteal osteosarcoma.

#### **Fibrocartilaginous mesenchymoma**

This tumor is extremely rare, and only a few cases are described worldwide in the literature. Only 17 cases have been reported to date [16], and we have personally observed two patients with this tumor. It occurs predominantly during adolescence and is usually located in the metaphysis. The x-ray shows a chambered osteolytic process with marginal sclerosis (▶ Chapter 4.5.5, ■ Fig. 4.63). Macroscopically the fibrous tissue is permeated with wound, cartilaginous strands of tissue that resemble shrimps.

- *Histological* examination reveals connective tissue and cartilage proliferations reminiscent of epiphyseal plates. The tumor is of low grade malignancy.
- As *treatment* we perform a wide resection, although metastases have not been described to date.

### 4.5.3.3 Tumors of the medullary cavity (Ewing sarcoma, primitive neuroectodermal tumor: PNET)

#### **Ewing sarcoma**

##### ➤ **Definition**

High-grade malignant, undifferentiated small-cell tumor, probably of neuroectodermal origin, and arising in the medullary cavity of bone.

##### **Occurrence**

The Ewing sarcoma typically occurs during childhood and adolescence. It is the third most common solid tumor of bone and is more common in boys than in girls. 90% of these tumors are observed before the age of 25.

### Site

In principle, the Ewing sarcoma can occur in any bone, but tends to show a predilection for the *diaphyses*, and also the metaphyses, of *long bones*, and is more frequently seen in the lower leg than the femur. It is particularly common in the pelvis, followed by the arms, ribs and spine.

### Etiology, pathogenesis

As with the osteosarcoma, *genetic factors* play an important role in the development of the Ewing sarcoma. The translocation  $t(11;22)(q24;q12)$  is highly characteristic and can be detected in almost 90% of all Ewing sarcomas [5]. The other Ewing sarcomas show similar translocations, e.g.  $t(21;22)(q22;q12)$ , which all lead to differing fusions of the *EWS* gene with members of the *ETS* family (transcription factors).

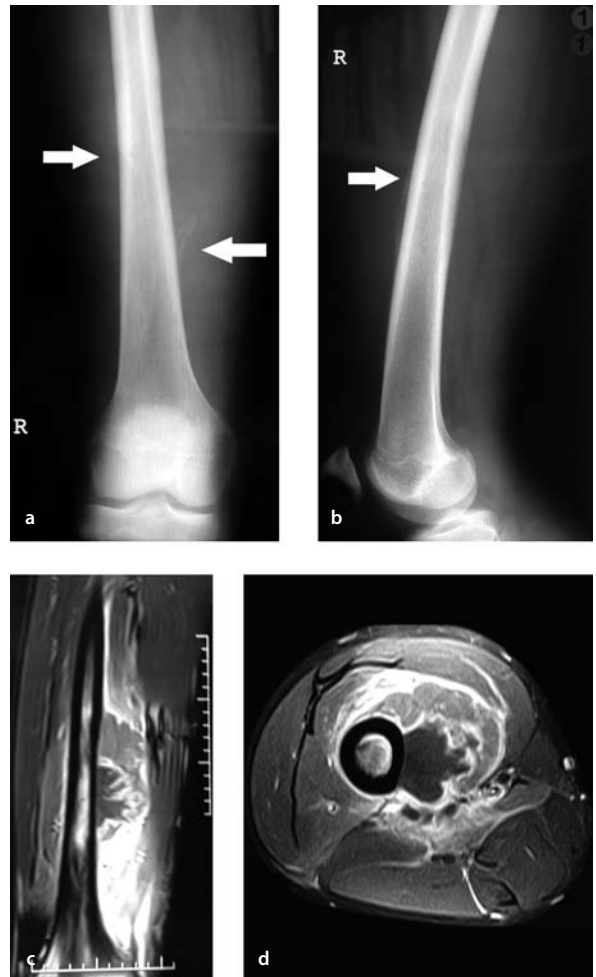
### Clinical features, diagnosis

The average period from the onset of symptoms to the diagnosis is longer for the Ewing sarcoma than the osteosarcoma, namely 4 months [51]. The tumor grows rapidly and is painful. A swelling is also often palpable. Febrile episodes and anemia also occur not infrequently. The blood count may show leukocytosis with an elevated erythrocyte sedimentation rate and increased CRP, although a normal blood test does not rule out the possibility of a Ewing sarcoma. An elevated serum lactate dehydrogenase level is an indication of the existence of metastases and therefore an unfavorable prognostic factor.

### Radiographic findings

The radiological findings on *plain x-rays* are highly variable. Often just minimal osteolysis and a slight periosteal reaction are observed (■ Fig. 4.49a, b). In other cases the x-ray shows a fairly large osteolytic tumor with penetration of the cortex, an onion-skin-like periosteal reaction and the formation of spicules. Much more information can be gleaned from the *MRI scan*, which usually shows a high proportion of soft tissue section in the tumor (■ Fig. 4.49c, d, also ■ Fig. 4.62a, ► Chapter 4.5.5).

The soft tissue spread can be very substantial. The tumor produces moderately intense signals in all weightings. Within the bone as well, the tumor usually shows a much greater spread than suggested by the overview images. »Skip metastases« on the other hand are very unusual with Ewing sarcoma. Other investigations include a *bone scan*, which shows massive uptake, and possibly other osseous foci as well, since the Ewing sarcoma also occurs as a multicentric tumor, with the simultaneous occurrence of foci in several bones. If a Ewing sarcoma is suspected, a *chest x-ray* and *CT scan* of the chest and abdomen can provide clarification. The definitive diagnosis must be confirmed by an open *biopsy*.



■ Fig. 4.49a–d. X-rays (a, b) and MRI (c, d) of the distal femur of a 17-year old male patient with *Ewing sarcoma*

! With a *Ewing sarcoma*, metastases appear at an earlier stage than with an *osteosarcoma*, whereas the diagnosis is confirmed, on average, at a later stage.

### Histology

The Ewing sarcoma consists of small, uniform cells with scant cytoplasm that are slightly larger than lymphocytes, with round or slightly oval nuclei and chromatin, usually finely distributed, without nucleoli. The cells contain intracytoplasmic glycogen, do not form reticular fibers, often show minimal mitoses and produce typical immunohistochemical findings [14, 42]. The fusion products of the  $t(11;22)(q24;q12)$  translocation can also be detected by molecular biological tests (ideally on unfixed tissue) [11].

### Differential diagnosis

Diagnosing a Ewing sarcoma on the basis of imaging procedures is not always easy since it can be confused not only with *osteomyelitis*, *Langerhans cell histiocytosis*,

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stress fractures, but also with *osteosarcoma* and *metastases*. Radiologically it cannot be differentiated at all from a *primitive neuroectodermal tumor (PNET)* which, on immunohistochemical testing, shows neuroendocrine markers such as synaptophysin and neuron-specific enolase (NSE), while histological examination will occasionally show rosette formation.

#### Treatment, prognosis

Up until the end of the 1970's only 10% of patients with Ewing sarcoma survived despite chemotherapy and radiotherapy. At the end of the 70's, high-dose chemotherapy was introduced and wide resection was now also routinely attempted. Treatment nowadays should be based on an internationally recognized protocol (► Chapter 4.5.5). Our hospital follows the EICESS guidelines or the recommendations of the EUROEWING study [7, 35, 36, 44].

**! As with osteosarcoma, the Ewing sarcoma should also be treated in a center in which all the necessary specialists with the appropriate experience work together.**

The *therapeutic strategy* is similar to that for osteosarcoma:

- confirm the diagnosis by means of a biopsy,
- chemotherapy for 3 months,
- wide resection of the tumor,
- further chemotherapy for 6 months,
- radiotherapy if there is doubt as to whether the resection extended into healthy tissue.

The initial (neoadjuvant) chemotherapy for three months enables the response of the tumor to chemotherapy to be assessed by the time of the resection. A good response means that over 90% of the tumor is necrotic. *Radiotherapy* can improve the outcome of treatment [7].

The *prognosis* for Ewing sarcoma is not quite as good as that for osteosarcoma, since micrometastases occur at a very early stage and are usually already present at the time of diagnosis. Nevertheless, five-year survival rates of 60–70% can still be achieved for tumors in the extremities. Tumors of the pelvis and spine, however, have a much worse prognosis [38, 50], since the chances of survival are just 10–30%.

**! Bear in mind that these survival rates are only achieved in major centers with the appropriate expertise. Doctors that only treat isolated cases must expect much worse results.**

As with osteosarcoma, amputations are hardly ever necessary these days for Ewing sarcoma. Only a wide rather than a radical resection is required. A compromise treatment in the area of major nerves and vessels is likely to be more successful with Ewing sarcoma than with the osteosarcoma since postoperative radiotherapy is possible. For very large tumors that present difficult resection problems we

occasionally administer preoperative radiotherapy as well. This reduces the risk of tumor metastasis during surgery.

On the other hand, the resection is more difficult in technical respects. In particular, the intraoperative blood loss during operations performed in the first few months after radiotherapy, especially for pelvic tumors, is even greater than usual. If a nerve passes through the tumor, as is invariably the case for example in the proximal fibula (peroneal nerve), we resect the tumor, together with the nerve, well into healthy tissue, and bridge the gap several weeks later with a graft. Naturally we try to avoid postoperative radiotherapy in this situation. Apart from local complications, there will still be a risk of secondary tumors [12, 29, 45].

Resection and bridging are discussed in ► Chapter 4.5.5.

#### Primitive neuroectodermal tumor (PNET)

This tumor is closely related to Ewing sarcoma and can be differentiated from the latter only by histological and immunohistochemical investigation. It also appears to behave slightly more aggressively than Ewing sarcoma [8]. The t(11;22)(q 24;q 12) translocation also occurs with the PNET [24]. The therapeutic principle is identical to that for Ewing sarcoma, although the chemotherapy is slightly different. Since this type of tumor was only detected as a separate entity and differentiated from the Ewing sarcoma by means of immunohistochemical markers around 12 years ago, statements about its prognosis are still limited [42].

### 4.5.3.4 Fibrohistiocytic tumors (fibrosarcoma, malignant fibrous histiocytoma)

#### Fibrosarcoma

This rare tumor is observed primarily in adults, but can also occur in adolescents in isolated cases [6].

- On the x-ray the tumor is predominantly osteolytic with ill-defined margins and with minimal surrounding sclerosis. Low-grade malignant fibrosarcomas can be very difficult to differentiate from a desmoplastic fibroma.
- *Treatment:* In juvenile patients chemotherapy is always indicated in addition to a wide resection.

#### Malignant fibrous histiocytoma(MFH)

This tumor is rarer than Ewing sarcoma, but commoner than fibrosarcoma. Although it is a tumor of middle age, isolated cases have also been observed during the second decade of life.

- The *radiological features* match those for fibrosarcoma.
- The *differential diagnostic considerations* are comparable.
- The *therapeutic protocols* are similar to those for osteosarcoma.
- The *prognosis* is moderately good.



#### 4.5.3.5 Malignant vascular tumors

##### **Hemangioendothelioma and angiosarcoma**

Vascular tumors can show all grades of malignancy, occur at any age between 10 and 70, primarily in the lower extremities. On x-rays, the tumors appear exclusively osteolytic and show only reactive, straggly sclerosis.

- The *histological picture* varies considerably. The tumor cells in *hemangioendothelioma* can show a broad eosinophilic cytoplasm that may contain lumina that shift the nucleus toward the periphery, producing the appearance of a signet ring. *Angiosarcomas* consist, at least in part, of vascular channels lined with highly atypical cells. The nuclei are enlarged, pleomorphic and hyperchromatic.
- The *treatment* and *prognosis* depend very largely on the degree of differentiation. A marginal resection is sufficient for highly differentiated hemangioendotheliomas, although these can also occur as multifocal tumors. The prognosis for high-grade malignant forms is poor, and wide resection is required in such cases. Although precise figures on the success of radiotherapy and chemotherapy are not available, the prognosis is generally poor.

##### **Hemangiopericytoma**

This is an extremely rare bone tumor that also occurs in children and adolescents. The tumor is osteolytic, shows permeative growth and induces a reactive sclerosis (■ Fig. 4.60, ► Chapter 4.5.5). A hemangiopericytoma is a low-grade malignant tumor, and complete recovery can generally be achieved with a resection extending into healthy tissue.

#### 4.5.3.6 Other malignant bone tumors

##### **Adamantinoma**

###### ► Definition

Low-grade malignant tumor consisting of mesenchymal and epithelial cells and occurring in the diaphyses and metaphyses of long bones, with a very strong predilection for the tibia. Very rare tumor that is completely unrelated to ameloblastoma of the jaw, which – in former times – also used to be known as an adamantinoma.

###### Occurrence, site

Very rare tumor occurring primarily between the ages of 10 and 40. Three quarters of the patients are male. Over 90% of all cases are located in the tibia, mainly in the diaphysis, and possibly also in the metaphysis.

###### Clinical features

Since the tumor grows very slowly it causes few symptoms, although diffuse pain can occasionally occur. The patient may notice a nodular, bumpy surface on the anterior as-

pect of the tibia. Pathological fractures occur. The x-ray shows honeycomb-like areas of osteolysis surrounded by sclerosis in the cortical bone, although these invariably infiltrate into the medullary cavity. The cortical bone may be widened, but is rarely penetrated (■ Fig. 4.50). Both the MRI and CT scans can help identify areas of discontinuous spread.

The most important and most difficult differential diagnosis is *osteofibrous dysplasia*, which shows a very similar radiological picture and also occurs preferably in the tibia, particularly in the shaft area. In osteofibrous dysplasia the sclerosis tends to predominate and the patients are younger. However, this tumor is restricted to the cortical bone and does not penetrate into the medullary cavity. Nevertheless, the differentiation can be difficult. In cases of very extensive osteofibrous dysplasia, a representative biopsy is needed therefore to rule out a malignant adamantinoma. The situation is made more complicated by the fact that both tumors can occur next to each other concurrently in the same patient [37, 45].

###### Histology

Diagnostic confirmation of an adamantinoma is provided by the appearance of epithelial cell nests em-



■ Fig. 4.50. AP and lateral x-rays of an adamantinoma of the tibia in a 17-year old female patient



### 4.5.3 · Malignant bone tumors

bedded in a spindle cell stroma. The picture under a light microscope is generally unequivocal. Occasionally, however, spindle cell epithelial formations occur that are almost impossible to differentiate from the stroma sections, in which case immunohistochemical investigations are required to identify the epithelial groups [25]. Cytokeratin-positive individual cells in an osteofibrous dysplasia-like stroma constitute a special variant (osteofibrous dysplasia-like adamantinoma) that is rarely able to metastasize [22].

#### Treatment, prognosis

The tumor must be resected widely, otherwise it will recur. If left untreated, or usually after several recurrences, it can also metastasize. Fatalities have been reported. It is very important, therefore, to differentiate it unequivocally from osteofibrous dysplasia, which is generally not treated but simply observed. An intralesional resection of the adamantinoma is not sufficient. Corresponding bridging procedures are required after wide resections (► Chapter 4.5.5). Fortunately, since the epiphyses are rarely involved, functionally effective bridging is usually possible.

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#### 4.5.4 Soft tissue tumors

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- This chapter focuses exclusively on those orthopaedically relevant tumors and tumor-like lesions in soft tissues that occur primarily in childhood and adolescence [12, 36]. The Enneking staging system can also be used for these tumors [10, 31] (► Chapter 4.5.1).

##### 4.5.4.1 Benign and locally aggressive tumors

###### Connective tissue tumors

###### Fibrous hamartoma of infancy

Synonym: subdermal fibromatosis

A *fibrous hamartoma of infancy* occurs almost exclusively in the area of the *shoulder* and *axilla*, primarily in boys. Although the tumor is rare, it is one of the most common soft tissue lesions in early childhood and usually manifests itself during the first three years of life. Histologically the lesion consists of three different components:

- bundles of intertwining fibroblasts which surround
- immature roundish cells embedded in a myxoid matrix, and which are also associated with
- lobularly structured fatty tissue.

A marginal resection is usually sufficient for removing this altered tissue.

###### Calcifying aponeurotic fibroma

Synonym: aponeurotic fibromatosis

A *calcifying aponeurotic fibroma* is a painless lesion that occurs primarily in boys under 18 on the aponeuroses of the *hands* and *feet*, and *very rarely in other sites as well*. Clinical examination reveals an ill-defined, solid compact mass. Macroscopically, the tumor appears pale grey and granular. Histologically, it consists of fibroblasts arranged in columns and forming moderate quantities of collagen. The central part of the tumor (except in toddlers) always features calcifications and chondroid metaplasia. Since the tumor infiltrates the surrounding

tissues it cannot usually be removed with a margin of healthy tissue. However, the tumor regresses after puberty and rarely recurs. The surgeon should therefore be very cautious in deciding whether resection is indicated since no further progression is expected after completion of growth. Since fibrosarcomas have been observed in, or after the excision of, a calcifying aponeurotic fibroma in just two cases to date, clinical follow-up is important [11].

### Myofibroma and myofibromatosis

Synonym: congenital (generalized) fibromatosis

These generally rare lesions can occur in a – more common – solitary form (*myofibroma*) or as multiple lesions (*myofibromatosis*). They can affect the internal organs or the skeletal system, particularly the long bones. Myofibromas are often diagnosed even at birth or during the first two years of life. They are nodular, probably hamartomatous lesions, consisting histologically of spindle-shaped myofibroblasts, collagenous fibers and a prominent capillary vascular network. Since their appearance can be highly variable myofibromas can resemble leiomyomas, neurofibromas and, in particular, hemangiopericytomas. In view of their necrotizing tendency they risk being confused with sarcomas. Marginal resection of the tumors is usually sufficient, and spontaneous cures can also occur. The prognosis is serious, however, if there is multifocal involvement of internal organs (lung, heart, bowel), particularly during infancy and early childhood.

### Fibromatoses, desmoid tumors

#### ➤ Definition

Fibromatoses are lesions showing invasive, permeative and progressive growth and consisting of fibroblasts and myofibroblasts and profuse quantities of collagen. They are relatively common, usually show the same histological pattern and differ according to their location (superficial or deep) or clinical presentation (e.g. abdominal fibromatosis in young women post partum). Some forms occur in small children, adolescents and young adults.

#### Superficial (fascial) fibromatoses

*Digital fibromatosis* or *inclusion body fibromatosis* usually affects the middle or distal phalanx of a *finger* or *toe*, occurs almost exclusively in small children and can even manifest itself at birth or shortly thereafter. The lesion is more commonly seen on the extensor side than the flexor side and is very rare. Histologically it consists of fibroblasts with intracytoplasmic eosinophilic round inclusions and a dense network of collagen fibers. Recurrences frequently occur after incomplete resection. However, since the lesion regresses spontaneously at a later date,

the surgeon should be very cautious in deciding whether resection is indicated. In fact, resection is required only to preserve function.

#### Deep (musculoaponeurotic) fibromatoses (desmoid tumors)

The *extra-abdominal fibromatoses (desmoid tumors)* are more common in male patients, in contrast with abdominal fibromatoses. Adolescents and young adults are preferably affected. The shoulder-neck region, hip, buttocks and extremities are usually involved.

- *Etiologically*, genetic (trisomies 8 or 20 in up to 30% of cases, familial occurrence, association with Gardner syndrome), hormonal (development during pregnancy) and traumatic factors have been discussed. Other skeletal anomalies have also been observed with increased frequency in patients with desmoid tumors [33].
- *Occurrence*: An annual incidence of 3–4 new cases per million inhabitants has been calculated for Finland [33].
- *Clinical examination* reveals a painless, coarse mass that can grow to an exceptionally large size. The tumor can displace muscles and thereby cause contractures. It infiltrates the skeletal muscles and occasionally spreads along the fascia. It can also compress or infiltrate nerves and thus trigger neurological symptoms.
- *Radiologically* the desmoid appears as an ill-defined soft tissue mass. The MRI scan reveals an ill-defined infiltrating lesion with a signal that is slightly more intense than that of muscle (■ Fig. 4.51).
- *Histologically* the tumor consists of relatively uniform spindle-shaped fibroblasts without atypia. The nuclei are usually pale and small, while the nucleoli are barely discernible. Mitoses are very rare and should arouse suspicions of a highly differentiated fibrosarcoma. The cells form profuse amounts of collagen, which pushes the tumor cells apart, thus producing the impression of a lesion with low cell content. Immunohistochemically, desmoids are positive for beta-catenin and estrogen receptor beta [1, 8]. A second histological form can occur in small children (*infantile fibromatosis*). This consists of small, immature round-oval fibroblasts embedded in a myxoid matrix.
- In the *differential diagnosis* the most important condition to consider is the highly differentiated *fibrosarcoma*. While both tumors can show a similar locally aggressive behavior, the desmoid does not metastasize. Desmoids grow progressively and show a great tendency to recur. A wide, non-mutilating resection should be attempted, but is often difficult to achieve, since the tumor appears to recur more rapidly after each reoperation.

**Fig. 4.51 a, b.** MRI scan of the distal femur (frontal section; **a**) in a 17-year old male patient with (histologically and immunohistochemically confirmed) *desmoid* that has infiltrated the distal half of the whole quadriceps muscle. **b** 6 months later after a cycling tour of over 5,000 km the tumor has shrunk to one third of its original size. The patient's condition has been stable for 4 years



— **Treatment:** A wide resection should be attempted at the first operation [32], since patients with incomplete, possibly marginal, resections are associated with a high risk of recurrence. The recurrence rate is higher in children and adolescents than in adults. It can be reduced with radiotherapy, hormones (tamoxifen) or preoperative chemotherapy [22, 30, 38]. Hormone therapy is promising if hormone receptors are found in the histological specimen. Rare spontaneous regressions have been reported in the literature [6]. We have observed spontaneous regression of the tumor in isolated cases, possibly in connection with intense sporting activity (■ Fig. 4.51). Exercise therapy may have a positive effect on desmoids.

## Fibrohistiocytic tumors

### Giant cell fibroblastoma

The giant cell fibroblastoma was first described as a separate entity just a few years ago. It occurs almost exclusively in children and appears to constitute the infantile variant of dermatofibrosarcoma protuberans [17]. Like this tumor, giant cell fibroblastoma presents the same translocation  $t(17;22)(q22;q13)$  resulting in the fusion of the collagen alpha 1 gene with the gene coding for platelet-derived growth factor beta that can be used for molecular diagnosis by Reverse Transcriptase Polymerase Chain Reaction (RT-PCR) [34]. Clinical examination reveals painless lumps or swellings in the skin or subcutaneous tissues, predominantly on the lower leg, groin area or chest wall, and usually in male patients.

— **Histological investigation** reveals spindle-shaped tumor cells embedded in a fibromyxoid stroma and infiltrating into the skin appendages or fatty tissues. A diagnostic pointer is the appearance, under the electron microscope, of cavities lined with giant cells, each with a large hyperchromatic giant nucleus showing multiple notches. Under the light microscope, however, this can give the impression of multinuclear cells. As a result of the myxoid stroma and nuclear

hyperchromasia, this tumor is fairly often confused with a sarcoma.

— **Treatment, prognosis:** Since giant cell fibroblastomas recur in almost fifty percent of cases, they should be excised widely. Metastases have not been reported to date.

### Plexiform fibrohistiocytic tumor

First described only in 1988, the plexiform fibrohistiocytic tumor occurs mainly in children and young adults [17]. Located in the upper or lower extremities, it presents as a painless swelling measuring just a few centimeters.

— **Histologically** it consists of multiple nodular infiltrates of histiocytic cells and giant cells surrounded by bundles of intertwining spindle-shaped fibroblasts. Occasionally the fibroblast component predominates. Atypia is not present and mitoses are sometimes observed.

— The **differential diagnosis** must particularly take account of the possibility of granulomatous inflammations, fibromatoses and fibrous histiocytomas of the skin.

— **Treatment, prognosis:** These tumors show a great tendency to recur, and lymph node metastases have been described in isolated cases. The tumors should therefore be resected with a correspondingly safety margin.

### Angiomatoid fibrous histiocytoma

The angiomatoid fibrous histiocytoma is observed particularly during childhood and adolescence and is currently considered to be a tumor of unclear differentiation with intermediate-grade malignancy since, although it recurs, it only metastasizes in exceptional cases [11]. It occurs mainly in the skin and subcutaneous tissues and primarily affects the extremities. It is occasionally associated with generalized symptoms such as anemia, fever and weight loss. Pain is fairly rare. The well defined lesions, often just a few centimeters in size, show irregular blood-filled cavities even on the cut surface and resemble a hematoma.



#### 4.5.4 · Soft tissue tumors

- *Histological examination* reveals, in addition to the blood-filled cavities, central accumulations of relatively uniform histiocytic cells with round-oval nuclei that may contain intracytoplasmic hemosiderin and, at the edges, inflammatory infiltrates some of which consist of follicularly arranged lymphocytes with the formation of germinal centers. Although atypia, and also hyperchromatic giant cells, can occur, these are not indicators of malignancy.
- *Treatment* ideally involves complete surgical removal, together with the pseudocapsule surrounding the tumor.

#### Fatty tissue tumors

Fatty tissue tumors occur mainly in adults. The relevant tumors during childhood and adolescence are *lipoblastoma* or *lipoblastomatosis*, the rare *diffuse lipomatosis* and the *intramuscular lipoma* – which can occasionally also occur in children. The *lipoblastoma* occurs exclusively during early childhood, primarily in boys, and affects the upper and lower extremities. Whereas a lipoblastoma is circumscribed and limited to the subcutaneous tissue, its diffuse form, *lipoblastomatosis*, also infiltrates the adjacent muscles. Both consist of immature, lobularly arranged fat cells embedded in a macroscopically visible myxoid matrix. Consequently, and also because of their immature spindle- and star-shaped cells, they are sometimes confused with a myxoid liposarcoma, which almost never occurs in this age group. Genetic investigations can clarify the situation in doubtful cases since these lesions show recombinations of 8q11–13 with 3q12–13, 7p22, 8q24 or other chromosomal sections.

Simple excision is usually sufficient as treatment, although a wide resection should be attempted for diffuse lipoblastomatosis in view of the poorly defined contours and the associated tendency to recur. The extremely rare condition of *diffuse lipomatosis* involves mature, excessively growing fatty tissue that can affect major parts of an extremity or sections of the trunk and infiltrate the subcutaneous tissues and muscles. The condition is often associated with skeletal hypertrophy. Since it sometimes spreads substantially, extensive surgical interventions are occasionally unavoidable. Although *intramuscular lipomas*, which consist of mature adipocytes, are rare during childhood and adolescence, because of their location, infiltrative growth and size they can cause considerable therapeutic problems. Occasionally the patient becomes aware of their presence when they become slightly painful.

- *Histologically*, because of their infiltrative growth into the skeletal muscles, these tumors are sometimes very difficult to differentiate from highly differentiated liposarcomas despite any preserved lobulation.
- *Therapeutically*, resection with a margin of healthy tissue is indicated, otherwise recurrences will occur.

#### Vascular tumors

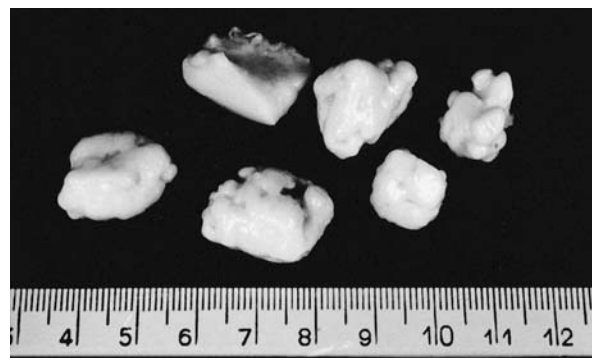
##### Angiomas

Angiomas are lesions that emulate the structure of blood (*hemangiomas*) or lymphatic (*lymphangiomas*) vessels. It can often be impossible to establish whether a genuine tumor or a malformation / hamartoma is present. In clinical respects, the site and extent (involvement of a body segment – angiomatosis/lymphangiomatosis) are the most important factors. Angiomatosis can affect several tissue types (skin, muscle, bone ) or several segments of the same tissue (several muscles) and can lead to hypertrophy of the extremity (▶ Chapter 4.6.6.4, Klippel-Trenaunay syndrome).

##### Hemangioma

Clinically, we make a distinction between superficial and deep hemangiomas. Whereas superficial hemangiomas are often already visible at birth, the deep hemangiomas only manifest themselves during adolescence. The intramuscular hemangiomas form the largest group. These are usually located in the thigh and can be painful. If they extend over large areas they can also lead to contractures or functional impairment. The easiest way to confirm the diagnosis is by means of the MRI scan (■ Fig. 3.452, ▶ Chapter 3.4.13).

- *Histologically*, hemangiomas can be subdivided into *capillary* and *cavernous* types. Capillary hemangiomas, particularly in neonates and toddlers, can be very cell-rich and show mitoses. The deep intramuscular hemangiomas almost invariably correspond to the capillary type. Since mitoses and endothelial proliferations (but not cellular atypia) also occur occasionally they can be confused with an angiosarcoma.
- *Treatment* involves resection, ideally with a substantial margin of healthy tissue. Since hemangiomas possess numerous offshoots extending along the vessels into the intermuscular septa and the surrounding soft tissues, they are particularly amenable to resection by laser surgery.



■ Fig. 4.52 Cartilage fragments in *synovial chondromatosis* of the knee in a 12-year old boy



### Capsulosynovial angioma

This very rare lesion usually occurs between the ages of 5 and 10. The tumor is almost invariably located inside the knee joint beneath the synovial membrane. Clinical examination reveals painful swellings and occasionally joint effusion. The diagnosis can be confirmed by MRI.

- The principal *differential diagnosis* to consider is pigmented villonodular synovitis.
- *Histological examination* shows the typical picture of a cavernous hemangioma.
- As far as *treatment* is concerned, the tumor must be resected widely, for which a combined approach to the joint from two sides is sometimes required. Provided the surgical treatment is adequate, recurrences are rare.

### Lymphangioma

Lymphangiomas may already be present at birth or occur during the first year. They are located primarily in the neck or shoulder area, but can also occur inside the abdomen. They are much rarer, on the whole, than hemangiomas. We make a distinction between cavernous and cystic lymphangiomas. These lesions must also be resected.

#### 4.5.4.2 Tumor-like lesions

##### Popliteal cyst

*Popliteal cysts* usually occur unilaterally and are more common in boys than girls. They usually originate from a bursa located under a tendon, either the tendon of the semitendinosus or semimembranosus muscle or the medial or lateral head of the gastrocnemius. Cysts also occasionally occur as a result of capsular herniation. The cysts (more accurately pseudocysts since they do not possess an epithelial lining) are bordered by a connective tissue membrane and are filled with synovial fluid. Clinically, the cyst is a firm, elastic, and sometimes very large, mass in the popliteal fossa.

The *sonogram* shows the presence of fluid. These cysts are rarely painful. Resection is rarely indicated since the cysts disappear spontaneously. We only resect very large and painful cysts. In such cases, recurrences occur if the pedicle is not removed from the tendon during the resection.

##### Ganglion

Ganglia occur at any age, but primarily during adulthood. They can originate from joint capsules, tendon sheaths or from the meniscus, have a relatively tough capsule, are firm and elastic and filled with a gelatinous fluid. While they can vary in size, they are rarely larger than 1–2 cm. Depending on its location, a ganglion can cause mechanical interference. In such cases, the ganglion should be resected. The surgeon should ensure that the pedicle is removed otherwise recurrences can occur.

### Synovial chondromatosis

Synovial chondromatosis is relatively rare. While it occurs mainly in adults, it can also occur in adolescents (■ Fig. 4.52). The condition tends to affect the major joints and, very rarely, the tendon sheaths. The knee is by far the most commonly affected site. The symptoms develop gradually, with pain, movement restriction and crepitations. Joint locking is rare and joint effusions are not particularly substantial. On the x-ray the roundish cartilage fragments are only visible at a late stage when they calcify or show secondary enchondral ossification (*osteochondromatosis*). By contrast, they are readily discerned on the MRI scan or during arthroscopy.

- *Histological examination* reveals nodular cartilage proliferation within the synovial membrane. Under no circumstances should the high cellular content and nuclear polymorphism at this site be interpreted as indicators of malignancy. In the context of cytogenetics, clonal chromosomal aberrations have been described. The disease progresses slowly (except in the very rare, aggressive pseudotumoral form).
- The *treatment* consists of the removal of loose joint bodies. In the event of a recurrence, synovectomy is recommended. Further recurrences are rare after this procedure, even if the synovectomy was not complete.

### Pigmented villonodular synovitis

While this disease tends to occur in adult patients, it is also observed in adolescents. A distinction is made between a *diffuse villous or villonodular form* and a *localized nodular form*, which is also known as a *synovial giant cell tumor*. In addition to *articular* manifestations, the condition can also occur in *extra-articular* sites. The commonest site is the knee (■ Fig. 4.53).

- The *symptoms* are relatively mild initially, and the condition is often diagnosed only after several years. Thickening of the joint capsule is observed, and recurrent effusions can occur. The joint aspirate is usually brownish. The x-ray shows erosions of the adjacent bone with intraosseous pseudocystic osteolytic areas. The diagnosis can usually be confirmed by an MRI scan, which shows the typical tumorous thickening of the synovial membrane and a signal pattern that is characteristic of hemosiderin.
- *Histology*: The brown discoloration of the thickened and coarsened synovial villi produced by the hemosiderin deposits is striking even at the macroscopic level. This is caused by infiltration with roundish-polygonal, siderin-loaded cells, intermixed with xanthoma cells and spindle-shaped fibroblasts. Alongside these are giant osteoclast-type cells and chronic inflammatory infiltrates. Hypocellular areas may be collagenized.
- *Treatment* involves as complete a synovectomy as possible. Two approaches are usually required in the



■ Fig. 4.53. MRI scan (sagittal section) of the knee in a 16-year old girl with pigmented villonodular synovitis (arrows show areas of nodular capsular thickening)

knee, while surgical dislocation may be needed in the hip. Since the synovectomy is almost never complete, recurrences occur repeatedly. We therefore perform a synoviorthesis with osmic acid or radionuclides six weeks after the resection, although only for patients that have stopped growing.

#### Myositis ossificans (heterotopic ossification)

This condition involves the posttraumatic or postoperative formation of new bone within the muscles or periosteum (periostitis ossificans). Substantial ossifications occur in children and adolescents mainly at the proximal end of the extremities (upper arm, shoulder, thigh, hip and knee). These are frequently observed in connection with neuromuscular disorders (cerebral lesions or paraplegias). The usual onset involves a coarse swelling of the soft tissues with slight pain 2–4 weeks after an (often minor and unremembered) trauma or after an operation. The lesion continues to grow over the following weeks, reaching its final size within around two months. During its growth phase the lesion is usually painful.

— *Radiologically*, the picture initially appears negative. After two months a, generally sharply defined, shadow zone that is accentuated towards the periphery becomes visible. Its radiopacity steadily increases on subsequent x-rays. While it is not always a straightforward matter to distinguish this from an extraskeletal osteosarcoma, osteosarcomas ossify from the center and the definition at the periphery is much more blurred, and they do not stop growing after two months.

— *Histological examination* reveals a zonally structured tissue with, in the center, immature proliferating fibroblasts with slight pleomorphism and significantly increased mitotic activity. Adjacent to these are immature, very cell-rich areas of new bone formation that slowly mature towards the periphery and form mineralized trabeculae surrounded here by typical cubic osteoblasts. In the case of an extraskeletal osteosarcoma, on the other hand, it is the unmaturing sections that are located at the periphery.

— The *treatment* consists of resection, though this should be delayed until maturation is complete, around 1 year later. Recurrences can occur particularly after premature excisions. The resection should be accompanied by the administration of high-dose prostaglandin inhibitors (e.g. indomethacin). In adult patients, low-dose radiotherapy immediately before or after surgery is another option [35]. Spontaneous regressions can also occur after one or two years.

#### 4.5.4.3 Malignant tumors

##### Rhabdomyosarcomas

A rhabdomyosarcoma is the most common malignant soft tissue tumor encountered during adolescence. Overall, it ranks fourth among the malignant soft tissue tumors (after malignant fibrous histiocytoma, liposarcoma and fibrosarcoma). A distinction is made between embryonal, alveolar and pleomorphic forms. The embryonal form of rhabdomyosarcoma, which can be subdivided into botryoid (grape-shaped) and spindle cell forms, occurs between the ages of 1 and 10, while the alveolar variant occurs between the ages of 10 and 25. The very rare pleomorphic variant is observed mainly in adults.

According to the experience acquired by the *Intergroup Rhabdomyosarcoma Study* a classification involving six subgroups would be more clinically relevant: botryoid, spindle cell, embryonal, alveolar, undifferentiated and rhabdomyosarcoma with rhabdoid components [29]. Typical sites are the head and neck, the genitourinary tract, the retroperitoneum and only then the extremities. The embryonal and spindle cells forms usually affect the head and neck, while the alveolar type predominates in the extremities [28].

— *Clinical features*: This is a moderately fast-growing destructive tumor that causes pain and rapidly becomes outwardly visible unless it grows in the abdominal cavity or retroperitoneum.

— The x-ray is not characteristic and calcifications are fairly unusual. Macroscopically it appears as a greenish-pink, usually lobed, tumor. On the MRI scan, the tumor signal is more intense than that of muscle and readily discernible as a rule (■ Fig. 4.54 and ■ Fig. 3.247, ► Chapter 3.2.13).

— *Histologically*, the embryonal type shows densely packed, usually undifferentiated cells with minimal cytoplasm and hyperchromatic nuclei next to varying numbers of cells with eosinophilic cytoplasm which contain glycogen and correspond to the rhabdomyoblasts. In our experience, cross striations are very rarely present and usually occur in more oblong cells containing more cytoplasm. They are readily recognizable however under the electron microscope.

The spindle cell rhabdomyosarcoma is considered to be the embryonal type with the better prognosis. The alveolar form involves weakly differentiated tumor cell aggregates in which »alveolar« spaces can form as a result of the loss of cohesion of the tumor cells. Necrosis is common. Since alveolar rhabdomyosarcomas have often already metastasized by the time of diagnosis they are associated with a poorer prognosis. They regularly show the translocation  $t(2;13)(q\ 37;q\ 14)$  [16]. During immunohistochemical testing the tumor cells react with muscle-specific markers.

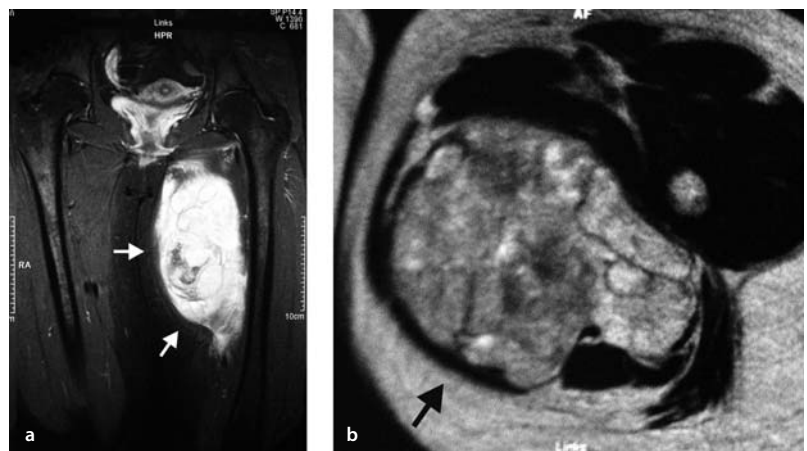
- *Prognosis*: Rhabdomyosarcomas grow very rapidly and aggressively and have a marked tendency to metastasize. The embryonal type has the best prognosis [2].
- The *treatment* consists of a combination of pre- and postoperative chemotherapy and wide resection [9, 18]. This is followed – except for the embryonal type – by radiotherapy [39]. Provided the tumors are located in relatively favorable sites, a five-year survival rate of 80% is achievable with this regimen [40]. The rhabdomyosarcoma is the malignant soft tissue tumor that best responds to chemotherapy [18]. If it has not proved possible to resect the tumor with a margin of healthy tissue, the tumor bed and regional lymph node stations are irradiated. Radiotherapy also has drawbacks however. Secondary tumors are not

especially rare [2, 20, 39]. In a major multicenter study with 1,770 patients with rhabdomyosarcoma, 22 patients with radiotherapy-induced tumors were identified after an average of 8.4 years (1.24%) [18]. On completion of the treatment, further surgical exploration of the tumor cavity is required. Generous biopsies can yield information about the completeness of tumor destruction and the resection and the need for further resection.

### Synovial sarcoma

This biphasic tumor consists of (predominantly) fibroblastic and epithelioid sections. After rhabdomyosarcoma this is the second commonest malignant soft tissue tumor encountered during adolescence, occurring mainly between the ages of 15 and 35.

- *Site*: Very few synovial sarcomas occur within the joints [25]. They are almost always extra-articular, occurring in contact with tendons, tendon sheaths or bursae, and occasionally with ligaments, aponeuroses or fasciae, and preferably affect the lower extremity, particularly the knee, ankle and foot.
- *Clinical features*: Synovial sarcomas are usually painful. A tumor mass of variable size is palpable. Since the tumor grows fairly slowly it is not usually diagnosed until several months or even years have elapsed. On the x-ray calcifications are seen more frequently compared to other sarcomas (in around 40% of cases; [26]). On the MRI scan, the tumor signal is more intense than that of muscle, but weaker than that of fatty tissue.
- *Histologically* a distinction is made between biphasic and monophasic (purely epithelioid or fibroblastic) or weakly differentiated variants. The classical biphasic type shows spindle-shaped fibroblasts with moderate atypia and usually few mitoses. These contain groups, swirls or strands of fairly large cells rich in cytoplasm, readily discernible borders and vesicular nuclei. Char-



■ Fig. 4.54a, b. MRI scans of an *alveolar rhabdomyosarcoma* in the medioposterior femoral muscles of an 18-year old girl. a Frontal, b horizontal section

#### 4.5.4 · Soft tissue tumors

acteristically, they are not surrounded by the reticular fiber network that is typical of the spindle cell component. On immunohistochemical testing they react positively with epithelial markers and show the typical chromosomal translocation t(X;18) with the gene fusion product *SYT/SSX*, which can also be detected by RT-PCR on the paraffin material [15, 27].

- *Prognosis*: Synovial sarcomas have a marked tendency to recur and metastasize, although it can take a long time for the metastases to appear since the tumor grows very slowly. Consequently, the five-year survival rate is not sufficient for evaluating the success of treatment, and the result can only be evaluated after 10 years. The initial size of the tumor has a significant predictive value [7].
- The *treatment* consists of a wide resection with subsequent polychemotherapy and radiotherapy [14, 24]. The regional lymph nodes should also be removed. As with other malignant soft tissue tumors, isolated limb perfusion with specific chemotherapeutic agents (tumor necrosis factor) can prove helpful [23, 37].

#### Extraskeletal Ewing sarcoma

This tumor is much rarer than the osseous form of Ewing sarcoma and occurs primarily in adolescents. It mainly affects the trunk.

- *Histologically* it is indistinguishable from the intraosseous Ewing sarcoma.
- The principal conditions to consider in the *differential diagnosis* are *neuroblastoma*, *rhabdomyosarcoma* and *malignant lymphoma*.
- The prognosis tends to be worse than for the skeletal type.
- *Treatment* is based on the protocol proposed by the cooperative soft tissue sarcoma study (CWS) and is similar to that for a skeletal Ewing sarcoma [21].

#### Infantile fibrosarcoma

Fibrosarcoma mainly affects adult patients over 30 years of age and is rare in children. Congenital cases do exist, however, and these *infantile or congenital fibrosarcomas* usually occur during the first 12 months of life, particularly in male patients, and are located predominantly on the distal parts of the extremities.

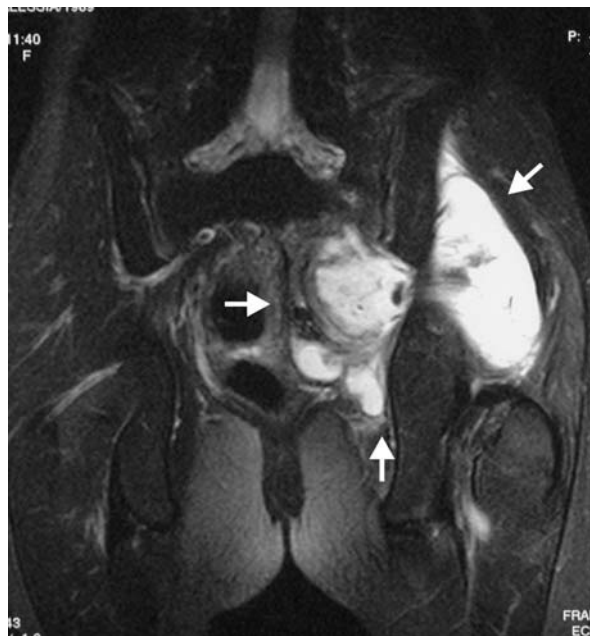
- *Histologically* the tumors consist of usually small, spindle-shaped, densely packed cells surrounded by variable amounts of collagen. The more collagen the tumor contains the more it resembles the adult type of fibrosarcoma. Both forms show numerous mitoses, an important differential diagnostic criterion, in distinguishing them from fibromatoses. In contrast with the adult form, lymphocytic infiltrates are often present, as are highly vascular hemangiopericytoma-like sections.

- The most important differential diagnoses are myofibromatosis, hemangiopericytoma, desmoid tumor and leiomyosarcoma.
- The prognosis for infantile fibrosarcoma is much better than that for the adult form.
- The *treatment* of congenital or infantile fibrosarcoma involves a wide resection. In view of the good primary prognosis, radiotherapy and chemotherapy is appropriate only for non-radically resectable or metastasizing tumors.

#### Malignant peripheral nerve sheath tumor (MPNST) (schwannoma)

Malignant peripheral nerve sheath tumor (MPNST) mainly affects young adults. It occurs as a solitary tumor in adolescents principally in connection with von Recklinghausen disease. Macroscopically they appear as grey-white tumors, usually in connection with a peripheral nerve (■ Fig. 4.55).

- The prognosis for sporadic MPNST is better than for the form associated with von Recklinghausen disease (75% survival compared to 30% recovery [4]). The treatment, where possible, consists of a wide resection. Since the tumors spread along the nerves the surgical procedure can prove problematic. Hardly any information is available about the effect of chemotherapy. Isolated limb perfusion with specific chemotherapeutic agents (tumor necrosis factor«) can prove successful for tumors on the extremities [36].



■ Fig. 4.55. MRI scan of a malignant peripheral nerve sheath tumor in an 11-year old girl, originating from the sacral nerve roots and spreading along the sciatic nerve through the sciatic notch



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### 4.5.5 Therapeutic strategies for bone and soft tissue tumors

» I was once Oscar. The child that no-one talks to because they're frightened of his illness. The child who suffers from the silence of his fellow humans, from the silence of heaven, from all the unanswered questions, but who nevertheless always retains an infinite zest for life. «  
 (Eric-Emmanuel Schmitt, author of the book »Monsieur Ibrahim and the flowers of the Koran« on his new book »Oscar and the lady in pink«, in which Oscar suffers from leukemia).

#### Introduction

The therapeutic strategies for tumors of the locomotor system have experienced *fundamental changes* over the last 20 years. This trend is attributable to:

- a better understanding of the nature of the tumor through *staging*,
- fundamental new developments in *diagnosis*,
- new options for *surgical treatment*,
- improvements in *drug treatment*.

Staging and the diagnostic possibilities were discussed extensively in ► Chapter 4.5.1.

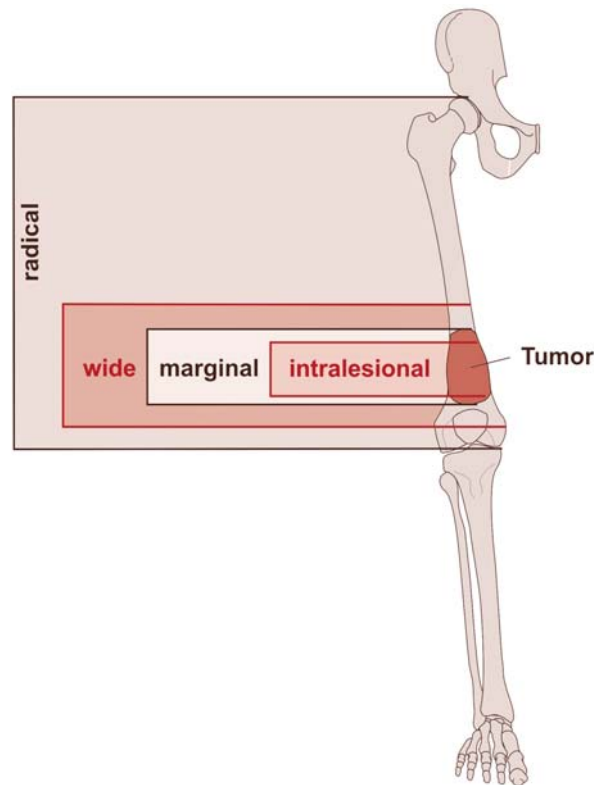
#### Resection

The following basic options are available for the resection of bone tumors, each based on the relationship between the resection margin and the tumor boundaries (► Fig. 4.56):

- *Intralesional resection*: The resection remains within the tumor (example: curettage).
- *Marginal resection*: The tumor is removed as a whole but within the pseudocapsule.
- *Wide resection*: The tumor is removed as a whole, including the pseudocapsule, with a margin of healthy tissue, but the resection remains within the affected compartment.
- *Radical resection*: The tumor is removed as a whole, including the affected compartment (usually feasible only as an amputation).

Another way of qualifying resections is provided by the *R system*, which is based on the investigation of the tissue preparation after resection. If the resected margins are free of tumor this is described as R0. If microscopic tumor residues are observed, the resection is classified as R1. If macroscopic tumor sections remain, the classification is R2.

The choice of resection depends very crucially on the *staging* of the tumor (► Chapter 4.5.1). In principle, any type of resection can be both »conservative« and ablative



► Fig. 4.56. Types of resection: intralesional, marginal, wide, radical

at the same time. Even an amputation through the middle of the tumor is classed as an »intralesional« treatment. On the other hand, a radical resection without amputation is possible in certain particularly favorable situations. But these latter options tend to be more in the realm of theory. In practice, we consider intralesional, marginal and wide resections to be limb-preserving treatments, whereas a radical resection is only feasible in the form of amputation.

#### Basic recommendations on the type of resection in relation to staging of the tumor

These are listed in ► Table 4.19.

#### Remarks on the various resection methods

##### Intralesional resection

Intralesional resection in bone is equivalent to a curettage. *Stage 1 tumors* can be curetted, although a surgical procedure is rarely indicated. The recurrence rate for a simple bone cyst depends not on the completeness of the curettage, but on the activity of the cyst. Nor does a Langerhans cell histiocytosis need to be completely removed by curettage, since a remission of the focus generally occurs after partial removal (or biopsy). Follow-up is required in such cases however.

❗ **For stage 2 and stage 3 tumors, the recurrence rate depends directly on the quality of the curettage. Whereas the recurrence rate for giant cell tumors, aneurysmal bone cysts, chondromyxoid fibromas etc. can be over 50% after curettage by inexperienced surgeons, this figure can be reduced to 10% in treatment centers in which bone tumors are frequently resected [4, 26, 37].**

The high recurrence rate is particularly problematic for giant cell tumor, since this tumor usually spreads through the epiphysis into the joint cartilage. If a recurrence oc-

curs, a subsequent clean resection generally proves to be more difficult than the initial operation. The recurrence rate for aneurysmal bone cyst depends greatly on its activity. Stage 3 lesions require more careful removal than aneurysmal bone cysts classified as stage 2. A curettage can never be complete if it is implemented only with the curettage spoon. The wall of the tumor or cyst always shows indentations and projections and the tumor can also penetrate between the trabeculae. Consequently, the tumor cavity must always be burr drilled with a *special drill* with an angled end for reaching into all the corners. At the end of the procedure we usually illuminate the

■ Table 4.19. Recommendations for the type of resection depending on the tumor stage

Stage	Typical tumors	Resection
<b>Benign, stage 1</b> (inactive)	<b>Bone:</b> juvenile bone cyst, enchondroma, fibrous dysplasia, Langerhans cell histiocytosis <sup>a</sup> <b>Soft tissues:</b> mucous cyst, pigmented villonodular synovitis <b>Bone:</b> osteochondroma <b>Soft tissues:</b> lipoma	(If indicated at all:) intralesional (curettage)  Marginal –
<b>Benign, stage 2</b> (active)	<b>Bone:</b> osteoid osteoma, osteoblastoma, chondroblastoma, chondromyxoid fibroma, aneurysmal bone cyst, giant cell tumor <b>Soft tissues:</b> angioma, glomus tumor	Marginal, poss. intralesional resection
<b>Benign, stage 3</b> (aggressive) (extracapsular, poss. extracompartmental)	<b>Bone:</b> osteoblastoma, chondroblastoma, chondromyxoid fibroma, aneurysmal bone cyst, giant cell tumor, desmoplastic fibroma <b>Soft tissues:</b> desmoid tumor	Marginal, if close to the joint poss. intralesional resection
<b>Malignant stage IA and IB</b>	<b>Bone:</b> chondrosarcoma (grade 1 and 2), parosteal osteosarcoma, central low-grade osteosarcoma, adamantinoma <b>Soft tissues:</b> liposarcoma (grade 1 and 2), hemangiopericytoma, fibrosarcoma (grade 1 and 2)	Wide resection  Wide resection, poss. preoperative radiotherapy, poss. isolated limb perfusion
<b>Malignant, stage IIA and IIB</b>	<b>Bone:</b> osteosarcoma, Ewing sarcoma, primitive neuroectodermal tumor (PNET), chondrosarcoma (grade 3), dedifferentiated chondrosarcoma, malignant fibrous histiocytoma (MFH) <b>Soft tissues:</b> liposarcoma (grade 3 and 4), rhabdomyosarcoma, synovial sarcoma, malignant fibrous histiocytoma (MFH), fibrosarcoma (grade 3)	Wide or radical resection  Wide or radical resection, poss. preoperative radiotherapy, poss. isolated limb perfusion, for synovial sarcoma or epithelioid cell sarcoma also sentinel node biopsy
<b>Malignant, stage III</b>	<b>Bone:</b> osteosarcoma, Ewing sarcoma, primitive neuroectodermal tumor (PNET), chondrosarcoma (grade 3), dedifferentiated chondrosarcoma, malignant fibrous histiocytoma (MFH) <b>Soft tissues:</b> liposarcoma (grade 3 and 4), rhabdomyosarcoma, synovial sarcoma, malignant fibrous histiocytoma (MFH)	Wide or radical resection (often only palliative, then conservative treatment if possible), poss. metastasectomy (particularly for osteosarcoma)  Wide resection, poss. preoperative radiotherapy, poss. isolated limb perfusion, for synovial sarcoma or epithelioid cell sarcoma also sentinel node biopsy

<sup>a</sup> As this tumor can also show aggressive biological behavior, follow-up over many years is required.

tumor cavity with an *arthroscope* and systematically look for tumor residues.

Surgeons have attempted to reduce the recurrence rate still further through the use of *necrotizing substances: liquid nitrogen (cryosurgery), phenol, methyl methacrylate* (e.g. Palacos). Liquid nitrogen and phenol can only be used in enclosed tumor cavities. If a leak is present these liquids can escape into the surrounding soft tissues and cause considerable damage. The drawback with methyl methacrylate is that, once set, it can be very laborious, and occasionally very difficult as well, to remove the hard plug at a later date. On the other hand, large cement plugs (particularly if they are above and close to joints) should not be left in the bone. Because of its hardness and weight, the cement gradually works its way downward, thereby enlarging the cavity and moving closer to the joint (particularly in the distal femur). Since most patients with bone tumors are relatively young, cement plugs should not be left in situ.

The quality of the curettage is much more important than the use of necrotizing substances in achieving a low recurrence rate. Recent studies have shown that equally low recurrence rates can be achieved with and without the use of such aids [4, 26].

#### Marginal resection

This should be attempted for all stage 2 or stage 3 benign tumors and is also usually possible provide the tumor is not located in the epiphysis close to a joint. The resection may be relatively limited and is performed through the pseudocapsule of the tumor. A histological distinction is made between »marginal 1« and »marginal 2«. »Marginal 1« signifies macro- and microscopic absence of tumor, while »marginal 2« means that the resection extended into macroscopically healthy tissue, but that the tumor reached the edges of the resection under the microscope (corresponding to an R1 resection).

#### Wide resection

The wide (R0) resection is now the standard procedure for all malignant tumors and involves the removal of the whole tumor in one piece together with a margin of healthy tissue around the tumor. The incision and the whole access path for the biopsy must also be resected at the same time.

For this reasons, it is very important to plan for the subsequent resection even at the biopsy stage so that the biopsy channel can be resected as well. Since, even before the operation, we can now assess the spread of the tumor in both bone and the soft tissues very precisely with modern imaging methods (particularly CT and MRI), radical resection is no longer required even for high-grade tumors. The margin of healthy tissue cannot be the same width all round. At unproblematic sites this should be 2 cm wide, but in the vicinity of major nerves and vessels may only

be a few millimeters. But even here, the resected fragment should include clearly healthy tissue. It may prove necessary to resect the relevant vessel or nerve with subsequent bridging. This is particularly important for an osteosarcoma. While a compromise is more feasible with a Ewing sarcoma, this must be followed by radiotherapy.

#### Radical resection

In a radical resection the whole compartment in which the tumor develops must be removed. Since high-grade malignant tumors generally spread out of the bone into the surrounding muscles, both the whole bone and all affected muscles must be resected at the same time. In principle, with a few exceptions, this implies amputation. A radical resection used to be required particularly for an osteosarcoma as skip metastases in the bone can occur remotely from the tumor. These are not visible on a normal x-ray and can mean that significant tumor sections are left in the body after a mere »wide« resection. Since the development of modern imaging techniques, particularly the MRI scan, skip metastases are now readily detectable. Nowadays, the borders of the tumor can be assessed much more accurately, thereby dispensing with the need for a radical resection.

In fact, a radical resection is no longer necessary even for high-grade tumors, and the current emphasis is on limb-preserving methods. Nevertheless, amputations are sometimes unavoidable in exceptional cases involving very large, extensive or unfavorably located tumors or recurrences, particularly if major nerves are also affected.

#### Treatment of benign and locally aggressive tumors

Some benign tumors are discovered as chance findings since they produce no symptoms, for example a non-ossifying bone fibroma, an enchondroma, possibly also a solitary bone cyst and fibrous dysplasia. These are *stage 1* tumors. The last two of these tumors or tumor-like lesions are occasionally discovered as a result of a pathological fracture. Provided the patient is free of pain, *non-ossifying bone fibromas* and *enchondromas* do not require treatment. This also usually applies, in the upper extremities, for a *simple bone cyst* or *fibrous dysplasia*.

On lower extremities, however, these conditions pose a greater problem as a result of bowing of the bone (fibrous dysplasia) or fractures very close to the epiphyseal plate (solitary bone cyst), in which case treatment may be indicated, at least for large lesions. The resection itself is not the main concern, but rather stabilization (► Chapter 4.5.3; use of cortisone, methylprednisolone and arming with flexible intramedullary nails). In the soft tissues *subcutaneous lipomas* only need to be removed if they are troublesome. Simple curettage is sufficient for stage 2 osteoid osteomas and osteoblastomas and also for Langerhans cell histiocytosis.

Chondroblastoma, chondromyxoid fibroma, aneurysmal bone cyst and giant cell tumor, by contrast, are more aggressive lesions with a strong tendency to recur unless they are completely removed. This is a particular problem with giant cell tumors, which can form very close to a joint. Very meticulous curettage techniques should be employed, possibly supplemented by the use of necrotizing substances. A marginal resection should be attempted for favorably located (not near a joint) tumors. In the soft tissues this also applies to angiomas and glomus tumors.

Stage 3 tumors are essentially the same as stage 2 tumors, but simply grow more eccentrically and more aggressively. A marginal resection should always be attempted and, if the tumor is very close to a joint, necrotizing substances administered. Since the recurrence rate for these tumors is very much lower in a specialist treatment center than in a hospital with very limited experience of surgery for such lesions, these tumors should be treated in a center. Of the soft tissue tumors, the *desmoid* falls into this category. A marginal resection frequently leads to a recurrence. For tumors located on the extremities, but not too close to the trunk, very intensive exercise therapy can lead to a diminution in the size of the tumors (► Chapter 4.5.4). Intralesional excisions, however, result in recurrences at increasingly shorter intervals, since the tumor reacts to the surgical *trauma* with proliferation. Radiotherapy may be indicated in cases that are not fully operable [19, 21].

#### Treatment of low-grade malignant tumors

This group includes *chondrosarcoma*, *periosteal osteosarcoma* and *adamantinoma*. These are usually stage IA or (rarely) IB lesions. All these tumors tend to occur in adulthood and are rare in adolescents. While they grow slowly and metastasize at a late stage, they are largely insensitive to cytotoxic drugs and radiotherapy. Treatment is therefore exclusively surgical. The patients usually have a good chance of survival provided the tumor is not too large or located in an unfavorable site – in the spine for instance – and has been correctly removed with a margin of healthy tissue. Where possible, these tumors should also be resected with a wide margin of healthy tissue, while a marginal resection may be sufficient at problematic sites (close to joints, major vessels and nerves).

*Isolated limb perfusion with tumor necrosis factor* may be appropriate for soft tissue sarcomas in this category (e.g. *synovial sarcomas*, *epithelioid cell sarcomas*, *liposarcomas*, *peripheral malignant nerve sheath tumors*) located close to major nerves and no more proximal than the lesser trochanter (lower extremity) or the deltoid insertion (upper extremity) [38]. In this technique, the blood supply to the tumor is isolated and treated with high doses of a cytotoxic drug prior to the resection.

A *sentinel node biopsy* is also worthwhile for synovial and epithelioid cell sarcomas. In contrast with all other malignant bone and soft tissue tumors, these sarcomas can metastasize via the lymph node stations. In this technique a radioactive substance is injected into the tumor and a subsequent bone scan then shows the uptake in the regional lymph node stations.

#### Treatment of high-grade malignant tumors

##### Bone tumors

The principal tumors in this group in relation to children and adolescents include the *conventional osteosarcoma*, the *Ewing sarcoma* and the *primitive neuroectodermal tumor (PNET)* and, among the soft tissue tumors, the *rhabdomyosarcoma*. These are classified as stage IIB or III. Stage IIA (intracompartmental) is very rare.

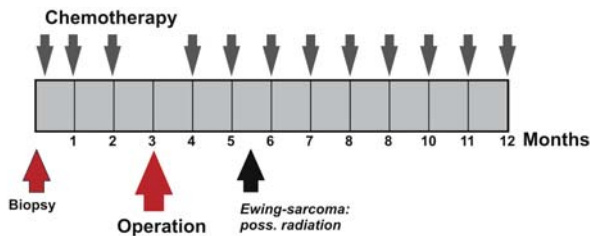
Towards the end of the 1970's a new strategy for *drug treatment* was introduced [33]. Whereas, in the past, surgeons tried to curb tumor development after operative removal of the tumor by administering moderate doses of cytotoxic drugs, it was subsequently realized that the tumor could largely be destroyed with doses almost 1,000 times higher. The effect of the highly toxic cytotoxic agents (particularly methotrexate) could then be cancelled again shortly after its administration by an antidote (folic acid), thereby avoiding major damage outside the dividing tumor. Nevertheless, the side effects can be substantial, and the chemotherapy-related complications (infections, heart failure, etc.) are associated with a mortality rate of approx. 3%.

The current therapeutic strategy (■ Fig. 4.57), after confirmation of the diagnosis by means of a biopsy, is to largely destroy the tumor and its metastases over a period of three months with a combination of various *cytotoxic drugs* at extremely high doses. The chemotherapy involves a combination of methotrexate and other drugs in very high doses, i.e. doxorubicin, cyclophosphamide, cisplatin and dactinomycin. After three months the tumor is surgically removed. The subsequent histological examination of the tumor then shows how much of the tumor has been destroyed by the cytostatic treatment. If over 90% of the tumor is necrotic, this means that the response to the drug has been good (*good responder*), and it can also be assumed that the metastases (predominantly in the lungs) have been destroyed.

The same chemotherapy combination is continued for a further 9 months. If the tumor has not responded well however (*poor responder*), the composition of the drugs is modified. This therapeutic protocol is now followed, subject to minor modifications, in all major centers worldwide.

The therapeutic principle is very similar for osteosarcomas and Ewing sarcomas, except that the surgical removal of the Ewing sarcoma can be followed by *radiotherapy* (particularly after a marginal resection).

## 4.5.5 · Therapeutic strategies for bone and soft tissue tumors



■ Fig. 4.57. The current *therapeutic strategies* for osteosarcoma and Ewing sarcoma are very similar. After the diagnosis is confirmed, chemotherapy is administered in three cycles at monthly intervals. The tumor is then resected. The histological examination shows the efficacy of the chemotherapy. If it shows a good response, the same chemotherapy regimen is continued for a further nine months. If the response is poor, different chemotherapeutic drugs are tried. Radiotherapy is usually prescribed in addition for Ewing sarcomas

Preoperative radiotherapy may also be indicated for tumors in unfavorable sites. 30–40 Gy are administered for this preoperative radiotherapy, whereas 60–70 Gy are required for the tumor radiotherapy. The combination of preoperative radiotherapy and hyperthermia has proved effective. The hyperthermia sensitizes the tumor to the radiotherapy (and also to the chemotherapy incidentally). The drawback of the radiotherapy is that it increases the bleeding tendency during the resection and the postoperative infection risk and that bony bridges show poorer osseointegration.

In Basel, osteosarcomas are treated according to the COSS (Cooperative OsteoSarcoma Study) protocol and Ewing sarcomas according to the EICESS (European Intergroup Cooperative Ewing Sarcoma Study) protocol [3, 30, 35]. These are controlled international studies that coordinate the treatment of these tumors in Germany, Austria, Italy, Great Britain and Switzerland. Recently these protocols have been integrated into the European and American Osteosarcoma Study Group (EURAMOS) and the EUROEWING study group). The tumors are treated according to standardized guidelines and evaluated in a coordinated manner. This is the only way of assessing and continually improving the effectiveness of treatment for these relatively rare tumors. These European studies are now being coordinated with American protocols.

The response to chemotherapy depends on whether *resistance* to the respective drugs is present or not. The causes of the development of resistance are the subject of intensive research. In particular, the presence of the P-glycoprotein, a membrane protein of the tumor cells, appears to be related to the development of resistance.

### Soft tissue tumors

The most important high-grade malignant soft tissue tumor during childhood and adolescence is rhabdomyosarcoma (▶ Chapter 4.5.4.3). This is treated according to the

*CWS-86 protocol* («Cooperative WeichteilSarkomstudie» = cooperative soft tissue sarcoma study). This study coordinates centers in Germany, Austria, Switzerland, Sweden, Poland and Hungary. The treatment regimen is similar to that for Ewing sarcoma. The 3-month period of neoadjuvant chemotherapy (possibly with preoperative radiotherapy depending on the site) is followed by resection and continuation of the drug treatment for a further six months. The response to the chemotherapy is graded as for bone sarcomas.

### Prognosis

Before the arrival of the new era, i.e. before 1977, the probability of survival for osteosarcoma was, at best, 20%. The hope at the time was simply to detect the tumor at an early stage and remove it completely with a margin of healthy tissue by means of amputation. The prognosis for Ewing sarcoma was even worse. Ewing sarcomas form metastases at a very early stage, and the surgical treatment was ineffective because metastases had always developed by the time of diagnosis. Treatment was limited to what we would now consider to be excessively low-dose chemotherapy and radiotherapy.

■ Fig. 4.58a presents the survival rates for patients with *osteosarcomas* now and 15 years ago. The mortality rate is highest during the first two years. Only a small proportion of patients die during the following few years, and as many as 72% are still alive after six years, compared to just 14% in the 1970's. The figures for these graphs are based on several large-scale American and European studies [3, 30, 35]. Overall, the data cover a sample of more than 1,000 patients. The cases treated in Basel have likewise been treated according to the COSS protocol since 1982. By 1998, over 1,700 patients in total had been treated according to this regimen.

The results are generally stated in the form of six-year survival rates. The following *factors* have proved to be the most significant in respect of their influence on the prognosis: The most important questions are whether *metastases* were already present at the time of diagnosis, whether the tumor has been removed *with a margin of healthy tissue* and whether the tumor *responded well or not to the chemotherapy*. The *size* and *site* of the tumor at the time of diagnosis also significantly affect the prognosis [3]. Thus, tumors located in favorable sites such as the upper arm or lower leg, and particularly if they respond well to the primary treatment, are now associated with a survival probability of over 90%. Even tumors on the thigh, the commonest site of the osteosarcoma, have a survival rate of 58%.

The situation is not so favorable for *Ewing sarcoma* because of the early formation of metastases. Nevertheless, a six-year survival rate of around 50% can still be achieved for tumors affecting the extremities (■ Fig. 4.58b) [35]. Survival probabilities of over 80% can be expected for



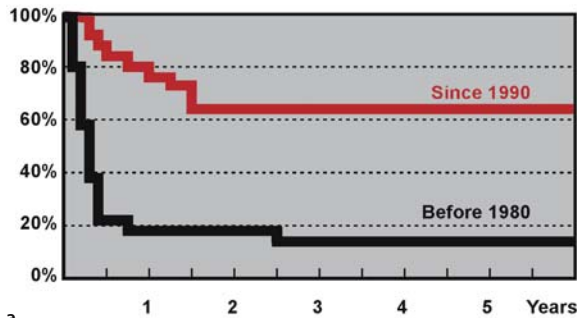
soft tissue tumors (*rhabdomyosarcomas*) depending on their location and provided the primary tumor does not metastasize [9].

### General aspects of the surgical treatment of malignant tumors

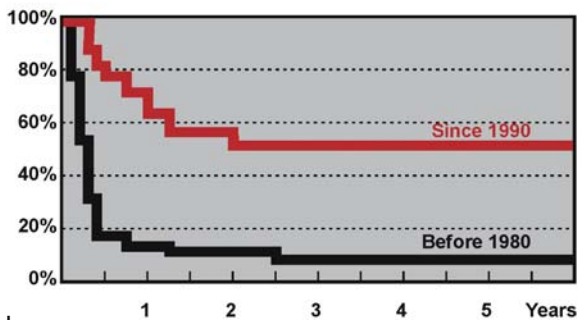
The goal of resection, whether *wide* or *radical*, is always the complete removal of the malignant tumor. As a rule this can be achieved by a limb-preserving technique

or by amputation. The spread of the tumor within or outside the compartment is a key factor when determining the appropriate treatment. Nowadays, amputation is unavoidable in just over 10% of cases, and is indicated particularly if major vessels and nerves are surrounded by the tumor.

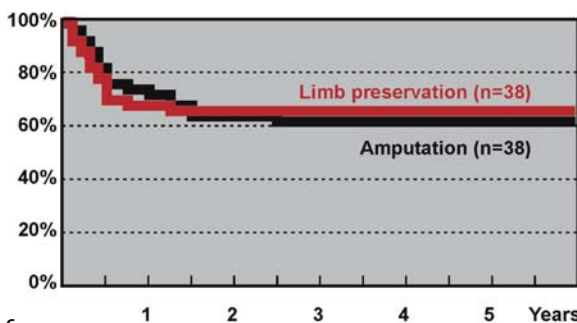
Thanks to imaging procedures we usually have very accurate information about the extent of the tumor prior to its removal. The resection must be planned very carefully as absolutely no compromise is possible. If necessary, important soft tissue structure must also be removed at the same time. The tumor should not be touched throughout the operation (*»it should not see the light of day«*). The access routes of earlier operations – for example biopsy channels – *must likewise be removed*. This is why the plan for the subsequent resection and reconstruction must be known even at the time of the biopsy, since a badly located biopsy scar can make it impossible to subsequently remove the tumor with a margin of healthy tissue without amputation. Depending on the spread of the tumor in each case, further specialists may need to be called in during the resection in order to perform a vessel- or nerve-bridging procedure or skin reconstruction. Close cooperation with vascular surgeons, plastic surgeons, and possibly neurosurgeons, is absolutely essential in a tumor center.



a



b



c

■ Fig. 4.58a–c. The two curves in **a** show the *six-year survival rates* before and after the introduction of the modern chemotherapy protocols for *osteosarcomas*. The probability of survival is currently over 60%, compared to less than 20% before 1980. Most fatalities occur during the first two years. The curves were produced on the basis of figures compiled from several European and American studies. **b** The survival rates for *Ewing sarcoma* are not as good, overall, as those for *osteosarcoma*, since the former metastasizes at a very early stage. Here too, the chances of survival have improved considerably since the introduction of the new treatment protocols. These curves were also based on figures from several European and American studies. **c** The same survival rate can now be achieved with *limb-preserving treatment* as was formerly possible with *amputation*

### Bridging options

The following options are available for bridging a bony defect:

- *autologous bone* from another site (e.g. the fibula, a rib or bone graft from the iliac crest) or locally (after autoclaving or irradiation),
- Insertion of *artificial bone and joint components* made from plastic and metal,
- *allogeneic bone* (foreign bone from another individual),
- *combination of joint prostheses with autologous or allogeneic bone.*

### Autologous bone

The use of autologous bone is the least problematic method in terms of subsequent osseointegration. The patient's own bone does not cause any rejection reactions, and the resulting healing process does not usually cause any problems. The disadvantage of this method is that joint sections can be replaced only in exceptional cases.

■ Fig. 4.59 and 4.60 show examples of reconstructions with autologous fibula. The removal of the fibula, leaving the most proximal and distal sections in situ, results in minimal impairment of load-bearing capacity and function of the lower leg. Whether the healing of the fibula at its new

site can be improved with a vascular pedicle remains a matter of dispute. Some experiments have shown hypertrophy occurring only with vascularized grafts, not with free grafts [17].

However, clinical experience has long demonstrated that hypertrophy also occurs to a comparable extent with non-vascularized grafts [25] (■ Fig. 4.60, 3.254, ► Chapter 3.2.13). Moreover, fibular regeneration at the old site is much better after removal without a vascular pedicle and thus without periosteum as well (■ Fig. 4.60f), reaching its original thickness even after just one year. The healing at the new site is also generally unproblematic without a primary vascular connection, at least in adolescents (■ Fig. 4.59 and ■ Fig. 4.60). Once the fibula has healed, remodeling occurs over time with functional weight-bearing, causing the transplanted fibula to adapt itself to the original bone.

If a *joint* is also affected, the reconstruction is much more difficult. Particularly in the case of children, the surgeon must try and manage without the use of an artificial joint. The two examples shown in ■ Fig. 3.250 and 3.251 (► Chapter 3.2.13) and in ■ Fig. 3.525 and 3.526 (► Chapter 3.5.7) show what can be achieved, although replacement with the patient's own bone may be required in some cases. The example in ► Chapter 3.5.7 also shows how the proximal humerus can be replaced by the clavicle.

If the proximal humerus has to be removed, together with the axillary nerve, as a result of a tumor, the implantation of a prosthesis is not a good solution since stable joint function is not possible without innervation of the deltoid muscle. In accordance with Winkelmann's [40] proposed technique, the clavicle was released from the sternum and folded down in the acromioclavicular joint. The clavicle then forms the proximal humerus, while the

acromioclavicular joint replaces the shoulder. While active shoulder mobility is, of course, restricted, the passive mobility is relatively good.

Another example of the resection of a tumor with a margin of healthy tissue despite joint involvement is presented in ► Chapter 3.2.13 (■ Fig. 3.250 and 3.251). This female patient had an Ewing sarcoma of the pelvis, and half of the acetabulum had to be removed at resection, thereby interrupting the continuity of the pelvic ring. Anchoring a substitute material in the pelvis is particularly problematic because of the relatively soft bone and the prevailing shear forces. Loosening of the implant rapidly occurs, and this can result in almost insoluble problems, particularly in young patients. The hip joint was transferred to the sacrum by rotating the acetabulum (■ Fig. 3.250 and 3.251, ► Chapter 3.2.13). Once the acetabulum has integrated with the surrounding bone, a stable situation is produced for the long term. While this procedure is associated with the drawback of leg shortening, this can be corrected at a later stage.

One special technique for using the patient's own bone is to remove the tumor together with the adjacent bone, sterilize the resected tissue either by boiling or irradiation in order to kill the tumor cells, and then *reinsert the resected tissue*. Whereas bone is severely destroyed by autoclaving, extracorporeal irradiation is very promising and represents a very effective alternative to prostheses and allografts, particularly for pelvic tumors [24]. The irradiated bone must be used in combination with an artificial joint since the devitalized cartilage is no longer usable as joint cartilage. Irradiated autologous bone, on the other hand (as with the non-vascularized fibula) appears to be revitalized. A precondition for this technique is that the bone should be mechanically stable enough, although this is often not the case depending on the individual tumor.

### Prostheses

As a rule, any bone or joint can be replaced by a prosthesis, which will need to be tailored to the needs of the individual patient depending on the resection. The main problem lies in anchoring these prostheses in healthy bone. The larger the removed fragment and thus the greater the lever action of the prosthesis, the more difficult will be the anchorage and thus the greater the likelihood of early loosening of the implant. Young, active patients in particular place a greater strain on their prosthesis than older patients. In this case, the problem of the unfavorable mechanical situation for tumor prostheses is compounded by the increased loading resulting from the higher activity level of the patient. Nevertheless, prostheses do not only help to avoid the amputation, they can, in many cases, largely preserve the function of the affected part and thus improve the patient's quality of life [28, 32].



■ Fig. 4.59. Left 15-year old boy with *Ewing sarcoma* in the iliac wing. Right Two years after resection of the tumor and reconstruction with autologous fibula. The patient is able to walk without a limp and jump, and regularly plays tennis



**Fig. 4.60a-f.** 5-year old boy with malignant hemangiopericytoma. **a** AP x-ray of the proximal femur. **b** Situation after wide resection and bridging with autologous fibula (without vascular pedicle). **c** One year later, a fracture of the fibula has occurred at the proximal end. **d** By the age of 7 years, a femur has already formed, albeit with a pseudarthrosis at the proximal end, which was stabilized with intramedullary flexible nails. **e** Situation at the age of 16 years. The pseudarthrosis was corrected with an angled blade plate and a vascularized iliac graft. A femur of normal thickness has developed from the fibula, and the leg length discrepancy is just 1.5 cm. **f** Four years postoperatively the original fibula has completely regenerated



#### 4.5.5 · Therapeutic strategies for bone and soft tissue tumors

Shaft implants can also be used [2]. In tumor patients we use a modular prosthesis system. In the past we used the system developed by Kotz (now called Global Modular Resection System, GMRS), which offers all sizes of implants for bone and joint sections of the lower extremity [28]. More recently we switched to the more versatile Modular Universal Tumor and Revision System (Mutars®).

The example in **Fig. 3.342** (▶ Chapter 3.3.12) shows a tumor resection in a distal femur and its replacement with a tumor prosthesis. A total femur replacement is shown in **Fig. 4.61**, and long-term results are now also available for GMRS type prosthesis: After 10 years, the implant has not loosened in 96% of proximal femurs, 76% of distal femurs or 85% of proximal tibias [28]. 88% still function well. The new generation of modular prostheses facilitates knee rotation in two planes, and includes the Global Modular Replacement System (GMRS), the Rotating Hinge Knee or the particularly versatile Modular Universal Tumor And Revision System (MUTARS). The latter type of prosthesis is also available in a silver-coated version, which is very effective in infection scenarios. A woven dacron tube surrounding the prosthetic joint facilitates the ingrowth of muscles.

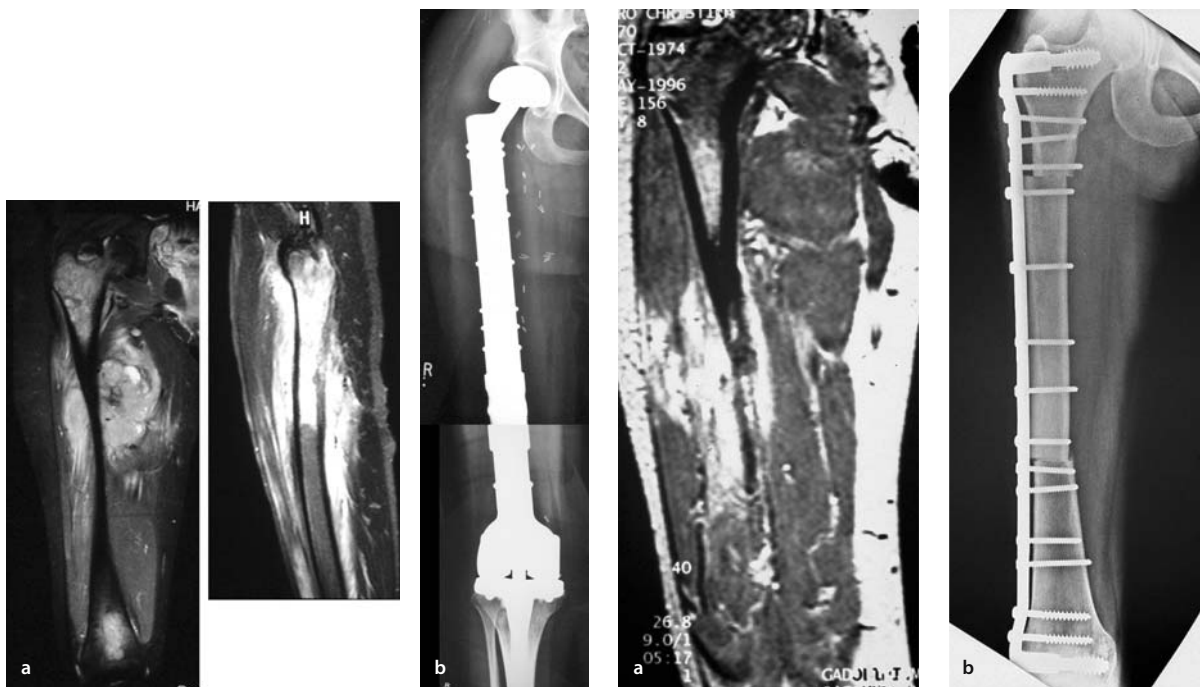
Special prostheses for use in the shaft but without a joint section are also available.

#### Allogeneic bone (allograft)

The third bridging option is to use foreign bone. Whereas the use of vital foreign bone is still in the experimental stage because of the rejection reactions and the need for immunosuppression, deep-frozen, devitalized grafts have been in use for some time [10, 18]. Such grafts must be stored for at least two months at  $-80^{\circ}$ , by which time the bone and cartilage are completely dead and rejection reactions are no longer expected.

We make a basic distinction between the replacement of exclusively bony sections (**Fig. 4.62**) and replacement involving joint sections (**Fig. 4.63**). While the allogeneic bone unites with the patient's own bone in the contiguous areas, most of the graft remains devitalized and is not integrated by the body.

The use of joint sections is problematic since the cartilage is dead and the periosteum does not have a nerve supply. Such joints degenerate after a short time. If a joint has to be removed with a large fragment of bone shaft, the *combination of allogeneic bone* for the shaft and a *joint prosthesis* has proved effective. The advantage of this method over the exclusive use of a prosthesis is that the foreign bone unites with the patient's own bone over the course of time, at least around the edges, and that the muscles can then anchor themselves more permanently compared to a metal implant. This also reduces the problem of prosthesis



**Fig. 4.61a, b.** 19-year old female patient with *Ewing sarcoma* of the proximal half of the femur and skip metastasis in the distal metaphysis. **a** MRI views of the right femur, frontal and sagittal. **b** AP x-ray with total femoral replacement (Mutars system) after resection of the femur

**Fig. 4.62a, b.** 18-year old girl with *Ewing sarcoma* in the center of the shaft. **a** MRI scan of the right femur (sagittal section). **b** After resection and intercalary bridging with an allograft. A special plate was prepared with angularly-stable dynamic screws on both ends





■ **Fig. 4.63a, b.** 11-year old male patient with *fibrocartilaginous mesenchymoma*. **a** AP x-ray and frontal MRI. **b** Two years after bridging with osteoarticular allograft

»vibration«. The use of the dacron tube, however, has reduced the need for this combination of a joint prosthesis with allogeneic bone as the dacron tube allows anchorage of muscles to the prosthesis.

■ Fig. 4.64 shows an example of a patient with a tumor resection in a proximal femur and the combined replacement with allogeneic shaft bone and a tumor prosthesis. Allografts are associated with high complication rates: fractures in 19% of cases, lack of bony union in 15%, infections in 11% [16].

#### **Specific aspects of resection and the bridging of tumors close to the physes during growth**

If a malignant tumor together with the epiphyseal plate is removed from a growing child, an increasing length



■ **Fig. 4.64a, b.** 14-year old male patient with *Ewing sarcoma* in the proximal femur. This tumor was also removed with a margin of healthy tissue. The reconstruction involved a combination of a specially prepared *hip prosthesis* and an *allogeneic bone graft* that replaced the whole upper section of the femur apart from the hip. On the one hand, the graft improves the anchorage of the implant while, on the other, the muscle insertions are more permanently anchored with the replaced bone section than would be possible with a purely metal implant

discrepancy in relation to the other side will become apparent. This problem is very serious particularly if the lower extremities are involved since differences of 10 cm or more can result, especially if the most active epiphyseal plate, i.e. in the distal femur, has to be removed. Basically, the *problem of growth cessation* after resection of a physis along with the tumor can be approached in the following ways:

- concurrent *closure of the epiphyseal plate on the other side*,
- *rotationplasty* according to Van Nes,
- use of an *extendable prosthesis*,
- conventional approach (insertion of an allograft, and possibly a prosthesis) and *leg lengthening* at a later date.



Large tumors close to a joint are usually restricted to a single bone as joint cartilage constitutes a very effective tumor barrier. For this reason, only one side of the joint needs to be resected in almost every case. If a tumor prosthesis is implanted, however, both joint-forming bone surfaces must be replaced, thereby destroying the epiphyseal plate of the bone on the other side of the joint. A knee prosthesis was therefore developed whose tibial anchorage was designed so as to allow the proximal tibial epiphyseal plate to continue growing [34]. This problem does not exist if allogeneic bone grafts (allografts) are used since the healthy part of the joint does not need to be replaced.

#### Concurrent closure of the epiphyseal plate on the other side

This is by far the simplest way of avoiding a major leg length discrepancy in children over 10 years who are predicted to grow fairly tall (over 5 ft 9 in or 175 cm). At a bone age of 10 years, the distal femur is expected to grow a further 6.5 cm in boys and 4.5 cm in girls, on average, while the anticipated further growth in the proximal tibia under the same conditions is 4.5 cm in boys and 2.7 cm in girls (■ Fig. 4.18, ► Chapter 4.2.2). Since patients with osteosarcomas tend to be fairly tall [27], this method can often be used in adolescents. The patient is spared from having to undergo other high-risk and stressful procedures at the cost of a loss in final height. The morbidity associated with the physal closure technique employed by us (► Chapter 4.2.3) is minimal, and the closure can be

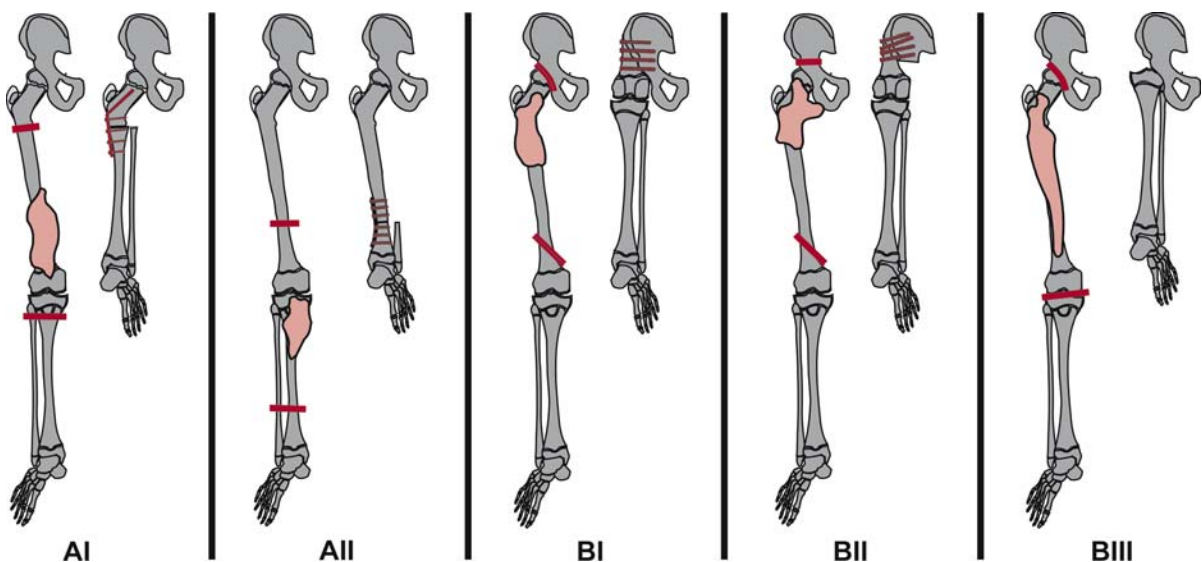
performed at the same time as the tumor resection. After two weeks full weight-bearing on the affected leg can be resumed.

#### Rotationplasty

The technique of rotationplasty was first mentioned by Borggraevé in 1930 [5] in relation to the treatment of a proximal femoral deficiency. A comprehensive description for the same indication was provided in 1950 by Van Nes [39], hence the more usual term in the English-speaking literature of »Van Nes rotationplasty«. The rotationplasty is a special form of amputation in which the upper ankle is rotated through 180° and functions as a knee. It was originally used for osteosarcomas of the distal femur as a way of improving function after tumor resections. Rotationplasties were also subsequently developed for tumor resections on the proximal tibia and proximal femur [41] (■ Fig. 4.65).

Even though the rotationplasty is not a limb-preserving method it does provide much better function compared to a standard amputation. Gait analyses have shown that children with rotationplasties can walk much faster than after amputations or arthrodeses [6, 20, 23, 32, 36]. They can even jump and climb stairs. The result is functionally equivalent to a lower leg amputation and can also be achieved by this method for tumors of the upper leg.

The problems are more of a psychological nature. The backward-facing foot (■ Fig. 4.66) is not accepted equally well by everyone. But it is not so much the patients themselves but rather the relatives that have trouble coping



■ Fig. 4.65. Types of rotationplasty according to Winkelmann [40]: Type AI for a tumor on the distal femur, Type AII and Type AI for a tumor on the proximal lower leg, Type BI for a tumor on the proximal femur,

Type BII for additional involvement of the muscles near the pelvis, Type BIII for a tumor affecting the whole femur



**Fig. 4.66a–c.** Example of a *rotationplasty* in a 9-year old boy with an osteosarcoma on the distal femur. **a** Clinical appearance 6 months postoperatively. **b, c** Demonstration of active mobility in the ankle that now acts as the new knee

with this situation. Since experience has been gained with hundreds of rotationplasties worldwide, this operation has established itself as a standard method for tumors close to the growth plate in children under 10 years of age [20, 23, 32, 41]. The chances of participating in athletic activities for several decades of life are better by far with a rotation plasty than with a tumor prosthesis.

#### Extendable prosthesis

Another option for bridging after resections during growth is the use of extendable prostheses [2, 11, 13, 14] that have been developed in certain centers. In addition to the drawbacks of a bulky prosthesis for a growing child, all are associated with the additional problem of the need for a new operation, with the corresponding risks, for every extension. Furthermore, since the shaft of a long bone grows not only in length but also in diameter the anchorage can loosen simply as a result of growth. Recent investigations, however, have shown a high complication rate, but a loosening frequency comparable with adult prostheses [2, 11, 14]. A recent innovation is the MUTARS prosthesis with an externally controlled extendable

section incorporating an electric motor in the manner of the Fitbone medullary nail. Initial results are encouraging although no firm recommendations can be provided at this stage.

#### Conventional approach with leg lengthening at a later date

This approach is a simple and logical way of equalizing the leg lengths. The main problem is the need for an external fixator for the lengthening process. Since this is anchored in the bone transcutaneously infections repeatedly occur. Large prostheses or allogeneic bone grafts are already at risk of infection, and the extension involves a not insubstantial risk of secondary infection.

#### Treatment of bone and soft tissue tumors – a multidisciplinary task

The objective of our efforts is to preserve the physical, and thus also the mental, integrity of the patient. Many specialists are involved in the decision concerning the appropriate treatment and its implementation. Since malignant bone tumors are rare the necessary experience

#### 4.5.5 · Therapeutic strategies for bone and soft tissue tumors

for this task can only be found in major *centers*. The team includes, in addition to the orthopaedic surgeon, an oncologist, radiologist and bone pathologist. All of these specialists should – if not exclusively at least primarily – be working in the field of bone tumors. In specific cases, a radiotherapist and other surgical specialists should be available, e.g. vascular, micro- and neurosurgeons and plastic surgeons.

In Basel, we are in the fortunate position of possessing all these specialists and being able to work closely with them. A particularly useful diagnostic resource at our disposal is the *bone tumor register*, which was set up in 1972 by the Basel Institute for Pathology and currently includes over 11,000 bone tumors and tumor-like lesions. Misdiagnoses can have fatal consequences. This particularly applies to bone tumors, since they are not only rare, but also subject to substantial variability in terms of their appearance and prognosis. Underestimating a tumor can lead to the death of the patient, while overestimating a tumor can result in the unnecessary sacrifice of major sections of an extremity.

A particular risk is inherent in the fact that bone tumors represent a »challenge« to the surgeon in both diagnostic and therapeutic respects. There is a great temptation to want to treat bone tumors oneself, despite a lack of experience. The saying »*good results come from experience – experience comes from bad results*« is particularly applicable to tumor treatment. It must also be acknowledged that *complications* also commonly occur, and are often unavoidable, even in major centers. Thus, it repeatedly happens that a resection does not include a full margin of health tissue despite a meticulous procedure. Moreover, the loosening rate for tumor prostheses and the infection rate associated with large allogeneic bone grafts are very high (approx. 11% [16, 29]).

*Psychological support* for the patients is an important part of the treatment. It is important to talk openly and honestly also with children and adolescents about their illness and prognosis. The affected children should be present in all discussions with the parents. Only then will you be able to gain their trust and give them the confidence needed to cope with these kinds of extensive treatment (see the quote at the start of ► Chapter 4.5.5). Much experience is required on the part of all those involved in order to manage the patients in the right way. Our team therefore includes a psycho-oncologist. Joint activities are organized for young tumor patients, e.g. annual holidays in Greece.

Another important aspect is the *cost issue*. A current treatment involving expensive diagnosis, chemotherapy and limb-preserving surgery can cost over 100,000 dollars per patient, whereas the former treatment of amputation and an early death were much »cheaper«. In the current discussion about the so-called »cost explosion« in health-care we should bear in mind that medical innovations

often lead to substantial increases in costs. But in return we can offer – at least in respect of bone tumors – a considerable improvement in life expectancy and quality of life, and this at an age when no-one can ask whether this is still »useful«.

**!** **The primary aim for the *clinical practitioner* is not to overlook a bone tumor. To this end, one simple rule should be borne in mind: If a patient complains of one-sided pain in the extremities that persists for several days or weeks and is not clearly activity-related (particularly if it occurs at night), an x-ray should always be arranged. If a bone tumor is suspected, the patient should be referred, if possible directly, to an appropriate center.**

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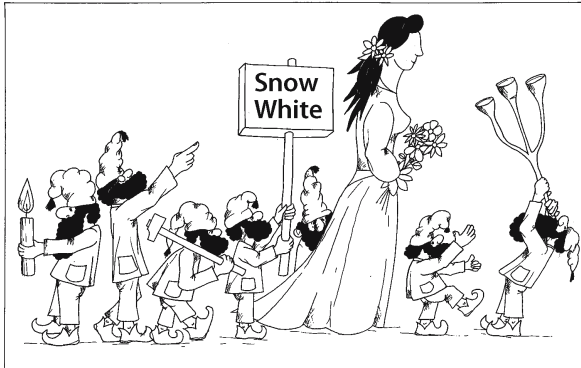
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## 4.6 Hereditary diseases

### 4.6.1 Of beggars and artists and clues in the quest for appropriate classification – Introduction

» *A dwarf standing on the shoulders of a giant can see farther than the giant himself.* « (J.J.W. Heinse)



The term »systemic diseases« covers all those illnesses that are not restricted to a body region or individual organ, but that affect a whole organ system. Pediatric orthopaedics, of course, is primarily concerned with those diseases in which the musculoskeletal system is (also) affected. Almost all these conditions are hereditary (i.e. genetically induced), and most are associated with small stature or dwarfism. Exceptions do exist, however, in this group that is extremely heterogeneous in terms of etiology, pathogenesis and manifestation. Thus, for example, Poland syndrome is not inheritable, and patients with Marfan syndrome can grow very tall.

*Small stature* is a common characteristic of many hereditary diseases. If a final height of less than 150 cm (4 ft 11 in) is reached we speak of »dwarfism«. *Historically speaking* Aristotle (384–322 BC) was probably the first person to write about dwarfism [3]. He also postulated theories about its cause (excessively small womb, poor diet). In the Middle Ages, the Dominican friar Albertus Magnus (1193/1206–1280) addressed this subject, describing a case of a 9-year old dwarfish girl who had not reached the size of a 1-year old. He attributed the deformity to the fact that only a small proportion of the father's seed had entered the mother's womb.

In his »Monstrorum historia« Ulisse Aldrovandi (1522–1605), naturalist and professor of medicine at Bologna, differentiated between mythical creatures and dwarves. Felix Platter (1536–1614), a city physician of Basel, made the important observation that, in addition

to genuine small stature, a diminished stature can also result when the spine collapses as a result of abnormal curvature (scoliosis or kyphosis) thus making the trunk too short, or when the lower limbs are shortened by deformation.

It was Isidore Geoffroy Saint-Hilaire (1805–1861), a Parisian zoologist, who founded the study of modern teratology with his »Histoire générale et particulière des anomalies de l'organisation chez l'homme et les animaux, les monstrosités, des variétés et vices de conformation«, which marked the start of the scientific examination of abnormalities and inherited diseases [3]. In the following description of the individual clinical conditions, the person that first described the relevant illness is mentioned in each case.

Dwarfs have always had a special significance in *human cultural history*. In Ancient Egypt, at a time when incest was not yet considered taboo (► Chapter 1.2), hereditary diseases were extremely common. That dwarfism was socially perfectly acceptable is demonstrated by the fact that some were even honored as gods (e.g. the gods Ptah and Bes). Deformed individuals in the ancient world were termed »monsters«. Although this term now has very negative connotations, dwarfs were rarely viewed as disagreeable individuals in historical times.

Dwarfs often used to work as beggars, at a time when beggars were not viewed as they have been since the start of the 18th century. Provided there was no urgent need to work in order to make a living, it was possible, particularly in Southern Europe, for a person with minimal needs to manage tolerably without working. Begging was a legal way of earning one's living. The beggar simply claimed his modest share of what others may have earned not necessarily as payment for work. Today's »work ethos« is only a phenomenon of the modern era.

The beggar was no more despised than other members of the lower social strata. In Ancient Greece he was under the specific protection of Zeus, while in the Middle Ages he was treated with respect and kindness, and even particularly honored if a mental defect could be interpreted as a prophetic talent. Popular belief ascribed to beggars the ability to cause harm, by their look or words, to those who were not willing to give. The fact that the courts of kings and rulers would regularly feed dwarfs and beggars and often hold open house for them on certain days is reflected in fairytales and ballads and shows that they were considered as bringers of good luck [4].

In the 18th century, dwarfs and deformed individuals were increasingly put on display in show booths and later in circuses. Even today, dwarfs (usually those with achondroplastic dwarfism) form part of the attraction in almost every circus.

The *world of fairytale and fantasy* is full of dwarfs, in most cases in a positive context. Thus, the seven dwarfs help Snow White before the prince manages to save her.

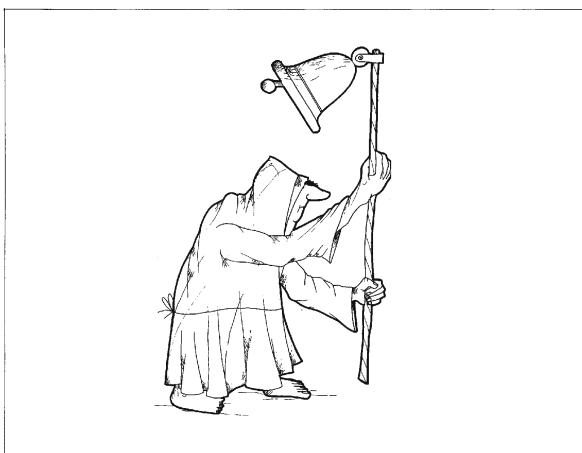




Rumpelstiltskin



Dwarf Long-Nose



Quasimodo

Even Rumpelstiltskin helps the miller's daughter to spin straw into gold until she becomes queen, although he subsequently claims the queen's child for himself. Perhaps this is due to the fact that dwarfs often remain childless. Dwarf Long-Nose, in the German fairytale, may have looked ugly (after all he owed his appearance to an evil witch), but he was the best cook that the king could ever wish for. The scoliotic dwarf Quasimodo has been portrayed in several films as the »hunchback of Notre-Dame«, based on the novel by Victor Hugo and also appears in a positive light despite his ugliness. The »Little People« (who appear all too rarely alas) are particularly considerate, helpful and kind creatures, while gnomes decorate the gardens of many homes as bringers of good luck.

Even though society may ascribe certain positive qualities to dwarfs there is no doubt that small stature is associated with considerable *psychological problems* for those affected. If we were all small then small people would not have any particular problems. Most people of small stature do not feel themselves to be handicapped. In a study of young people aged between 14 and 20 with heights between 85 cm (2 ft 9 in) and 150 cm (4 ft 11), 85% did not consider themselves to be disabled [1]. Very small individuals experience restrictions in their everyday lives because they are unable to reach light switches, elevator buttons, washbasins or shop counters, but these functional handicaps can at least be minimized by practical appliances and a certain amount of creativity.

Of much greater consequence and far more difficult to cope with is the »social handicap«: the stigmatization based solely on a difference in height. The fact that other people turn round and stare, almost as a reflex action, simply because one is particularly short. The fact that small individuals automatically become the center of attention whenever they venture into the outside world: on the street, in restaurants, on public transport, everywhere. Wherever they live, they soon become known by all. The reactions they encounter range from astonishment, insecure or very unusual behavior, sympathy, mockery extending to maliciousness, depending on the maturity of the onlooker.

Height therefore appears to play an extremely important role in our society. One reason is the link between height and social status. A study by an anthropologist investigating the connection between shortness and tallness with properties that are attributed to them showed that a tall person was considered to be healthier, stronger, more interesting, more serious, more active, safer, tougher and more open than a short person [7]. This range of properties is also commonly ascribed to successful individuals. In this context, the wish of many small people to be made taller with the help of advances in modern medicine is perfectly understandable. Many are prepared to invest a great deal of time and effort and tolerate considerable pain in order to achieve their goal.

However, in view of the many complications associated with even the latest procedures (► Chapter 4.2.2) it is certainly appropriate to establish from the outset whether psychological support might be a better approach for resolving the problem than a distressing bilateral leg lengthening procedure.

For *parents* the realization that the child's growth will be stunted usually comes as a shock. The way in which this »crisis« is managed is hugely important for the child's future. The greater the parents' self reproach, the greater the risk that the child will be spoilt and incapable, in later life, of living independently despite having the necessary intellectual abilities. It is important therefore for the treating doctors to avoid stirring up any feelings of guilt on the part of the parents. The family and pregnancy history must be taken with extreme sensitivity. Specific details should only be questioned if they are actually relevant to the diagnostic process. Seemingly trivial routine questions (such as the administration of drugs or the drinking of alcohol during pregnancy) can very easily lead to lifelong (unjustified) self reproach on the part of the mother.

» *Childhood illnesses of the soul first manifest themselves in adults* «  
(Hans Weigel)

Many patients with hereditary illnesses are perfectly normal in mental respects. Some may show outstanding *artistic talent*. Thus the painter Henri de Toulouse-Lautrec, who probably suffered from pyknodysostosis, characterized a whole era with his pictures at the end of the 19th century. The recently deceased Michel Petrucciani, whose small stature was associated with osteogenesis imperfecta, was one of the greatest jazz pianists of this time. Even the *powerful figures* of the world were not always tall and strong. Attila, the king of the Huns, King Charles III of Naples and Sicily and Napoleon were all said to be small in stature.



The association »Little People of America« was founded in 1957 in America in order to achieve better representation of their common interests and help short people cope with their problems. Associations of little people exist in many countries of the world. Their addresses can readily be found on the internet.

» *small is beautiful...* «

### Classification

Classifying such a heterogeneous group as the congenital disorders of the musculoskeletal system is not a simple undertaking. But such a classification is needed in order to create a common basis for professional discussions. The »Committee on Nomenclature on Intrinsic Diseases of Bones« of the European Society of Paediatric Radiology was the first body to undertake this classification in 1971 (»Paris Nomenclature«), which has since been revised several times, most recently in 2001 [5]. This primarily clinically oriented classification was recently restructured to take account of the findings of molecular genetics [8]. This international classification currently comprises 36 groups, including 33 with generalized disorders (osteochondrodysplasias) and 3 with localized deficiencies (dysostoses). While the gene defect in most disorders is already known [2, 5, 8], the gene product still remains unclear in many cases. In view of the rapid development of genetics, these gaps are also expected to be closed in the near future.

### Classification skeletal dysplasias according to the »Committee on Nomenclature on Intrinsic Diseases of Bones«

#### Osteochondrodysplasias

##### 1. Achondroplasia group

- Thanatophoric dysplasia
- Achondroplasia
- Hypochondroplasia

##### 2. Severe spondylodysplastic dysplasias

##### 3. Metatropic dysplasias

- Metatropic dwarfism

##### 4. Short-rib dysplasias

- Asphyxiating thoracic dysplasia (Jeune syndrome)
- Chondroectodermal dysplasia (Ellis-van-Creveld syndrome)

##### 5. Atelosteogenesis-omodysplasia group

##### 6. Group of diastrophic dysplasias

- Diastrophic dysplasia

##### 7. Group of dyssegmental dysplasias

##### 8. Type II collagenopathies

- Achondrogenesis II and hypochondrogenesis
- Spondyloepiphyseal dysplasia
- Kniest syndrome
- Stickler syndrome

**9. Type XI collagenopathies**

- Otospondylomegalepiphyseal dysplasias

**10. Other spondyloepi-(meta)-physeal dysplasias****11. Multiple epiphyseal dysplasia**

- Multiple epiphyseal dysplasia
- Pseudoachondroplasia

**12. Chondrodysplasia calcificans punctata**

- Chondrodysplasia calcificans punctata, Rhizomelic type
- Zellweger syndrome
- Chondrodysplasia calcificans punctata, Conradi-Hünemann type

**13. Metaphyseal dysplasia****14. Spondylometaphyseal dysplasia****15. Brachyolmia spondylodysplasias****16. Mesomelic dysplasias****17. Acromelic dysplasias**

- Trichorhinophalangeal syndrome
- Brachydactyly

**18. Acromesomelic dysplasias****19. Dysplasias with predominant involvement of flat bones**

- Cleidocranial dysplasia

**20. Dysplasias with bent bones**

- Camptomelic (or camptomelic) dysplasia
- Stüve-Wiedemann dysplasia

**21. Dysplasias with multiple dislocations**

- Larsen syndrome

**22. Dysostosis multiplex group**

- Mucopolysaccharidoses

**24. Dysplasias with decreased bone density**

- Juvenile osteoporosis
- Osteogenesis imperfecta

**25. Dysplasias with defective mineralization**

- Osteomalacia
- Renal osteodystrophy
- Hyper-/Hypoparathyroidism (primary, idiopathic, Pseudohypoparathyroidism)

**26. Increased bone density without modification of shape**

- Melorheostosis
- Osteopoikilosis
- Osteopetrosis

**28. Increased bone density with metaphyseal involvement**

- Metaphyseal dysplasia (Pyle's disease)

**29. Craniotubular digital dysplasia****30. Neonatal severe osteosclerotic dysplasia****31. Disorganized development of cartilaginous and fibrous components of the skeleton**

- Dysplasia epiphysealis hemimelica («Trevor's disease»)
- Multiple osteochondromas
- Metachondromatosis
- Multiple enchondromatosis (Ollier syndrome)

- Maffucci syndrome

- Fibrous dysplasia, (monostotic, polyostotic, McCune-Albright syndrome)

- Fibrodysplasia ossificans progressiva

- Cherubism

**32. Osteolyses**

- Familial multicentric carpal/tarsal osteolysis

- Torg syndrome

- Familial expansile osteolysis

- Gorham syndrome

**33. Patella dysplasias**

- Nail-patella syndrome (onycho-osteodysplasia)

- Meier-Gorlin syndrome

**Dysostoses (localized hereditary skeletal deformities)****A. Dysostoses with predominant cranial and facial involvement**

- Apert syndrome (Acrocephalosyndactyly)

**B. Dysostoses with predominant axial involvement**

- Spondylocostal dysplasia (Jarcho-Levin syndrome)

**C. Dysostoses with predominant involvement of the extremities**

- Fanconi syndrome

- Coffin-Siris syndrome

- Rubinstein-Taybi syndrome

Certain hereditary diseases with orthopaedic implications that are not included in this classification also exist. We have grouped them in subsequent chapters according to the following aspects:

**Chromosomal abnormalities**

- Down syndrome

- Trisomy 8

- Trisomy 5

- Trisomy 18

- Turner syndrome

- Klinefelter syndrome

- Fragile X syndrome

**Syndromes with neuromuscular abnormalities**

- Arthrogryposis

- Pterygium syndromes

- Popliteal pterygium syndrome

- Progressive form of multiple pterygium syndrome

- Recessive form of multiple pterygium syndrome

- Pterygium syndromes

- Goldenhar syndrome (oculoauriculovertebral syndrome, hemifacial microsomia)

- Moebius syndrome

- Cornelia-de-Lange syndrome (Brachmann-de-Lange syndrome)

- Pierre-Robin syndrome

- Williams-Beuren syndrome

- Prader-Willi-Labhard syndrome
- Rett syndrome
- Dandy-Walker syndrome

#### Various syndromes with orthopaedic relevance

- Gaucher disease
- Neurofibromatosis (type I and II)
- Proteus syndrome
- Klippel-Trenaunay-Weber syndrome
- Marfan syndrome
- Homocystinuria
- Ehlers-Danlos syndrome
- Silver-Russell syndrome
- Holt-Oram syndrome
- Poland syndrome
- Hemophilia
  - Hemophilia A
  - Hemophilia B
  - von Willebrand disease
- Hypothyroidism
- Sotos syndrome
- Weaver syndrome
- Beckwith-Wiedemann syndrome

#### Epidemiology of the diseases

Reliable epidemiological data are not always available for every hereditary condition. Figures on prevalence are available, however, for the most important skeletal dysplasias thanks to an excellent study in Great Britain [9]. This survey extended over a period of 30 years, and most epidemiological figures in the text are based on this work.

#### Definition

As regards *morbidity*, a distinction must be made between two important *terms*:

##### Incidence:

The number of new patients contracting illness I in the year Y in region R in a specified population.

##### Prevalence:

Patients with illness I on day D in region R.

The figures in the text on the individual clinical conditions are not cited uniformly and may refer to an incidence or a prevalence depending on the source. While figures on incidence and prevalence cannot be compared directly with each other, we have converted the incidence to a population size of 100,000 and the prevalence to a population of 1 million (this applies to all epidemiological figures in this book).

Fig. 4.67 shows the relative frequency (prevalence) of the principal skeletal dysplasias in the form of a bar chart. An incidence of 25/100,000 neonates has been calculated in the USA for all skeletal dysplasias. In this country the commonest forms are achondrogenesis, achondroplasia, osteogenesis imperfecta and thanatophoric dysplasia.

#### Differential diagnosis

During the examination of a child with an unknown syndrome a single symptom can point the way. Clues in the overall picture can facilitate the diagnostic process.

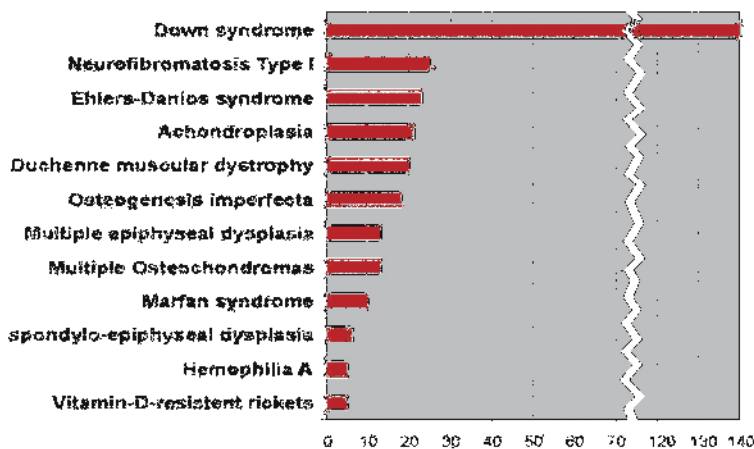


Fig. 4.67. Incidence of the most common hereditary disorders with orthopaedic relevance [6, 9]. The incidence of disorders not listed in the chart is less than 5/100,000 births

### Differential diagnoses

#### ■ Prognosis (life expectancy)

- Lethal  
Achondrogenesis, Conradi chondrodysplasia calcificans punctata, hypochondrogenesis, myopathies (lethal forms), osteogenesis imperfecta (Vrolik type), thanatophoric dwarfism
- Severely restricted (<10 years)  
Cornelia-de-Lange syndrome (poss.), cri-du-chat syndrome, Dandy-Walker syndrome (poss.), Down syndrome, Holt-Oram syndrome, metatropic dwarfism, mucopolysaccharidosis Pfaundler-Hurler, osteopetrosis (infantile malignant form)
- Moderately restricted  
Arthrogryposis (poss.), homocystinuria, Larsen syndrome (poss.), Marfan syndrome, Möbius syndrome, myopathies (Duchenne), osteogenesis imperfecta (Lobstein type) (poss.), mucopolysaccharidoses (various types), multiple epiphyseal dysplasia, Pierre-Robin syndrome, Prader-Willi syndrome, Rett syndrome, spondyloepiphyseal dysplasia (congenital type), spinal muscular atrophy, trisomy 18,

#### ■ Expected final height

- Severe dwarfism (final height <100 cm or 3 ft 3 in)  
Diastrophic dysplasia (poss.), osteogenesis imperfecta (poss.), pseudoachondroplasia (poss.)
- Moderate dwarfism (final height 100–130 cm, or 3 ft 3 in – 4 ft 3 in)  
Achondroplasia, Conradi-Hünemann chondrodysplasia calcificans punctata, Cornelia-de-Lange syndrome, diastrophic dysplasia, Kniest syndrome, Morquio mucopolysaccharidosis, metatropic dwarfism, osteogenesis imperfecta, pseudoachondroplasia, Silver-Russell syndrome, spondyloepiphyseal dysplasia (congenital type)
- Slight dwarfism (final height 130–150 cm or 4 ft 3 in – 4 ft 11 in)  
Ehlers-Danlos syndrome (type VII), Hunter syndrome, hypochondroplasia, Meier-Gorlin syndrome, Pfaundler-Hurler mucopolysaccharidosis, pterygium syndrome, Williams-Beuren syndrome
- Small stature (final height over 150 cm or 4 ft 11 in)  
Arthrogryposis, brachyolmia, Larsen syndrome, multiple epiphyseal dysplasia (poss.), multiple cartilaginous exostoses (poss.), trisomy 8,

- Tall stature

Beckwith-Wiedemann syndrome, Homocystinuria, Klinefelter syndrome, Marfan syndrome, Sotos syndrome, Weaver syndrome

#### ■ Mental retardation

Apert syndrome (poss.), arthrogryposis (poss.), Cornelia-de-Lange syndrome, Cri-du-chat syndrome, Dandy-Walker syndrome, Down syndrome, Gaucher syndrome, Goldenhar syndrome (poss.), hypochondroplasia (poss.), mucopolysaccharidosis, Möbius syndrome (poss.), muscular dystrophy, myelomeningocele (poss.), neurofibromatosis (poss.), osteopetrosis (poss.), Pfaundler-Hurler syndrome, Pierre-Robin syndrome, Rett syndrome, Rubinstein-Taybi syndrome, Sotos syndrome, Williams-Beuren syndrome, cerebral palsy

#### ■ Gait abnormalities

Arthrogryposis, Cri-du-chat syndrome, Dandy-Walker syndrome, Gaucher syndrome, homocystinuria (poss.), mucopolysaccharidosis (poss.), Möbius syndrome (poss.), muscular dystrophy, myelomeningocele, Pierre-Robin syndrome, Rett syndrome, Williams-Beuren syndrome, cerebral palsy

#### ■ Altered body proportions

Achondroplasia (short extremities), brachyolmia (short trunk), chondrodysplasia calcificans punctata, rhizomelic type (shortened upper arms and thighs), diastrophic dysplasia (short extremities), Leri-Weill dyschondrosteosis (shortened forearms and lower legs), Ellis-van-Creveld syndrome (short extremities), hypochondroplasia (short extremities), spondyloepiphyseal dysplasia (only moderate since the short extremities are associated with a short trunk)

#### ■ Hemihypertrophy

Klippel-Trenaunay syndrome, neurofibromatosis, Proteus syndrome, Silver-Russell syndrome

#### ■ Obesity

Prader-Willi syndrome, hypothyroidism

#### ■ Facial abnormalities

Achondroplasia (saddle nose, bulging forehead, prognathism), Apert syndrome (wide head), Conradi-Hünemann chondrodysplasia calcificans punctata (flat face), Cornelia-de-Lange syndrome (upper lip, eyebrows), cherubism (mandibular deformity), Down syndrome, Goldenhar syndrome (facial asymmetry), Larsen syndrome (flat face, bulging forehead, hypertelorism), Kniest syndrome (hypertelorism, flat nose), mucopolysaccharidoses (gargoylism), Marfan syndrome (dolichocephaly), Möbius syndrome (facial palsy, mandibular hypo-



plasia), osteogenesis imperfecta (triangular face), pterygium syndrome, Silver-Russell syndrome (triangular face, downturned corners of the mouth), Sotos syndrome, (hypertelorism, epicanthus), Williams-Beuren syndrome (dental abnormalities)

#### ■ Cleft lip and palate

Atelogenesis, Goldenhar syndrome, Pierre-Robin syndrome, Pterygium syndrome, spondyloepiphyseal dysplasia

#### ■ Eye changes

Apert syndrome (Impairment of ophthalmic nerves), Conradi-Hünermann chondrodysplasia calcificans punctata (congenital cataract), Down syndrome, Ehlers-Danlos syndrome (ectopia lentis in type VI), Gaucher syndrome (strabismus), homocystinuria (ectopia lentis), Kniest syndrome (myopia), Marfan syndrome (ectopia lentis), Möbius syndrome (abducent nerve palsy), osteogenesis imperfecta (blue sclerae, poss. retinal detachment), spondyloepiphyseal dysplasia (poss. cataract, myopia, retinal detachment), Stickler syndrome (myopia, glaucoma)

#### ■ Ears (hearing loss, deafness, changes in the pinna)

Diastrophic dysplasia (pinna), Goldenhar syndrome (pinna), Kniest syndrome (deafness), Meier-Gorlin syndrome (missing ears), neurofibromatosis type II (vestibular schwannoma), osteogenesis imperfecta (deafness due to otosclerosis)

#### ■ Dental problems

Chondroectodermal dysplasia, Ehlers-Danlos syndrome (periodontosis in type VIII), Ellis-van-Creveld syndrome, osteogenesis imperfecta (impaired dentinogenesis)

#### ■ Changes in the skin and appendages

Albright syndrome (pigmentations), Conradi-Hünermann chondrodysplasia calcificans punctata (dry skin), chondroectodermal dysplasia, Ehlers-Danlos syndrome (hyperelasticity, pigmentations, hematomas), Ellis-van-Creveld syndrome (nails), fibrodysplasia ossificans progressiva (calcifications), Gaucher disease (pigmentations), Klippel-Trenaunay-Weber syndrome (vascular nevi), Maffucci syndrome (hemangiomas), nail-patella syndrome (nail dystrophy), neurofibromatosis type I (café-au-lait spots, fibroma molluscum), Proteus syndrome (tumorous changes), pterygium syndrome (popliteal fossae; nail changes)

#### ■ Chest deformity

Down syndrome, Ellis-van Creveld syndrome (too short ribs), homocystinuria (poss. funnel

chest) Jeune syndrome (too short ribs), Marfan syndrome (poss. funnel chest), Poland syndrome (aplasia of the pectoral muscle, nipple hypoplasia), pterygium syndrome (poss. keeled chest), spondyloepiphyseal dysplasia, rickets (rachitic rosary) Sotos syndrome,

#### ■ Heart defects

Cornelia-de-Lange syndrome, Ellis-van-Creveld syndrome, Holt-Oram syndrome, Marfan syndrome, Pierre-Robin syndrome, Turner syndrome

#### ■ Abnormalities of tendons and muscles

Fibrodysplasia ossificans progressiva (calcifications), myopathies (muscular dystrophies), pterygium syndrome

#### ■ Ligament laxity

Down syndrome, Ehlers-Danlos syndrome, fragile X syndrome, homocystinuria, Larsen syndrome, Marfan syndrome, osteogenesis imperfecta, pseudoachondroplasia,

#### ■ Habitual dislocations

Atelogenesis, Down syndrome, Ehlers-Danlos syndrome, Ellis-van-Creveld syndrome (patella), homocystinuria, Marfan syndrome, Larsen syndrome, nail-patella syndrome (patella, elbow), Rubinstein-Taybi syndrome (poss. patellar dislocation),

#### ■ Joint contractures

Arthrogyrosis, Conradi-Hünermann chondrodysplasia calcificans punctata, diastrophic dysplasia, fibrodysplasia ossificans progressiva, hemophilia (poss.), homocystinuria, Marfan syndrome (with contractural form), metatropic dwarfism, pterygium syndrome, spondyloepiphyseal dysplasia (congenital type), trisomy 8

#### ■ Reduced bone strength

Chondrodysplasia punctata, Ehlers-Danlos syndrome, Gaucher syndrome (pathological fractures), juvenile osteoporosis, Klinefelter syndrome, mucopolysaccharidoses, osteogenesis imperfecta

#### ■ Altered radiographic bone density

Familial expansile osteolysis, Gorham syndrome («vanishing bone»), infantile cortical hyperostosis (Caffey), juvenile osteoporosis, Klinefelter syndrome (osteopenia), melorheostosis (localized sclerosis), osteopoikilosis, osteopetrosis (increased bone density), primary hyperparathyroidism (pseudotumors)

#### ■ Delayed skeletal maturation

Cleidocranial dysplasia, hypothyroidism, camptomelic dysplasia, spondylometaphyseal dysplasia, cerebral palsy

- 4
- Epiphyseal radiographic changes  
Chondrodysplasia calcificans punctata (calcifications), dysplasia epiphysealis hemimelica, mucopolysaccharidoses (necroses), metatropic dwarfism, multiple epiphyseal dysplasia, pseudoachondroplasia, spondyloepiphyseal dysplasia, Stickler syndrome
  - Metaphyseal radiographic abnormalities  
Achondroplasia, Albright syndrome (tumors), fibrous dysplasia (tumors), enchondromatosis (Ollier disease), Gaucher syndrome (widening), hypochondroplasia, Kniest syndrome, metachondromatosis, metaphyseal dysplasia (Pyle's disease), metatropic dwarfism, multiple cartilaginous exostoses (osteochondromas), pseudoachondroplasia
  - Axial deviations  
Achondroplasia (genua vara, excessively long fibula), Leri-Weill dyschondrosteosis (bowing of the radius and tibia), Ellis-van-Creveld syndrome (genua valga), enchondromatosis (Ollier disease), hypochondroplasia, camptomelic dysplasia, Kniest syndrome, Larsen syndrome (poss.), metaphyseal dysplasia (Pyle's disease), metatropic dwarfism, multiple cartilaginous exostoses (osteochondromas) (poss.), neurofibromatosis type I (poss. tibial bowing and pseudarthrosis), osteogenesis imperfecta, pseudoachondroplasia (genua valga), rickets, renal osteodystrophy, spondyloepiphyseal dysplasia (coxa vara), Stüve-Wiedemann dysplasia
  - Abnormalities of the hands (▶ Chapter 3.5.3)  
Apert syndrome (synostoses), cri-du-chat syndrome (clinodactyly, shortening of the metacarpals), Cornelia-de-Lange syndrome (ray defects, small hands), diastrophic dysplasia (abducted thumb), cleidocranial dysostosis (short little fingers), Down syndrome (palmar crease, short hands, Ellis-van-Creveld syndrome (polydactyly), Holt-Oram syndrome (shortening of the thumb), Marfan syndrome (arachnodactyly), metachondromatosis (exostoses), Möbius syndrome (syndactylies), Poland syndrome (sybrachydactyly), Prader-Willi syndrome (small hands), Proteus syndrome (macroductyly) Rubinstein-Taybi syndrome (deviation of thumb), Silver-Russell syndrome (clinodactyly V), trichorhinophalangeal syndrome (brachyphalangia), trisomy 8 (camptodactyly, clinodactyly), trisomy 18 (excessively long index fingers, flexion contracture of little fingers),
  - Deformities of the forearms and elbow  
Atelogenesis, Cornelia-de-Lange syndrome (radial deformation, radial head dislocation), Leri-Weill dyschondrosteosis (Madelung deformity, dislocations), Holt-Oram syndrome (radial aplasia, shortening of the ulna), camptomelic dysplasia (radioulnar synostosis), Klinefelter syndrome (poss. radioulnar synostosis), multiple osteochondromas (cartilaginous exostoses), TAR syndrome (thrombocytopenia-absent radius syndrome), Williams-Beuren syndrome (poss. radioulnar synostosis)
  - Spinal changes (▶ Chapter 3.1.11)  
Achondroplasia (lumbar kyphosis, short pedicles), Apert syndrome (segmentation defects), Conradi-Hünermann chondrodysplasia calcificans punctata (kyphosis, scoliosis), diastrophic dysplasia (pronounced kyphoses), Down syndrome (atlanto-axial instability), Ehlers-Danlos syndrome (poss. scoliosis), fragile X syndrome (poss. scoliosis), Goldenhar syndrome (poss. formation defects), hypochondroplasia (short pedicles), fibrodysplasia ossificans progressiva (strikingly small vertebral bodies in the cervical spine), Klippel-Trenaunay-Weber syndrome (scolioses, kyphoses, tumors), Kniest syndrome (platyspondylia), Larsen syndrome (kyphoses, segmentation defects), Marfan syndrome (scoliosis, spondylolisthesis, widening of the lumbar interpedicular distance), metatropic dwarfism (atlantoaxial instability, scoliosis, kyphosis), mucopolysaccharidoses (platyspondylia), neurofibromatosis type I (scolioses), osteogenesis imperfecta (scoliosis, kyphosis), osteopetrosis (spinal stenosis), Pierre-Robin syndrome (atlantoaxial instability), pseudoachondroplasia (flattened vertebral bodies, atlantoaxial instability), Rett syndrome (scoliosis), Rubinstein-Taybi syndrome (cervical spondylolisthesis), Sotos syndrome, spondyloepiphyseal dysplasia (atlanto-axial instability, platyspondylia, kyphosis, scoliosis), trisomy 18 (scoliosis, congenital anomalies), Williams-Beuren syndrome (disk calcifications, poss. scoliosis)
  - Changes in the hips (▶ Chapter 3.2.7)  
Arthrogyrosis (poss. hip dislocation) epiphyseal dysplasia (deformation), cleidocranial dysostosis (coxa vara), Down syndrome (poss. secondary dislocation), hemophilia (poss. femoral head necrosis), mucopolysaccharidosis (femoral head necrosis), metaphyseal dysplasia (deformation), pseudoachondroplasia (fragmented ossification centers), rickets (coxa vara), spondyloepiphyseal dysplasia (coxa vara with late onset),

- Changes in the patella  
Diastrophic dysplasia (poss. agenesis), Down syndrome (poss. dislocation), Ehlers-Danlos syndrome (poss. dislocation), Ellis-van-Crevelde syndrome (poss. dislocation), Marfan syndrome (poss. dislocation), Meier-Gorlin syndrome (agenesis), nail-patella syndrome (hypoplasia, poss. dislocation), osteogenesis imperfecta (poss. dislocation), pterygium syndrome (popliteal fossae; poss. agenesis), Rubinstein-Taybi syndrome (poss. dislocation),
- Abnormalities of the feet (▶ Chapter 3.4.5)  
Apert syndrome (synostoses), arthrogryposis (clubfeet), Cornelia-de-Lange syndrome (small feet), diastrophic dysplasia (clubfeet), Down syndrome, Ehlers-Danlos syndrome (clubfeet), Ellis-van-Crevelde syndrome (polydactyly), fibrodysplasia ossificans progressiva (shortening of the great toe), fragile X syndrome (flatfeet), homocystinuria (pes cavus), Klippel-Trenaunay-Weber syndrome (poss. macro-/syn-/poly-/camptodactyly) Larsen syndrome (multiple ossification centers), Marfan syndrome (clubfeet) (poss.), Möbius syndrome (clubfeet), Pierre-Robin syndrome, Prader-Willi syndrome (small feet), Proteus syndrome (macroductyly), pseudoachondroplasia (flatfeet), pterygium syndrome (poss. clubfeet), Rubinstein-Taybi syndrome (deviation of great toes), spondyloepiphyseal dysplasia (poss. clubfeet), trisomy 8 (clubfeet), trisomy 18 (clubfeet), (flatfeet)

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## 4.6.2 Skeletal dysplasias

» *I cried because I had no shoes, until I met a man who had no feet.* «  
(Helen Keller)

### ► Definition

The classification of skeletal dysplasias is based on the »International Nosology and Classification of Constitutional Disorders of Bone« 2001 [31]. The number after the chapter number »4.6.2« corresponds to the number of this classification.

### 4.6.2.1 Achondroplasia group

The common feature in this group is the chromosomal locus of the defect: 4p16.3. This is the fibroblast growth factor receptor 3 gene (FGFR3).

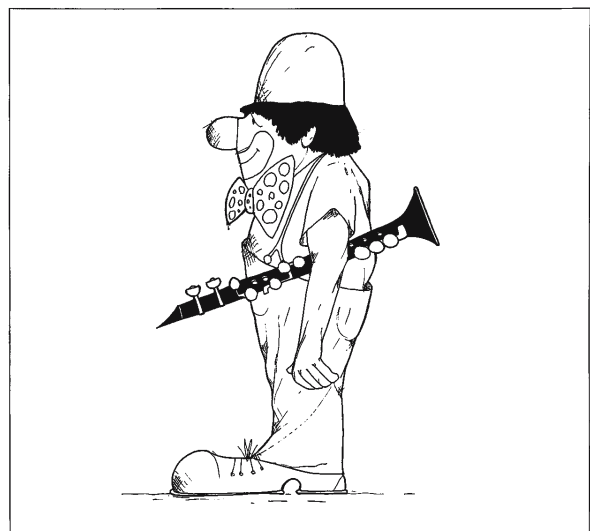
#### Thanatophoric dysplasia

Very severe, disproportionate small stature, even at birth, with macrocephaly (poss. cloverleaf skull) and very narrow thorax with severely shortened ribs, which cause respiratory insufficiency, from which the children usually die immediately after birth. Relatively common in the USA, but the orthopaedist is rarely confronted with this condition in view of the early lethality.

#### Achondroplasia

### ► Definition

Autosomal-dominant inherited disorder with dwarfism and impaired enchondral ossification, but normal periosteal bone formation. The disease is characterized by short extremities with a normal trunk length and head size, protruding forehead and saddle nose, a constricted lumbar spinal canal and typical pelvic changes.



### Historical background

The term *achondroplasia* was coined in 1878 by Parrot [87], who also described the Egyptian god Ptah as suffering from achondroplastic dwarfism. The term *chondrodystrophy* was introduced by Kaufmann in 1892 [46] and has, for a long time, been more commonly used in preference to achondroplasia in German-speaking countries.

### Etiology, pathogenesis, occurrence

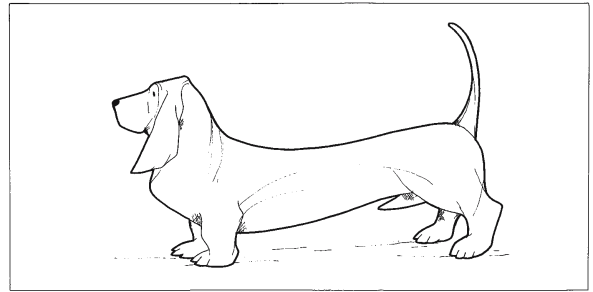
Achondroplasia is inherited as an autosomal-dominant condition, although most cases occur as spontaneous mutations in normal parents. The disorder affects enchondral ossification as a result of a defect in the fibroblast growth factor receptor 3 gene [117]. Cartilage production is reduced and its normally present palisade layer is lacking, being replaced by mucoid degeneration. Since periosteal ossification remains unaffected, the diaphyses show a normal diameter. Achondroplasia is one of the more common hereditary diseases, with a calculated prevalence of 4.3/million [115].

Achondroplasia is also known in the animal kingdom. Individual dog breeds (particularly the dachshund) are, in »physiological« terms, achondroplastic dwarfs with short legs and a normal trunk length.

### Clinical features, diagnosis

The diagnosis can usually be made even prenatally or at birth.

- Compared to the head and trunk, the extremities are shortened. This feature is particularly evident in the proximal segments (femur and humerus). The head is disproportionate with a bulging forehead. The face is broad, the nose is flattened and saddle-shaped, and the lower jaw protrudes slightly (prognathism), producing a very characteristic overall facial picture. Dental development is normal. The lumbar lordosis is accentuated and the abdomen bulges outward, and the habitus is very typical. In the lower extremities, genua vara is often present since the fibula is too long relative to the tibia [64, 110]. The patients look muscular. The skin is creased and there appears to be too much skin. The patient has a waddling gait and a normal intelligence.
- *Radiological examination* reveals changes in all long bones (■ Fig. 4.68). The metaphyses are widened but the epiphyses are of normal width. The clavicle and fibula are not as severely affected as the other long bones, whereas the humerus and femur are the bones that are most often altered and shortened. The sacrum is narrow and strongly tilted, while the acetabula are horizontal and wide. The ilium is likewise broader than normal, with a reduced depth. This produces a change in the contours of the inner pelvic ring, which becomes oval and wider than normal (■ Fig. 3.214, ► Chapter 3.2.7). The spine is normal in length, but on the lateral x-ray the ossification centers of the vertebral bodies are smaller than normal. A frequent finding is



The dachshund is a »physiological« achondroplastic dwarf



■ Fig. 4.68. AP x-ray of both legs of a 6-year old boy with *achondroplasia*. Typical findings are the swollen bulb-like metaphyses in the long bones and the excessively long fibulas

a long drawn-out kyphosis extending to the lumbar spine above an acutely-angled lordosis. The interpedicular distance decreases from L1 toward L5 (normally the opposite applies). The pedicles are also shortened, which can lead to premature spinal stenosis.

- The most important *differential diagnoses* to consider are *hypochondroplasia* and *pseudoachondroplasia*. The distinguishing features are listed in ■ Table 4.20. Confusion, at birth, with other (lethal) forms of dwarfism (such as thanatophoric dwarfism or achondrogenesis) is possible.

### Treatment, prognosis

No known *treatment of the underlying illness* currently exists. Trials with the administration of growth hormone have not proved convincing [117]. The intrinsic *prognosis* is very good. The individuals remain in good health and

## 4.6.2 · Skeletal dysplasias

have a largely normal life expectancy. Arthroses are not especially common since the epiphyses themselves are not affected by the disease. By contrast, patients suffer prematurely from spinal stenosis. The *psychological effects* of the small stature are not inconsiderable.

The *orthopaedic treatment* focuses on three factors:

- axial deviations,
- height,
- spinal stenosis.

The *axial deviations* mainly affect the knee. The excessively long fibula frequently results in the development of genua vara. Useful *preventive* measures include the wearing of splints to stabilize the lateral ligaments and an appropriately timed closure of the proximal fibular epiphyseal plate. If the latter procedure is omitted, a corrective osteotomy,

generally at the tibial subcondylar level, usually has to be performed at a later date. In this operation the tibia must be lengthened (without lengthening the fibula, which is fixed to the distal part of the tibia during the lengthening procedure) so as to restore the correct length. The bones heal completely normally after the osteotomies. We stabilize the situation in the growing patient with crossed Kirschner wires and fit a plaster cast for 4 weeks.

Patients with achondroplasia occasionally express a wish for *bilateral leg lengthening*. Such lengthening procedures are common particularly in Russia and Southern Europe [3, 56]. However understandable this wish may be, the surgeon must be very careful in deciding whether the procedure is indicated. In practice, the lengthening of extremities is advantageous only if it results in a normal height, i.e. at least 150 cm (4 ft 11 in). This implies a total

■ Table 4.20. Differential diagnosis of achondroplasia, hypochondroplasia and pseudoachondroplasia

	Achondroplasia	Hypochondroplasia	Pseudoachondroplasia
<b>Inheritance</b>	Autosomal-dominant, more rarely poss. autosomal-recessive	Autosomal-dominant	Autosomal-dominant, more rarely poss. autosomal-recessive
<b>Pathology</b>	Metaphyseal	Metaphyseal	Meta- and epiphyseal
<b>Frequency</b>	4,3 : 1 00,000	2,6 : 100,000	4,3 : 100,000
<b>Final height</b>	120–130 cm (3ft 11in–4ft 3in)	130–140 cm (4ft 3in–4ft 7in)	100–115 cm (3ft 3in–3ft 9in)
<b>Proportions</b>	Disproportionate	Disproportionate	Proportionate
<b>Time of diagnosis</b>	At birth	Aged 2–3 years	Aged 2–3 years
<b>Face</b>	Macrocephaly with »balcony forehead« and »saddle nose«, prognathism, poss. hydrocephalus	No change	No change
<b>Extremities</b>	Shortening, genua vara, extension deficit of elbow, brachydactyly with trident hand	Shortening, Genua vara, extension deficit of elbow, brachydactyly	Shortening, early osteoarthritis, ligament laxity (genu valgum and recurvatum), brachydactyly
<b>Radiographic findings</b>	Swollen bulb-like metaphyses in the long bones, excessively long fibulas, brachydactyly	Swollen bulb-like metaphyses in the long bones, excessively long fibulas, brachydactyly	Swollen bulb-like metaphyses, delayed development of the epiphyses with small, fragmented ossification center, excessively long fibulas
<b>Spine, trunk</b>	Hyperlordosis, protruding abdomen, thoracolumbar infantile gibbus, lumbar kyphosis	Hyperlordosis	Hyperlordosis
<b>Radiographic findings</b>	Wedge-shaped deformation of the vertebral bodies, deformation of the posterior wall, short pedicles, reduced interpedicular distance at lumbar level, spinal stenosis	Slightly shortened pedicles, no or slight spinal stenosis	Flattened oval vertebral bodies, in some cases »triangular«, normal pedicles, no spinal stenosis
<b>Pelvis, hip; Radiographic findings</b>	Horizontal acetabular roof and square iliac wing	Horizontal acetabular roof	Wide and late-ossifying triradiate cartilage, acetabular dysplasia, coxa vara
<b>Gait</b>	Waddling, toddling gait	Waddling gait	Waddling gait
<b>Habitus</b>	Hypotonia	Muscular	
<b>Intelligence</b>	Normal	Normal (diminished in 10%)	Normal



lengthening of approx. 30 cm (12 in) (in the upper and lower legs). Since the arms will then look out of proportion, the upper arms will need to be lengthened as well.

Such a complicated lengthening process is associated with huge problems: On the one hand, it will involve a total treatment period of four years in several stages. On the other hand, the possible complications increase substantially with lengthening of more than 8 cm. Lengthening therefore has to be effected in several stages. Before the doctor embarks on such a treatment, the lengthening candidate should meet other patients who have already been through the whole process. Only if the patient has a realistic understanding of, and is prepared to accept, the risk of complications may this elaborate treatment be initiated (► Chapter 4.2.2).

*Spinal stenosis* can lead to problems as early as young adulthood. It is treated by laminectomy or widening of the vertebral foramen. The orthopaedist should not wait too long before providing this treatment, particularly if neurological signs and symptoms occur.

### Hypochondroplasia

#### ► Definition

Milder form of achondroplasia with less serious changes, particularly affecting the skull and spine. The condition is not usually diagnosed at birth, but only around the age of 2–3 years.

#### Historical background, etiology, pathogenesis, occurrence

Hypochondroplasia was first described in 1913 by Ravena [95]. It is an autosomal-dominant disorder. Here, too, the condition affects enchondral bone formation as a result of a defect of the fibroblast growth factor receptor 3 gene on chromosome 4p16.3 [117]. However, most cases usually occur sporadically as new mutants. While a prevalence of 2.6/million is reported [115], this figure is probably on the low side since the condition cannot be diagnosed at birth and some mild cases occurring at a later date probably remain unrecognized.

#### Clinical features, diagnosis

The stunted growth first becomes apparent around the age of 2–3 years, or in some cases not until the age of 5 or 6. The children otherwise have a normal outward appearance apart from the disproportionately small stature, which is attributable solely to the shortening of the extremities. Flexion contractures of knees and elbows develop over time, and general ligament laxity is observed. A genu varum often forms. At the spinal level, a lumbar hyperlordosis is apparent. The facial features and the hands are not unusual. Intelligence is also usually normal although, for reasons unknown, it is impaired in 10% of cases.

The *x-ray findings* are similar to those for achondroplasia but less pronounced. The long bones are broad and short (■ Fig. 4.69) [65]. The skull, pelvis and hands are normal. At hip level, the acetabular roof is often broader and more horizontal than normal. The greater sciatic foramen is occasionally slightly smaller than normal. At spinal level there is a reduction in the intrapedicular distance, and the pedicles themselves may be slightly shortened, although to a lesser extent than in achondroplasia. The most important *differential diagnosis* is *achondroplasia*, and the differences are listed in ■ Table 4.20.

#### Prognosis, treatment

- Patients with hypochondroplasia attain a height of 130–140 cm (4 ft 3–4 ft 7 in), and occasionally 150 cm (4 ft 11). The life expectancy is normal. No serious handicap is expected and spinal stenoses are rare. The main problem is posed by the psychological effects of the small stature.
- No specific *treatment of the underlying disease* is known.

*Orthopaedic treatment:* Corrective osteotomies are sometimes required for the genua vara. See the section on achondroplasia for details of the problems associated with bilateral lengthening of the extremities and the treatment of spinal stenosis.



■ Fig. 4.69. AP x-ray of both legs of a 6-year old girl with *hypochondroplasia*. The changes are much less pronounced than in achondroplasia, but the bones are short and wide

## 4.6.2 · Skeletal dysplasias

### 4.6.2.2 Severe spondylodysplastic dysplasias

This group includes lethal forms such as platyspondylic skeletal dysplasia and type 1 A achondrogenesis. The chromosomal defect is not yet known.

### 4.6.2.3 Metatropic dysplasias

#### **Metatropic dwarfism**

Metatropic dwarfism was first described in 1966 by Maroteaux [67]. This is a very rare disease, with just isolated reports in the literature. The location of the gene defect in this autosomal-dominant disorder is not known. There are various forms of metatropic dwarfism. Histological investigations have shown an absence of normal cancellous bone formation in the metaphysis in metatropic dwarfism, resulting in impaired enchondral bone formation. Enchondral and perichondral growth become uncoupled [8].

- **Clinical features, diagnosis:** Some forms are lethal even at birth. In others, the *trunk* is normal in length while the extremities are shortened (■ Fig. 4.70). However, scoliosis and kyphosis, develop at an early stage, thereby shortening the trunk as well, while the face appears normal. *Atlantoaxial instability* is occasionally present and can cause a tetraplegia. In the *long bones* both the metaphyses and epiphyses are altered. Flexion contractures of the *knees* and *hips* can also occur.
- The *prognosis* for metatropic dwarfism is poor. Respiratory problems can prove fatal even during childhood. Very premature osteoarthritis can occur as a result of the changes in the epiphyses.
- No *treatment* exists for the underlying disease. The orthopaedic treatment addresses the *deformities* in the joints and the *kyphoscoliosis*.

### 4.6.2.4 Short-rib dysplasias

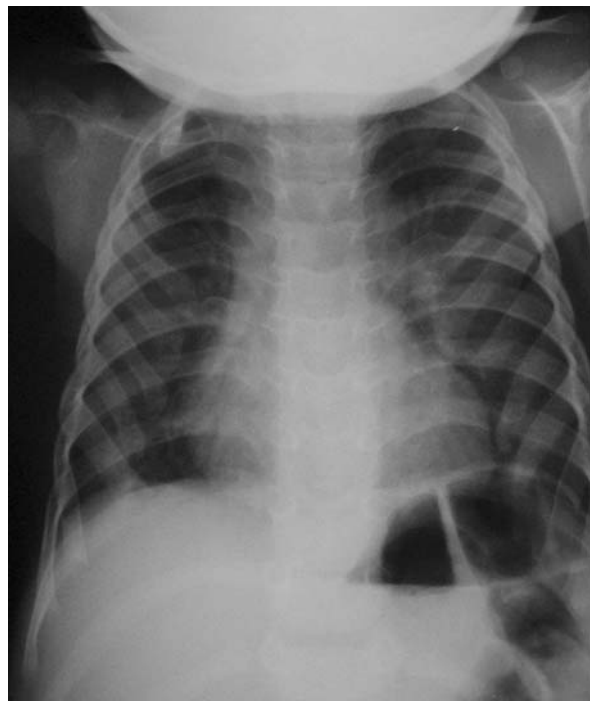
#### **Asphyxiating thoracic dysplasia (Jeune syndrome)**

This syndrome was described by Jeune et al. in 1954 [39]. The location of the gene defect in this autosomal-recessive disorder is not yet known. Two subtypes are distinguished: Type I is characterized by the spotty distribution of enchondral ossification in the epiphyseal plates, an irregular metaphyseal junction and islands of poorly mineralized cartilage in the metaphyses. In type II the disorganized enchondral ossification is regular.

- **Clinically,** the most striking features are the short extremities and excessively short ribs. The thorax is too narrow (■ Fig. 4.71), thus substantially impairing the development of the lungs.
- In terms of *treatment* the pulmonary problems deserve special attention. Attempts have been made in recent years to lengthen the ribs with external fixators [77], although a more promising approach involves the use of titanium ribs (VEPTR; ► Chapter 3.1.7).



■ Fig. 4.70. AP x-ray of both legs of an 11-year old boy with *metatropic dwarfism*, with swollen bulb-like metaphyses in the long bones, epiphyseal changes and fibular shortening



■ Fig. 4.71. Chest x-ray of a 2-year old boy with *Jeune syndrome*. The thorax is very narrow and the take-off point of the ribs is unusually horizontal

### **Chondroectodermal dysplasia (Ellis-van-Creveld syndrome)**

This condition was described in 1940 by Ellis and van Creveld [20]. The location of the gene defect in this autosomal-recessive disorder is not yet known. The disease is extremely rare with an incidence of <0.1/100,000. The parents are usually related to each other.

- The condition can usually be *diagnosed* at birth, or even prenatally with ultrasound [86], although the signs and symptoms become more marked as growth proceeds. The *long bones* are shortened, and the forearm and lower legs are more severely affected than the upper arm and thigh. Since the *trunk* is normal in length, the dwarfism is disproportionate. Dislocation of the *radial head* is often present. The proximal end of the *tibia* is widened, while the ossification center of the proximal tibial epiphysis is hypoplastic and shifted medially, producing a very pronounced genu valgum, particularly during adolescence. The *fibula* tends to be too short rather than too long (in contrast with the situation in achondroplasia). The *patella* frequently subluxates in a lateral direction.

The *finger phalanges* are usually substantially shortened (■ Fig. 4.72). At *wrist* level, an additional carpal bone is frequently present. *Polydactyly* is also common, usually in its postaxial form, i.e. on the ulnar side.

The ectodermal changes affect the *nails*, the *teeth* and the *hair*. A *heart defect* is present in around two-thirds of cases. *Intelligence* is usually normal, although mental retardation has been observed in isolated cases.

- *Differential diagnosis*: It is possible that *Jeune syndrome*, *renal-hepatic-pancreatic dysplasia* and *chondroectodermal dysplasia* are different manifestations of the same disorder [11].
- There is no known *treatment* for the underlying disorder. The most important problem is the *heart defect*, which often requires surgical correction. The valgus deformities of the knees occasionally require a *varization osteotomy* [109]. *Lengthening* of the disproportionately short *lower legs* in order to improve the proportions may be appropriate. *Recentering of the patella* is indicated if it has dislocated laterally, and sometimes requires the detachment and medial transfer of the whole quadriceps muscle (► Chapter 3.3.5).

#### **4.6.2.5 Atelosteogenesis-omodysplasia group**

*Atelosteogenesis* (3 types) is an extremely rare autosomal-recessive syndrome with short extremities and characteristic, wide face with widely spaced eyes and a hypoplastic nose. Multiple joint dislocations, cleft lip and palate, clubfeet, broad, shortened long bones are also features of the disease.

*Omodysplasia* (autosomal-dominant inheritance) is characterized by short upper arms, facial abnormalities

(indented, broad nose, long philtrum), elbow changes (hypoplastic, everted condyles, radioulnar diastasis and anterior dislocation of the radial head). Only a few isolated cases of this condition have been described [68].

#### **4.6.2.6 Group of diastrophic dysplasias**

The location of the gene defect in this group is 5q32-q33. The group also includes type IB achondrogenesis (lethal form), type II atelogenesis (likewise lethal) and a recently described rare autosomal-recessive form of multiple epiphyseal dysplasia [63].

### **Diastrophic dysplasia**

#### **► Definition**

Autosomal-recessive disorder with severe, disproportionate dwarfism, major deformities of the joints with clubfeet, characteristic abduction of the thumb pronounced kyphoscoliosis of the spine, and often severe kyphosis of the neck.

The name derives from the Greek (*diastrophein* = distortion).

#### **Historical background, etiology, pathogenesis, occurrence**

The disorder was first described in 1960 by Lamy and Maroteaux [52]. The mode of inheritance is autosomal-recessive. It appears to involve an enzyme defect that prevents the formation of normal collagen and regular cartilage tissue. The epiphyseal plates lack the normal columnar cartilage. Calcification is severely disrupted, preventing the usual enchondral ossification. There is a congenital defect of chondrogenesis that can affect any type of cartilage (i.e. including the fibrous cartilage of the ear, larynx or trachea). The disorder is rare in Central Europe (no epidemiological figures are available), but less so in Finland [103].

#### **Clinical features, diagnosis**

The disorder can be diagnosed even at birth. There is disproportionate dwarfism with *contractures* of the joints, severe *clubfeet*, characteristic deformities of the *ears* and abduction of the *thumb*. A cleft palate is also frequently present. The hands are short. The abduction of the thumb is caused by an abnormally short 1st metacarpal, and the thumb subluxates radially at the metacarpophalangeal joint. Clubfeet are invariably present, and here too the 1st metatarsal is short and triangular in shape. The rearfoot is in an extreme varus and equinus position. The joints can either be very stiff or very lax. Contractures predominate in most cases. The hips and knees show flexion contractures, which can severely interfere with walking ability. The hips are often dislocated on both sides. We have also observed the complete absence of the patella.

The elbow joints may be contracted. The spinal changes are particularly severe [97]. Major kyphoscoliosis very

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■ Fig. 4.72a–d. Photographs and x-rays of the hands (a, b) and legs (c, d) of a 16-year old boy with *Ellis-van-Creveld syndrome*. Note the

short phalanges and the genu valgum resulting from the excessively short fibulas

frequently develops (■ Fig. 3.107, ► Chapter 3.1.11). The kyphosing of the cervical spine is particularly problematic, potentially reaching 180° during the first few years of life. This usually results in additional secondary damage to the spinal cord which, combined with the contractures, further restricts the ability to walk. The vertebral arches are usually abnormally shaped (■ Fig. 3.108, ► Chapter 3.1.11). Moreover, the interpedicular distance can decline towards the lumbar level, and the pedicles may be short, as in achondroplasia, resulting in the development of spinal stenosis. Thoracic or thoracolumbar kyphosis is almost invariably present. At the lumbar level this is accentuated by the flexion contracture of the hips.

The severity of the condition varies greatly. Sometimes the dwarfism is extreme, with a final height as low as 80 cm (2 ft 7 in), but in other cases the stunted growth may only be slight, with a final height of 140 cm (4 ft 7 in). On x-rays, the ossification centers of the epiphyses appear at a late stage, and are deformed, flattened or triangular in shape. The metaphyses are widened. A coxa vara is often observed, with widening and irregularities of the femoral neck. The long bones are short and thick, resembling those in achondroplasia. But in contrast with achondroplasia, the appearance of the epiphyseal centers is delayed. The flat bones do not show any changes. The patients are of normal intelligence.



- *Differential diagnosis:* Since the stunted growth is apparent even at birth, the disorder must be differentiated from *achondroplasia*, *spondyloepiphyseal dysplasia* and *chondrodystrophia calcificans congenita*. In view of the joint contractures it must also be differentiated from *arthrogryposis*.

#### Prognosis, treatment

The severity of the disorder is highly variable. Milder forms are often observed in Finland [89]. Patients with severe forms are greatly disabled, although life expectancy does not appear to be significantly restricted.

- No known *treatment* exists for the *underlying disease*. *Orthopaedic treatment:* Numerous orthopaedic problems are posed by diastrophic dwarfism and the treatment is very demanding. The deformities are difficult to treat and have a great tendency to recur.

The *clubfeet*, with the extreme equinus deformity, should be surgically corrected during the first year of life after pretreatment with cast straightening. The aim is to produce a plantigrade standing position. If, as often occurs, the *patella is dislocated*, this should be reduced to its normal position with a soft tissue release. This procedure should be performed at a relatively early stage since the reduction is always more difficult later on. Occasionally the patella is missing. The *hips* are usually very contracted and, as with a teratologic dislocation, the reduction is difficult. Whether a hip reduction should even be attempted must be discussed with the parents. In view of the cartilage changes the prognosis for the hips is poor even when they are centered.

Special attention must be paid to the *spine*. Some patients develop a very severe *kyphosis* even in infancy, particularly at cervical level. An attempt can be made to influence the kyphosis with a cervical collar or a halo. In some cases, however, an early anterior and posterior spondylodesis cannot be avoided. A cervical spina bifida occulta is also often present. A severe kyphosis also often develops at thoracic level. Attempts to keep this within limits with a corset do not always prove successful. In recent years we have tried to control such kyphoses with titanium ribs (VEPTR instrumentation; ► Chapter 3.1.7). Nor is it easy to achieve success with this method either because the titanium ribs themselves have a kyphosing effect. If they are used appropriately, however, the kyphotic correction can prove successful. After the child has started walking, the orthopaedist's attention should turn to the *knee contractures*. Since the energy required to walk with flexed knees is extremely high, an attempt should be made to stretch the knees, possibly using the Ilizarov apparatus or the Taylor Spatial Frame (► Chapter 3.3.13). A recurvation osteotomy is occasionally required in order to achieve sufficient extension.

#### 4.6.2.7 Group of dyssegmental dysplasias

Dyssegmental dysplasia occurs in two autosomal-recessive forms (Silverman-Handmaker and Rolland-Desbuquois). Since both forms are lethal they will not be discussed any further at this point.

#### 4.6.2.8 Type II collagenopathies

This group includes the lethal forms of achondrogenesis II (Langer-Saldino) and hypochondrogenesis and the congenital forms of spondyloepiphyseal dysplasia, the Kniest and Stickler syndromes [29].

These disorders affect type II collagen, which makes up 80% of the collagen in the cartilage matrix. The location of the gene defect is 12q13.1-q13.3.

#### *Achondrogenesis II and hypochondrogenesis*

Very severe, disproportionate stunted growth, hydrops, death at birth or the neonatal period.

#### *Spondyloepiphyseal dysplasia*

##### Definition

Inherited disorder with disproportionate dwarfism that mainly affects the spine, but also involving the epiphyses of the long bones. The *congenital types* can be diagnosed at birth and the mode of inheritance is autosomal-dominant, whereas the *late-onset types* are X-linked or autosomal-recessive conditions (► Chapter 4.6.2.10).

- *Classification, etiology, occurrence:* Although a clear inheritance pattern has been found for both forms, most cases occur sporadically and are attributable to new mutations. Histochemical investigation has shown a degenerative lysosomal process in the formation of proteoglycan. As mentioned above, type II collagen is affected [116] and the gene defect is on 12q13.1-q13.3. Certain cases show similarities with mucopolysaccharidosis [18]. A recent report has described a new autosomal-dominant form [96]. Various epidemiological studies have calculated the prevalence of all forms as ranging from 7 to 11.1/million [115].

- *Clinical features, diagnosis:* Patients with the *congenital form* have a pronounced *coxa vara* and dwarfism even at birth. Hip flexion contracture and hyperlordosis of the lumbar spine are also observed. On the x-ray the femoral neck configuration is reminiscent of femoral neck pseudarthrosis, with a pronounced varus position. The greater trochanter is displaced upwards, while the femoral heads are normally centered, but flattened and exhibiting a pear-shaped deformity (► Fig. 3.215, ► Chapter 3.2.7). The vertebral bodies are deformed and drawn out in the shape of a tongue (► Fig. 3.109, ► Chapter 3.1.11). The dens is dysplastic, and there is a risk of *atlantoaxial instability*. *Kyphoses* and *scolioses* frequently develop at a later date. The



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patient usually appears pigeon chested. The other epiphyses can also be affected. Possible *associated abnormalities* include cleft lip and palate, deafness, myopia, cataracts and clubfeet. The hips are exposed to a considerable risk of osteoarthritis.

- **Differential diagnosis:** The most difficult task is to distinguish SED on the one hand from *mucopolysaccharidosis* and, on the other, from *pseudoachondroplasia*. Both conditions show similar spinal changes.
- **Treatment:** No known treatment of the *underlying condition* is known, nor is there currently any prospect of such a treatment being developed.

From the orthopaedic standpoint the most serious problem is the *atlantoaxial instability*. An occipitocervical spondylodesis is often required at a very early stage. The *coxa vara* must also be corrected at an early stage by a valgization osteotomy, in order to avoid any further exacerbation of the varus position and the formation of a pseudarthrosis as a result of highly abnormal weight bearing. At the spinal level, *scoliosis* and *kyphosis operations* are indicated unless the corresponding deformity can be managed with conservative measures (corset), which is not usually the case with the congenital form.

#### **Kniest syndrome**

This syndrome was described in 1952 by Kniest [49]. It is a very rare disease and only a few isolated reports appear in the literature. The parents are often related. In contrast with the above-mentioned type II collagenopathies, the Kniest syndrome involves deletions of several amino acids [29].

- **Clinical features, diagnosis:** At birth the *trunk* is of normal length, whereas the extremities are shortened. But the development of scoliosis and kyphosis subsequently cause the trunk to become shortened as well. The *face* is unusually flat, with hypertelorism and flattening of the nose. Both the metaphyses and epiphyses of *long bones* are affected. Some patients suffer from severe flexion contractures of *knees* and *hips*. *Deafness* and *myopia* may also be present. Osteoarthritis can occur at a very early age as a result of the epiphyseal changes. In addition to the invariably present deafness, vision is also permanently impaired as a result of retinal detachment.
- No *treatment* exists for the underlying condition. The orthopaedic treatment involves correction of the joint deformities and the *kyphoscoliosis*.

#### **Stickler syndrome**

The Stickler syndrome, also known as *arthro-ophthalmopathy* is a genetically and phenotypically heterogeneous group of diseases, with variations in both the signs and symptoms and the age at onset. Most striking from the *clinical* standpoint are the ocular problems (myopia, retinal detachment, glaucoma and blindness) [29]. The patient typically suffers from micrognathia/retrognathia, usually with a median cleft palate and hearing loss. The patients are of normal

height and sometimes show a marfanoid habitus. Mitral valve prolapse is often present. The joint involvement manifests itself clinically as joint stiffness and pain. The *x-rays* of some patients will show slight dysplasia of the vertebral bodies and relatively flat epiphyses with remodeling defects and fairly narrow diaphyses (■ Fig. 4.73). Signs of osteoarthritis are observed as early as young adulthood.

#### 4.6.2.9 Type XI collagenopathies

##### **Otospondyloomegaepiphyseal dysplasias (OSMED)**

The site of the gene defect in these autosomal-recessive disorders is 6p21.3. They affect the type XI collagen, which is jointly responsible for the formation of collagen fibrils. The *signs and symptoms* include sensorineural hearing loss, metaphyseal abnormalities, vertebral body abnormalities, restricted joint mobility, flat nose, flat midface/molar hypoplasia, mesomelic micromelia, nostrils facing forward, submucosal cleft palate/cleft uvula, failure to thrive, susceptibility to infections, kyphosis, capillary hemangioma/portwine stain, abnormal lacrimal ducts, strabismus/squint, ventricular septal defect, and fused carpal bones.

#### 4.6.2.10 Other spondyloepi-(meta)-physeal dysplasias

All of the hereditary disorders in this group are very rare:

- **X-linked dominant inherited spondyloepiphyseal dysplasia tarda** (gene locus Xp22.2-p22-1). This condition only affects boys. They are normal in size at birth, but gradually become disproportionately small, with a short trunk, as growth progresses. X-rays show platyspondylia and coxa vara. Early cases of osteoarthritis are common.
- **Progressive pseudorheumatoid dysplasia** starts at school age with swelling of the joints. The radiographs show widening of the epiphyses, narrowing of the joint space and flattening of the vertebral bodies. The mode of inheritance is autosomal-recessive and the gene locus is 6q22-q23. Increasing destruction of the joints occurs resulting in an early need for artificial joint replacement.



■ Fig. 4.73. X-ray of the hips in an 8-year old boy with *Stickler syndrome*, with remodeling defects both in the femoral heads and the acetabula

#### 4.6.2.11 Multiple epiphyseal dysplasia and pseudoachondroplasia

##### **Multiple epiphyseal dysplasia (MED)**

###### ➤ Definition

Autosomal-dominant hereditary disease with highly variable severity. It involves a disruption of enchondral ossification of the epiphyses, particularly the femoral head epiphysis. Three forms are distinguished:

- *severe form* according to *Fairbank* with a delayed appearance of the ossification centers of most epiphyses, crude fingers and toes and moderately stunted growth [23],
- *milder form* according to *Ribbing* with only minimal involvement of the fingers and toes. In the majority of cases only the femoral heads are significantly affected [98],
- *localized mild form* according to *Meyer* with exclusive involvement of the femoral heads (*dysplasia epiphysealis capitis femoris*) [71].

The *Ribbing* and *Meyer* types only affect the hips and are discussed in ► Chapter 3.2.7 (■ Fig. 3.211 and 3.212). Consequently, only the *Fairbank* type, which is very rare, will be described below. It is an autosomal-dominant condition and the gene locus is 19p13.1.

- The disorder is not usually *diagnosed* until early childhood. Affected patients show moderately *stunted growth* (a final height of 145–160 cm or 4 ft 9 in–5 ft 3 in, can be expected). Hip problems often occur, prompting the radiographic investigation. The hip symptoms are load-related.
- The *x-ray* shows delayed and irregular ossification of the femoral head center, which is usually widened, although the joint cartilage is not thickened. Coxa vara is occasionally present. The x-ray of the hand shows crude phalanges and typical epiphyseal changes. The ratios of wrist length to wrist width and of the height of the femoral head epiphysis to the width of the metaphysis are also abnormal [38].
- *Differential diagnosis*: Distinguishing multiple epiphyseal dysplasia from *Legg-Calvé-Perthes disease* can sometimes be difficult. As a rule, the presence of an epiphyseal dysplasia should always be suspected if both femoral heads are involved. In MED the joint cartilage is not thickened, and there is no lateral calcification or subluxation. The course of the disease during growth is more benign and the metaphyses are not usually involved. By contrast, the acetabulum is more severely affected than in Legg-Calvé-Perthes disease. Certain mixed forms can also evidently occur [90]. *Spondyloepiphyseal dysplasia* can be ruled out if there is no visible changes in the vertebral bodies. In *hypothyroidism* the femoral heads are abnormally small and show irregular ossification. If the clinical presentation is atypical, the bone age estimated from

the hand x-ray can provide a clue to the diagnosis as this is greatly retarded.

- The *long-term prognosis* is not particularly good if, on completion of growth, a flattened and widened femoral head with acetabular changes is present. In these cases, incipient osteoarthritis of the hip can be expected to occur as early as 30 years of age [112].
- The negative course of Fairbank multiple epiphyseal dysplasia cannot be influenced to any great extent by *therapeutic measures*. Since osteotomies do not improve the prognosis, early total hip replacements are often required.

##### **Pseudoachondroplasia**

This is an inherited disorder with proportionate dwarfism, and in which the epiphyses and metaphyses are altered and the spine shows pathological features. The condition was first differentiated from spondyloepiphyseal dysplasia in 1959 by Maroteaux and Lamy [65]. It is associated with a mixed *inheritance pattern*, being autosomal-dominant in most cases and, more rarely, autosomal-recessive. The gene locus is 19p12–13.1. Most cases occur sporadically as new mutants. An abnormal proteoglycan is stored in the cartilage tissue. Accordingly, this is actually a storage disease that differs fundamentally from achondroplasia, which only involves impaired enchondral growth, but no storage of abnormal proteoglycan.

- The prevalence of pseudoachondroplasia has been calculated at 4.3/million inhabitants [115].
- From the *clinical* standpoint the severity of the disease varies greatly. Pseudoachondroplasia is not usually diagnosed until the age of 2–3 years. The *shortening* affects both the extremities and trunk, and is therefore less disproportionate than in achondroplasia. On the other hand, the dwarfism overall is more pronounced, and a final height of just 100–115 cm is reached. Striking findings include marked ligament laxity, which is associated with genua valga and recurvata, instability of the ankles and pronounced pes planovalgus. A kyphoscoliosis can develop in a trunk that is already shortened in any case. The patient is of normal intelligence.
- *X-rays* reveal a spine with flattened, oval vertebral bodies, in some cases with a triangular shape. Hypoplasia of the dens is a typical feature. Functional views often show atlantoaxial instability, and kyphoscoliosis is often present. The long bones are short and broad. Not only do the metaphyses appear swollen and bulb-like, but there is also delayed development of the epiphyses with small fragmented ossification centers (■ Fig. 4.74). The hips and knees are usually the worst affected joints, with coxae varae and dysplasia of the acetabula. The triradiate cartilage is wide and ossifies at a late stage. In contrast with the situation in achondroplasia, the greater sciatic foramina are normal. At knee level, and as in achondroplasia and hypochon-

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droplasia, the fibula is relatively overlong, with the possible consequent formation of genua vara.

- **Differential diagnosis:** The differences compared to *achondroplasia* are listed in ■ Table 4.20. Otherwise, pseudoachondroplasia has to be differentiated from *spondyloepiphyseal dysplasia* and *diastrophic dwarfism*. The latter is characterized by joint contractures, whereas the ligament laxity predominates in *pseudoachondroplasia*.
- The *prognosis* is good and life expectancy is normal. Since the epiphyses (in contrast with achondroplasia and hypochondroplasia) are affected, joint changes and osteoarthritis occur prematurely.
- No specific *treatment exists for the underlying condition*. *Orthopaedic treatment:* Axial restoration is particularly important if the pseudoachondroplasia is associated with major deviations. Premature joint damage occurs even if the axes are normal, and this is further promoted by pronounced axial deviations. If osteoarthritis has become established, *total hip replacements* may be indicated.

Under no circumstances should *bilateral leg lengthening* be undertaken to achieve a normal height in pseudoachondroplasia. On the one hand, the overall gain in length would need to be 50 cm, which is unrealistic in any case. On the other hand, the joints are



■ **Fig. 4.74.** AP x-ray of both legs of a 7-year old girl with *pseudoachondroplasia*, with swollen bulb-like metaphyses in the long bones and excessively long fibulas. In addition, the epiphyses are irregular and consist of several ossification centers

particularly at risk of osteoarthritis, and the problem is greatly aggravated by any lengthening procedure. Occasionally, orthotic, or even surgical, treatment is indicated for scolioses and kyphoses.

#### 4.6.2.12 Chondrodysplasia calcificans punctata *Chondrodysplasia calcificans punctata, rhizomelic type*

This is an autosomal-recessive hereditary disease with 3 subtypes depending on the gene locus in each case (type 1: 6q22-q24; type 2: 1q42; type 3: 2q31). There is a deficiency of alkyl glycerone phosphate synthetase. Clinically, the disease manifests itself as a rhizomelic micromelia (i.e. shortening primarily of the proximal parts of the extremities in the thigh and upper arm), restricted joint mobility, epicanthus, cataract/lenticular opacity, flat face, ichthyosis and sparse/thinning hair/baldness, metaphyseal abnormalities, microcephaly, microplasia, scoliosis/kyphosis, spina bifida occulta, severe mental retardation. The spinal deformity, in particular, is problematic, since it can lead to severe kyphoses, which are almost impossible to control in early childhood and require early anterior and posterior fusions [69].

#### *Zellweger syndrome*

An autosomal-recessive inherited disorder that proves lethal shortly after birth. Clinical features include hypotonia, dysmorphic skull, poor eyesight, hepatomegaly, dysphagia, severe mental retardation. There are isolated reports of patients surviving for longer periods. Widely varying genes have been blamed for the peroxin deficiency associated with this syndrome.

#### *Chondrodysplasia calcificans punctata, Conradi-Hünemann type*

Chondrodysplasia calcificans punctata was described in 1914 by Conradi [14]. This was a rhizomelic type. In 1931, Hünemann described the milder, non-lethal form [35], which is an X-linked dominant condition. The gene locus is Xp11 and most cases are sporadic. This milder form (Conradi-Hünemann) must be differentiated from the lethal forms (Conradi) [9].

- **Clinical features, diagnosis:** The face is flat, the skin dry, congenital *cataracts* are observed, and the extremities are asymmetrically shortened. The x-rays show *stippled calcification* on the ends of the long bones in the epiphyses, but also in the vertebral bodies and the ilium. Scolioses and *kyphoses* can develop.
- The *prognosis* for this mild form is relatively good, and the stippled calcification disappears around the age of 5. However, pronounced asymmetry of the extremities occurs with substantial leg length discrepancies.
- *Treatment* in this condition primarily involves *leg length equalization*, either by epiphysiodeses or targeted lengthening procedures.

#### 4.6.2.13 Metaphyseal dysplasia

Metaphyseal dysplasia occurs in various forms, e.g. as *McKusick cartilage-hair hypoplasia*, an autosomal-recessive syndrome (gene locus 9p21-p12) with very stunted growth, fine hair, crooked legs and general ligament laxity. Since the immune system is impaired, these patients tend to suffer from infections and tumors. Radiologically flattened epiphyses and very delicate, in some cases irregular, bones.

An autosomal-dominant form was described by *Jansen*. Characteristic features include dwarfism, crooked legs and forearms, irregular metaphyses, wide epiphyses and diffuse osteopenia.

#### 4.6.2.14 Spondylometaphyseal dysplasia

Of the diseases in this group, the autosomal-dominant type named for *Kozlowski* is the most well known. Metaphyseal and vertebral abnormalities are combined with a small stature, kyphosis, short hands and feet, as well as a gait abnormality. Skeletal maturation is distinctly retarded.

#### 4.6.2.15 Brachyolmia spondylodysplasias

All types of *brachyolmia* (Greek for »short trunk«) are characterized by small stature, generalized platyspondylia without major epiphyseal or metaphyseal changes. Four types with differing modes of inheritance are distinguished. They are usually associated with scolioses and/or kyphoses.

#### 4.6.2.16 Mesomelic dysplasias

»Mesomelia« involves short extremities with predominant shortening of the middle (intervening) segments. The most well known type is *Leri-Weill dyschondrosteosis*, which is characterized by bowing of the radius and tibia, dislocations in the area of the elbow and Wrist (with Madelung deformity), fibular hypoplasia, shortening of the forearms and lower legs. The gene locus is on the common region of the X- and Y-chromosomes (Xpter-p22.32). Accordingly, the Leri-Weill syndrome occurs particularly in connection with Turner syndrome [6]. If a Madelung deformity occurs in combination with stunted growth, then the possibility of this syndrome should be borne in mind.

#### 4.6.2.17 Acromelic dysplasias

*Trichorhinophalangeal syndrome* is an autosomal-dominant disorder in which *brachyphalangia* is associated with a conical change of the phalangeal epiphyses as well as a *characteristic face* with a pear-shaped nose with a long philtrum and sparse hair growth. *Ossification abnormalities* of the femoral heads that are reminiscent of epiphyseal dysplasia or Legg-Calvé-Perthes disease are also frequently present.

The group of acromelic dysplasias also includes the hereditary forms of *brachydactyly* (6 types). All are autosomal-dominant conditions, and some are associated with other abnormalities and mental retardation.

#### 4.6.2.18 Acromesomelic dysplasias

Rare forms of dwarfism with short extremities in which the segments away from the trunk are mainly affected. The long bones are shortened and widened, the radius bowed and its head usually displaced. Platyspondylia with kyphosis are also frequently present. Several, usually autosomal-recessive, forms of this hereditary condition exist.

#### 4.6.2.19 Dysplasias with predominant involvement of flat bones

##### *Cleidocranial dysplasia*

— *Synonyms:* Marie-Sainton syndrome, cleidocranial dysostosis

This is a disease involving abnormal periosteal ossification, which primarily affects the clavicles, skull and pelvis. The gene locus of this autosomal-dominant condition is 6p21.

- *Clinical features:* Although the flat bones are primarily affected, enchondral ossification is also impaired to a lesser extent, resulting in moderately stunted growth. The missing gene regulates osteoblast differentiation.
- This dysplasia is *diagnosed* on the basis of delayed ossification of the skull and the resulting very wide fontanelles, which can remain open until adulthood. The skull appears very large, with a pronounced frontal bulge, and the face rather small. The shoulders appear narrow because of the missing clavicles. The thorax is narrow and the symphyses are very wide. Additional features include short middle phalanges of the little fingers, delayed bone maturation, coxa vara, occasionally scolioses, hearing loss and slight mental retardation.
- *Treatment:* Orthopaedic treatments are needed to correct a coxa vara and, occasionally, scoliosis.

#### 4.6.2.20 Dysplasias with bent bones

##### *Camptomelic (or camptomelic) dysplasia*

The gene locus in this autosomal-dominant condition is 17q24.3.-q25.1.

- *Clinical features:* Typical features include missing or hypoplastic clavicles, hypoplasia of the genitalia (pseudohermaphroditism), severe bowing of the femur and tibia, cleft palate, clubfeet, delayed bone maturation, radiohumeral synostosis, fibular hypoplasia, craniosynostoses, general ligament laxity, knee dislocations, macrocephaly, platyspondylia, small scapulae, possible severe kyphosis.



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- *Treatment* is based on the deformities. The bent legs must be straightened and fixed with telescopic nails (■ Fig. 4.77 and 4.87), while spinal deformities must be corrected, possibly at an early stage, with titanium ribs (■ Fig. 3.86, ► Chapter 3.1.7).

### Stüve-Wiedemann dysplasia

This rare condition, described by Stüve and Wiedemann in 1971 [111], involves pronounced bowing of the long bones, stunted growth, camptodactyly, occasionally dyspnea and hyperthermia, and mental retardation. Most patients die during childhood, usually in connection with a bout of fever. This autosomal-recessive condition is frequently found in Arab countries.

### 4.6.2.21 Dysplasias with multiple dislocations

#### Larsen syndrome

##### 🔍 Definition

This inherited disorder is characterized by a flat face, bulging forehead, hypertelorism and multiple congenital dislocations (usually hips, knees, poss. radial head) and segmentation defects of the cervical spine.

The syndrome was first described by Larsen et al. in 1950 [54]. Both autosomal-dominant and autosomal-recessive inheritance patterns have been observed. The gene locus in the more common dominant variant is 3p21.1–014.1. It appears to involve a generalized mesenchymal defect that affects all of the connective tissues. The disease is very rare and the literature only contains isolated cases. There is a striking accumulation in La Réunion, where 38 cases have been described [55]. We ourselves have encountered four cases.

- As regards the *clinical* picture, the knee and hip dislocations are apparent even at birth. Not infrequently, these are combined with clubfeet or pes equinovagus. The tarsal bones often show multiple ossification centers. A tracheomalacia in infancy and early childhood can cause major problems. The spine is affected by malformations that lead to kyphosis or scoliosis (■ Fig. 3.110, ► Chapter 3.1.11). An atlantoaxial instability may be present [40, 61]. The face is likewise very striking, with a flat nose and bulging forehead. Heart defects are not unusual [55]. The intelligence of the children is usually normal, although a few are mentally retarded.
- *Differential diagnosis*: Larsen syndrome can be confused with arthrogryposis multiplex congenita, in which the joints can also be severely deformed or dislocated. A pronounced stiffness is generally present in arthrogryposis however, which is not the case with Larsen syndrome. Since significant ligament laxity also occurs in Ehlers-Danlos syndrome, this must also be differentiated from Larsen syndrome.

- *Prognosis*: Some children die at an early stage because of heart defects or tracheomalacia [55]. In the series from La Réunion, 14 of the 38 children died from such complications. The life expectancy of the remaining children does not appear to be significantly restricted.

- *Orthopaedic treatment*: The dislocated joints should be treated at a very early stage [100]. The hips must be reduced openly, which is technically difficult as these are highly unstable joints. Nevertheless, a stable centering of the femoral heads certainly must be attempted. The knees can usually be treated conservatively, and reduction by slow straightening generally proves successful. The feet also usually require surgery to correct the pronounced equinus deformity.

! **It should be noted, in connection with the operations, that the tracheomalacia and cervical changes pose particular anesthesiological problems.**

The spine should be monitored carefully. In addition to fixed scoliosis, an atlantoaxial instability can also develop, requiring early occipitocervical fusion.

### 4.6.2.22 Dysostosis multiplex group

#### Mucopolysaccharidoses

##### 🔍 Definition

The mucopolysaccharidoses form a group of conditions involving defective lysosomes. Lysosomes are enzymes involved in mucopolysaccharide metabolism, and their failure can lead to the storage of mucopolysaccharide components. Six types are distinguished, depending on the enzyme defect in each case.

- *Synonym*: Dysostosis multiplex

#### Classification, occurrence, etiology

■ Table 4.21 shows the *classification* of mucopolysaccharidoses in six types, based on the enzyme defect and listing the main features. The authors of a 30-year study in Great Britain calculated a prevalence for mucopolysaccharidoses of 1.7/million inhabitants [115]. The mucopolysaccharidoses involve lysosomal defects. As can be seen from ■ Table 4.21, a different enzyme is affected in each type.

#### Historical background

*Type I* mucopolysaccharidosis was first described by Gertrude Hurler in the year 1919 [37]. The term »gargoylism« was coined by Ellis, Sheldon and Capon [19], and refers to gargoyles, those grotesque figures on Gothic cathedrals that spit out the rainwater. A description of *type II* was published by Hunter in 1917 [36], while *type III* (Sanfilippo syndrome) was first mentioned in 1961 by Harris and described in 1963 by Sanfilippo [33, 104]. *Type IV* was independently described by both Morquio [74] and Brailsford [10] in 1929. *Type V* was mentioned by the ophthalmologist Scheie in 1962 [105]. *Type VI* was mentioned for the first time in 1963 by Maroteaux [66].



### Clinical features, diagnosis, differential diagnosis

#### — Clinical features:

Those mucopolysaccharides that are not converted directly by enzymes, i.e. heparan sulfate, dermatan sulfate and keratan sulfate are stored and excreted in the urine in excessive quantities. This can be established by biochemical analyses. Most mucopolysaccharidoses affect height. The skeletal changes affect the skull, which is enlarged, and the sella turcica, which is widened. The clavicles are wide, particularly towards the sternoclavicular joint, and the ribs are broader at the front than the back.

Scolioses are often present. The vertebral bodies are flattened and oval with very irregular ends. In Morquio disease, the vertebral bodies protrude centrally in the shape and the apophyseal ring is notched. A highly characteristic finding is thoracolumbar kyphosis with vertebral slippage in this area (■ Fig. 3.106, ► Chapter 3.1.11) [91]. The ilium is widened, and coxa valga is often present. In type IV mucopolysaccharidosis (Morquio), the femoral head epiphysis is also often very irregular, resembling a case of Legg-Calvé-Perthes disease (and often being confused with this condition; ■ Fig. 3.216, ► Chapter 3.2.7).

- Another possibility that must be considered in the differential diagnosis is multiple epiphyseal dysplasia. The long bones are shortened, as are the scapulae. The

metacarpals taper to a point at their distal end, and their epiphyses are narrowed. The individual types of mucopolysaccharidoses cannot be differentiated solely on the basis of the skeletal dysplasias, but rather on the basis of the biochemical analyses and the other clinical findings.

- The condition can usually be *diagnosed* during the first few months of life. Hypertelorism is usually present, the cornea is cloudy and hearing loss is observed. The nose is broad, and the children often suffer from chronic rhinitis and have to breath through their mouth. In most forms the neck is shortened, and psychomotor development is impaired to a greater or lesser extent. The heart and lungs are also often impaired and limit the life expectancy.

### Treatment

- *Treatment of the underlying disease:*

Advances have been made in recent years in the treatment of mucopolysaccharidoses. Three approaches have been pursued. Successful results have recently been obtained with enzyme replacement therapy, particularly in type I patients. The transplantation of *homologous hematopoietic stem cells* has also proved successful in types I and II, whereas the results for types III and IV have been disappointing to date [76]. *Gene therapy* is still in its initial stages, but certain approaches are highly promising. Special attention

■ Table 4.21. Classification of mucopolysaccharidoses

Type	Enzyme defect	Secreted substance	Inheritance	Face	Height	Skeletal changes	Mental retardation	Prognosis
MPS I (Pfaundler-Hurler)	α-L-iduronidase	Dermatan sulfate ++ heparan sulfate+	Autosomal-recessive	Gargoylism	Moderately small stature	Thoracolumbar kyphosis	Severe	Death usually at the age of approx. 10 years due to cardiopulmonary problems
MPS II (Hunter syndrome)	Sulfoiduronate sulfatase	Heparan sulfate++ dermatan sulfate +	X-linked recessive (all patients male)	Gargoylism, less pronounced than in type I	Moderately small stature	Not very pronounced	Moderate	Survival up to the third decade
MPS III (San-Filippo syndrome)	N-heparan sulfatase or α-acetylglucosaminidase	Heparan sulfate++	Autosomal-recessive	Little changed	Normal	Widening of the medial ends of the clavicles	Severe	Survival up to the third or fourth decade
MPS IV (Morquio syndrome)	N-Ac-Gal-6 sulfate sulfatase	Keratin sulfate++	Autosomal-recessive	Coarse, wide mouth, prominent maxilla	Pronounced dwarfism	Platyspondylia, kyphosis, irregular femoral head epiphyses	None	Almost normal life expectancy
MPS V (Scheie syndrome)	α-L-iduronidase	Dermatan sulfate ++ heparan sulfate+	Autosomal-recessive	Gargoylism	Normal	Small epiphyses on the hands	None	Almost normal life expectancy
MPS VI (Maroteaux-Lamy syndrome)	N-Ac-Gal-4 sulfatase	Dermatan sulfate++	Autosomal-recessive	Coarse	Pronounced dwarfism	Thoracolumbar kyphosis	None	Shortened life expectancy

#### 4.6.2 · Skeletal dysplasias

should always be paid to the *cardiopulmonary problems*, which must be treated accordingly.

##### — Orthopaedic treatment:

An important problem is the atlantoaxial instability, since this occasionally results in constriction of the spinal cord in the upper cervical spine, making treatment essential. Usually an occipitocervical spondylosis is performed in such cases. The atlantoaxial instability must also always be borne in mind during any induction of anesthesia. The second problem that occasionally requires treatment is the *thoracolumbar kyphosis*, which generally leads to spondylolisthesis in this area. Posterior tension-band wiring prevents any further slippage and accentuation of the kyphosis. If the *coxa vara* is very pronounced, a valgization osteotomy may be indicated. A severe genu valgum must also be corrected. In some cases the valgus deformity of the rearfoot is severe enough to require a subtalar or even a triple arthrodesis.

##### 4.6.2.23 Dysplastic slender bone group

Only isolated reports exist for the autosomal-recessive *microcephalic osteodysplastic dysplasia*, which occurs in three types. The patients show moderately stunted growth (which only becomes apparent during the course of growth), have short fingers, are microcephalic but with normal intelligence, the bones are generally very thin, and *coxa vara* and abnormally shaped epiphyses are typical features of the disease.

##### 4.6.2.24 Dysplasias with decreased bone density *Juvenile osteoporosis*

In juvenile osteoporosis there is an imbalance between bone formation and bone resorption [92]. Overall bone mass decreases as a result of the increased resorption by osteoclasts, although there are no qualitative abnormalities. The process mainly affects the cancellous bone [92]. The laboratory tests do not show any major abnormalities apart from a negative calcium balance. This is an extremely rare condition [12] that is probably caused (like osteogenesis imperfecta) by an anomaly of type I collagen as a result of a mutation [17].

- The *clinical* onset of juvenile osteoporosis as a temporary, self-limiting disease occurs during puberty. The spine is always affected [12]. The principal symptoms are dull back pain, possibly with symptoms in the extremities, particularly the feet, with a consequent adverse effect on gait. Muscle weakness may also be present.
- The main *complications* are spontaneous fractures of the spine, which can ultimately result in fixed hyperkyphosis [12]. Fractures also often occur in the extremities, particularly in the metaphyseal area.

- The *x-ray* shows a normal, but possibly diminished, trabecular structure. The cortices are thin, and the vertebral bodies may show depressions, giving them the appearance of »fish vertebrae«. In the limbs, the osteoporosis is usually at its most pronounced in the metaphyses (■ Fig. 4.75).
- An important task in the *differential diagnosis* is to rule out *secondary osteoporosis*, which occurs in the following disorders: hyperthyroidism, hyperparathyroidism, hypogonadism, Cushing syndrome, homocystinuria, vitamin C deficiency, rickets, Lowe syndrome, uremia, Turner syndrome, leukemia, lymphoma and osteogenesis imperfecta. Treatment with *corticosteroids*, for instance in juvenile rheumatoid arthritis or Langerhans cell histiocytosis, can lead to severe osteoporosis.
- No specific *treatment* exists. In terms of drug treatment, successful results have been obtained with the bisphosphonate pamidronate (Aredia) [108]. An important requirement is to preserve mobility and encourage plenty of exercise and movement. A corset treatment is sometimes needed to correct a pronounced kyphosis. In the event of a fractured extremity, the possibility of inactivity-induced osteoporosis should be avoided. Where possible, such fractures should not be treated with cast fixation, and internal plate fixation is also highly inadvisable. A more appropriate response is to use intramedullary load-bearing implants, provided these are compatible with open epiphyseal plates (e.g. flexible intramedullary nail).



■ Fig. 4.75. Lateral x-ray of the spine of a 5-year old boy with *idiopathic juvenile osteoporosis*. Note the multiple compression fractures with vertebral bodies of reduced height

## Osteogenesis imperfecta

### ➤ Definition

Group of autosomal-dominant or autosomal-recessive disorders with abnormal bone fragility, blue sclerae, hearing loss, general ligament laxity and increased vulnerability of the skin to a greater or lesser extent.

— **Synonyms:** Fragilitas ossium, osteopsathyrosis idiopathica, Lobstein disease, Vrolik disease, brittle bone disease

### Historical background

The disease with increased bone fragility has been known since ancient times, although without any distinction being made from rickets. The first detailed descriptions date back to Lobstein in 1835 [59] and Vrolik in 1849 [114].

### Classification, occurrence

The condition was first classified by *Looser* in the year 1906 [60]. He made a distinction between an osteogenesis imperfecta *congenita*, in which multiple fractures are already present at birth, and a *tarda form*, in which the fractures occur later on. The first form was subsequently referred to as the *Vrolik* type, and the second form as the *Lobstein* type. The current *classification*, based on *genetic factors*, subdivides the condition into five groups (■ Table 4.22). (More recently, as many as seven groups were differentiated, but these are not yet clearly distinctive [94].) Since this genetic classification takes little account of the orthopaedic problems, in 1992 *Hanscom* [32] proposed a *radiological classification*, based on 64 cases, that takes better account of the orthopaedically relevant factors (■ Table 4.23).

The prevalence of osteogenesis imperfecta is 16.3/million inhabitants [115]. The commonest form is type I, type II is less common, while types III–V are extremely rare.

### Etiology, pathogenesis

The underlying problem in osteogenesis imperfecta is the impaired maturation of type I collagen fibers from the reticulin fibers. While osteoblast activity is brisk, the cells are incapable of forming normal collagen. The »cross-links« play an important role in the maturation of the collagen and their formation is impaired in osteogenesis imperfecta, thereby preventing the production of polymerized collagen. The enzyme defect appears to be different in the various types. Histological examination reveals thin bone trabeculae and decreased ground substance. The bone possesses numerous areas of fibrous bone with an abnormal proteoglycan content. Fracture healing is not impaired, and very large amounts of callus are formed as a result of the increased production of cartilaginous callus.

### Clinical features, diagnosis

The clinical manifestation varies greatly depending on the type involved. The orthopaedist rarely encounters *type II*



■ Fig. 4.76. X-ray of both legs of a 3-year old boy with *osteogenesis imperfecta* and bowing of the long bones after suffering multiple fractures

as the affected children do not survive. This corresponds to type E or type F according to *Hanscom* [32]. Multiple fractures even occur during the delivery process. The skull is soft and the ribs are not ossified. Death occurs as a result of intracranial bleeding and lung collapse. Skeletal deformities occur in varying degrees in the other types (*type I, III, IV, V*) (■ Fig. 4.76).

The *classification according to Hanscom* [32] helps us assess the severity of the condition. In *Hanscom type A* the vertebral bodies show normal contours, and the extremities, particularly the legs, are only slightly bowed. In *type B* there is clear bowing of the upper and lower legs, with widened cortices. The vertebral bodies are biconcave, and scoliosis and/or kyphosis not infrequently develops. An additional factor in *type C* is the development of acetabular protrusion around the age of ten years. In *type D* cystic changes are observed from the age of five years on x-rays of the distal femur and proximal tibia, and the epiphyseal plates close very prematurely. Very serious spinal deformities are regularly present in *types C* and *D*. In *type E*, additionally, the cortices of the long bones are not ossified, while in *type F* the cortices of the ribs are also missing. Neither of these two types is compatible with survival.

The following *non-osseous signs and symptoms* may be observed: The *sclerae* are blue. Although the sclerae of all neonates are bluer than those in small children, the blue sclerae persist in patients with osteogenesis imper-

■ Table 4.22. Genetic classification of osteogenesis imperfecta

Type	Inheritance	Characteristic features	Frequency	Gene locus
I	Autosomal-dominant	Generalized osteoporosis Abnormal bone fragility Blue sclerae and deafness in adulthood A distinction is made, depending on whether teeth formation is impaired, between type A (no impairment) and type B (impaired dentinogenesis)	1/30,000	17q21-q22
II	Autosomal-dominant	Extreme bone fragility Death at birth or in early childhood (lethal form) Delayed cranial ossification	1/60,000	7q22.1
III	Autosomal-recessive	Severe bone fragility Severe growth disorder, although adulthood is reached The sclerae are blue at birth, but a normal white color in adulthood	extremely rare	7q22.1
IV	Autosomal-dominant	Osteoporosis and bone fragility vary, less pronounced than in type III Sclerae are blue at birth and normal during adolescence If dentition is normal = type IV A, if dentition is impaired = type IV B	Unknown, extremely rare	17q21-q22
V	Autosomal-dominant	Hypertrophic callus formation and calcifications of the interosseous membrane on the forearm	Unknown, extremely rare	?

■ Table 4.23. Classification of osteogenesis imperfecta. (After Hanscom [32])

Type	Bowing of the long bones	Biconcave vertebral bodies	Cloverleaf pelvis	Cysts	Absent cortex on the long bones	Absent cortex on the ribs	Frequency (%)
A	+						31.3
B	+	+					7.8
C	+	+	+				17.2
D	+	+	+	+			15.6
E	+	+	+	+	+		9.4
F	+	+	+	+	+	+	3.1

fecta (except in types III and IV). The following are also present: general ligament laxity with hypermobility of all joints, atlantoaxial subluxation, flexible flatfeet, habitual patellar dislocation, muscle hypotonia, capillary fragility (positive Rumpel-Leede test), gastrointestinal problems, bulging forehead, triangular face, hyperopia, poss. retinal detachment, impaired dentition with bluish discoloration of the teeth, deafness (in 40% in type I), frequently also otosclerosis, small stature.

— **Differential diagnosis:** Osteogenesis imperfecta can be confused with *campyelic dwarfism*, in which bowing of the long bones also occurs. Fractures increasingly occur also in *cystinosis* and *pyknodysostosis*. It is important to rule out *child abuse*, in which multiple fractures are often observed at the same time.

Distinguishing features are the absence of both the blue sclerae and impaired dentinogenesis. Abnormal bone brittleness is also present in *juvenile osteoporosis*, while the various forms of *osteomalacia* also need to be considered in the differential diagnosis.

#### Prognosis

This has already been discussed in relation to the classification. No further mention need be made of type II (lethal form). In the other types, the ability to walk is of crucial significance for the patient's quality of life (and probably for life expectancy as well). A study in the UK showed that those children who are able to sit up freely by the age of 10 months will very probably be capable of walking and that walking will also be their main form of locomotion (80%),

whereas most children who cannot sit up by 10 months are also rarely able to walk at a later age (only 18%) [16].

### Treatment

No known treatment currently exists for the *underlying condition*. A form of gene therapy aimed at replacing the missing enzyme, at least for type I, is currently in development [80]. Encouraging results have recently been obtained with treatment designed to improve bone strength through the administration of bisphosphonates (pamidronate = Aredia) [93, 118].

The *orthopaedic treatment* must address two main problems: the fractures and bowing of the long bones and the spinal deformities.

- The fractures, which occur mainly during childhood represent a huge problem for the affected patients and their whole environment. In the past such patients had to be immobilized for prolonged periods because of the fractures, thereby compounding the bone brittleness caused by the underlying condition with inactivity-induced osteoporosis. Not infrequently the children lost the ability to walk as a result of this secondary osteoporosis, even though they might enjoy largely normal bone strength on completion of growth thanks to the osteogenesis imperfecta.

Classical fracture management for children involved conservative treatment with cast fixation or surgical procedures with plate fixation or an external fixator. These two operations are totally unsuitable in osteogenesis imperfecta since, in addition to the existing bone brittleness they introduce further predetermined breaking points. Because of their rigidity and the phenomenon of stress shielding they further reduce bone strength, and repeated fractures usually occur during the rehabilitation phase.

Consequently, early attempts were made to splint the bone with nails. The problem that usually arose was that the nails became too short with growth, resulting in new fracture sites at the nail end. A breakthrough emerged with *telescopic nails*, which were introduced by Bailey [4, 45]. These are two-part nails in which a pin slides inside a tube. The nails are inserted into both epiphyses and advanced through the epiphyseal plate into the bone marrow. The pin is then introduced into the tube and the two parts pushed together. As the bone grows the two parts gradually slide apart, thus ensuring that the whole bone remains splinted, until the bone has doubled in length. From the standpoint of its operating principle, this system has obvious advantages since it allows very early weight-bearing and also prevents fractures occurring later on. No major growth disturbance is expected even though the nails are pushed through the epiphyseal plate. In particular, and provided the nail remains in situ, the epiphyseal plate does not close but continues to grow.

Our experience with this system, however, has revealed a number of *disadvantages*: The nails must be inserted via the joints, thereby damaging the joint cartilage. In the long term, the telescopic nails cause additional iatrogenic joint damage, above and beyond the damage that is already produced by the disease, particularly in the ankle joint.

The nails must be custom made for each patient, and the size and thickness of the nails must fit precisely. However, it is often not possible, particularly with fresh fractures, to produce the nails quickly enough. Inserting the nails through bone that is often bowed is difficult. In some cases, the bone must be osteotomied in several places. The bowing tendency of the bone places eccentric loads on the nails. Pushed together, the two components of the nail can become wedged, thereby reducing the telescopic effect.

Since the problem of iatrogenic joint damage is particularly pronounced in the ankle joint, we have developed a special telescopic nail whose two parts can be inserted from the knee and which can be locked in the distal epiphysis (■ Fig. 4.77; [27]). For the femur we have developed a telescopic Gamma nail, which can be inserted from the greater trochanter and is locked with a screw in the distal epiphysis. It allows correction of the varus deformity, which is always present in the bowed femur (■ Fig. 4.87).

Alternatively, two *flexible intramedullary nails* can be used. These nails are more readily available since they can also be used for the management of conventional fractures. They can be inserted from outside the joint through the epiphysis into the bone. In the femur one nail is inserted in each case from the proximal and distal ends. This is not possible in the tibia. The end of the nail is bent and turned inwards into the bone of the epiphysis so as to prevent migration of the nail. The joints remain unaffected in this procedure. The flexible intramedullary nails are much easier to introduce than telescopic nails and custom manufacture is not required. Considerable caution is also indicated during the insertion of the flexible intramedullary nails since the bone is very brittle and can easily perforate. Postoperatively, the patients tend to be feverish even though no infection is present [25].

- *Spinal deformities*: Scolioses, in some cases severe forms often with a pronounced kyphotic component, occur in around half of the patients with type I osteogenesis imperfecta. The treatment of these scolioses is difficult since corset treatment is ineffective. Surgical treatment is usually required. Posterior straightening with spinal instrumentation anchored to as many segments as possible generally proves sufficient (■ Fig. 3.104, 3.105, ▶ Chapter 3.1.11). In very severe forms, an anterior spondylodesis is additionally indicated, particularly if a pronounced kyphotic component is present.





■ **Fig. 4.77a, b.** AP x-ray of both legs of a 10-year old boy with *osteogenesis imperfecta* after suffering multiple fractures. The long bones were splinted by the insertion of *conventional telescopic nails in the femur (a)* and *lockable telescopic nails in the tibia (b)*. The lockable nails are introduced from the knee end, and the inner section is locked in the distal epiphyses by means of a screw. This avoids any damage to the ankle joint

The *anesthesiological risks* are not inconsiderable, and the possibility of atlantoaxial instability should also be borne in mind.

#### 4.6.2.25 Dysplasias with defective mineralization *Osteomalacia (rickets)*

##### ➤ **Definition**

Softening of the bone as a result of inadequate mineralization.

— **Synonyms:** Vitamin D-deficiency rickets, vitamin D-resistant rickets = hypophosphatemia = phosphate diabetes, Albright syndrome

##### **Classification**

- Simple vitamin D-deficiency rickets
- Rickets in malabsorption syndromes
- Vitamin D-resistant rickets

##### **Etiology, pathogenesis**

Vitamin D increases the absorption of calcium in the small intestine. After it has been consumed in foods however (particularly dairy products), vitamin D must be converted by sunlight from its biologically inactive form into the biologically active vitamin D<sub>3</sub>. The hydroxylated form of vitamin D<sub>3</sub> is produced in the liver. The kidneys, in turn, convert this form into dihydroxycalciferol. In vitamin D deficiency the intestinal absorption of calcium is inadequate. The low calcium level stimulates the parathyroid gland, thereby elevating the calcium level, but at the same time reducing the phosphate absorption in the kidney. The low phosphate level leads to inadequate mineralization of bone matrix.

The vitamin D deficiency may be caused either by an inadequate intake or by a malabsorption syndrome. In *vitamin D-resistant rickets* the conversion of the hydroxylated form does not take place owing to a defect either in the glomerular or tubular renal system. *Renal osteodystrophy* is associated with *chronic renal insufficiency*, which leads to *hyperphosphatemia* and consequent *hypocalcemia*.

##### **Occurrence**

Vitamin D-deficiency rickets used to be an extremely common disease in past centuries, but has now largely disappeared in the developed world. It occasionally occurs in small-for-gestational-age babies. An increase in the number of cases has recently arisen as a result of the use of certain milk substitute products and inadequate exposure to sunlight [101]. In developing countries, on the other hand, vitamin D-deficiency rickets is still a common disorder. Vitamin D-resistant rickets, on the other hand, is the commonest metabolic bone disease in developed nations, although precise figures are not available. The condition is hereditary and inherited as an X-linked dominant disorder in two-thirds of cases. It occurs twice as often in girls as in boys.

##### **Clinical features, diagnosis**

— **Vitamin D-deficiency rickets:** The affected children show muscle weakness and a general lack of drive. The abdomen is typically bulging, and the children start sitting, standing and walking at a later stage than normal. The bones in the area of the malleoli, knees and wrists are thickened. The skull bones are soft. If the infant usually lies on its back, the back of the head is mechanically flattened and the forehead, together with the base of the skull, is widened. Another typical features is bulging of the bony/cartilaginous attachments of the ribs (rachitic rosary). The lower limbs become

bowed as a result of weight bearing. Depending on the initial findings at the onset of walking, the bones can either show a valgus (more rare) or varus (more common) deformity. Coxae varae can also form, possibly followed by the development of scoliosis.

On the x-ray the epiphyseal plates appear thickened and ill-defined, while the epiphyses are widened with a ragged border with the metaphyses. The cortical bone in the diaphyses usually shows decreased radiodensity. Depending on the stage of the illness, bowing may also be visible (■ Fig. 4.78).

- **Vitamin D-resistant rickets:** The signs and symptoms are very similar to those of vitamin D-deficiency rickets, but generally more pronounced and not rectifiable by the administration of vitamin D. The condition is usually diagnosed at around the age of 2 years, but severe forms can manifest themselves after just a few months of life. The laboratory tests show hypophosphatemia and an elevated alkaline phosphatase level. The serum calcium is normal or only slightly reduced. The other electrolytes and the pH are usually within normal limits.
- **Differential diagnosis:** Cases of very marked genua vara after the onset of walking are usually *idiopathic*, but can also occur in connection with *Blount disease*. The possibility of *renal osteodystrophy* should also be considered in the differential diagnosis.

#### Treatment

- Treatment of the *underlying condition*:
  - *Vitamin D-deficiency rickets* can be prevented or corrected by the daily administration of 500 IU of



■ Fig. 4.78. AP x-rays of both knees of a 6-year old girl with vitamin-D-resistant rickets. Note the widened epiphyseal plates and the beaker-shaped metaphyses

vitamin D. Since breast milk and cow's milk are relatively low in vitamin D, vitamin D substitution is required for infants. The administration of excessively high doses should be avoided since vitamin D hypervitaminosis is dangerous. Adequate exposure to sunlight should also be ensured in addition to the sufficient vitamin intake.

- *Vitamin D-resistant rickets* must initially be treated by a pediatrician specialized in metabolic disorders so that the nature of the defect can be established. Treatment involves very high doses of vitamin D (between 50,000 and 100,000 IU). Phosphate must also be replaced depending on the serum concentration in each case.
- **Orthopaedic treatment:** We consider that the once common treatment with *splints or cast fixation* is *not appropriate*. Cast fixation results in osteoporosis in addition to the osteomalacia, thus further promoting the bone brittleness and bowing. *Splints* have also failed to prove effective. Children with rickets lack drive and start to walk at a late stage. The extra burden of splints will make them even less inclined to move, which is counterproductive and also liable to promote osteoporosis. Moreover, splints are not even capable of neutralizing the forces involved. A lower leg splint on its own can never correct a pronounced genu varum or valgum, while upper leg orthoses, which severely hamper walking, do too little to counteract the forces. No specific treatment is required for a patient with vitamin D-deficiency rickets with genua valga or vara provided the axial deviation is less than 15°. Vitamin D replacement will correct the osteomalacia in a relatively short time, and the axial deviation will normalize itself spontaneously. If the axial deviation is greater than 15°, a *corrective osteotomy* should be considered, since the displacement of the force resultants limits the possible spontaneous correction. If the pressure on the epiphyseal plates is excessive on one side, they react with bone resorption instead of bone formation. This particularly applies in vitamin D-resistant rickets. The correction should be made at the site of the deformity, usually in the lower legs, although the thighs may also be bowed. Bowing can also affect the femur and tibia. The most important goal is to achieve a horizontal knee. If both the femur and tibia are bowed, then both bones will need to be corrected, ideally at supracondylar level in the femur and at infracondylar level in the tibia, i.e. below the tibial tuberosity. The fibula must always be osteotomied at the same time. In the case of small children, we always perform the osteotomies without wedge removal, preferring to place the bone in the desired, straightened position and fix it with two crossed Kirschner wires. This is followed by the fitting of a long-leg cast for four weeks. After two weeks we place

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the children on a standing frame in order to counteract the development of osteoporosis. After four weeks, the cast and transcutaneously inserted Kirschner wires are removed. An external fixator can be used for older children [41]. If bowing affects the proximal femur, the new telescopic Gamma nail can be used (■ Fig. 4.87).

#### Renal osteodystrophy

Renal osteodystrophy occurs in chronic renal insufficiency and is very rare in children and adolescents. The most common causes of renal failure in children are chronic pyelonephritis with polycystic kidney, congenital renal hypoplasia or renal aplasia, chronic glomerulonephritis or cystine storage in the end stage of vitamin D-resistant rickets.

- Various factors play a role in the *development* of the disease. The renal insufficiency leads to secondary hyperparathyroidism with high serum concentrations of parathyroid hormone. The increased secretion of the parathyroid hormone is probably caused by hypocalcemia, although acidosis is also probably involved.
- *Clinically* the patients show the signs and symptoms of renal insufficiency, with polyuria, albuminuria, nitrogen retention and metabolic acidosis. The children are of small stature, occasionally with dwarfism. The bones of the legs are more affected than those in the arms and painful bowing with knock knees or bow legs are also often present [85].
- The *diagnosis* should always be confirmed with an iliac crest biopsy. The *x-ray* shows generalized osteoporosis with thinning of the cortices and bony trabeculae. The epiphyseal plates are widened, and epiphyseal separations are common. The skeletal maturity is delayed.
- The principal *differential diagnosis* to consider is *vitamin D-resistant rickets*.
- Treatment of the underlying disease: The prognosis for patients with renal osteodystrophy was considerably improved with the introduction of dialysis and kidney transplants. The conservative treatment usually involves the administration of vitamin D in very high doses (up to 200,000 IU).  
*Orthopaedic treatment:* As with rickets, *splint treatments* and *cast fixation* should be *avoided*. On the other hand it is important to ensure that the children's ability to walk is preserved for as long as possible. Deformities should be *corrected surgically* however as soon as they exceed a certain level. The increased perioperative risks should be taken into account (anemia, hypertension, bleeding tendency, disrupted electrolyte balance). Pinning is indicated for epiphyseal separations.

#### Hyper-/Hypoparathyroidism

##### Primary hyperparathyroidism

- This condition involves primary diffuse hyperplasia or neoplasia of the parathyroid glands and is ex-

tremely rare in children. Familial forms exist [34]. The increased secretion of parathyroid hormone produces elevated serum calcium levels accompanied by decreased serum phosphorus and raised alkaline phosphatase levels. Another effect of the parathyroid hormone is to increase the activity of the osteoclasts, resulting in the formation of holes in the bone (or *pseudotumors*). The holes are filled with fibrous tissue (hence the alternative name of the disease of osteitis fibrosa [64]; ■ Fig. 4.79).

- *Clinically*, gastrointestinal symptoms such as constipation and abdominal discomfort are observed. The mental state may also change, with the onset of lethargy. In addition to generalized osteoporosis, the *x-ray* shows stippled zones of resorption. On *histological examination* these zones are filled with fibrous connective tissue, enriched with giant cells, inflammatory cells, macrophages and hemosiderin. These so-called »brown tumors« are very typical of hyperparathyroidism.
- The *treatment* focuses primarily on the underlying disease, i.e. the removal of the hyperplastic or neoplastic parathyroid tissue.
- In the *differential diagnosis* it is important not to confuse the pseudotumors with genuine tumors.



■ Fig. 4.79. Soft tissue calcification in *pseudohyperparathyroidism* in the area of the proximal phalanx of the middle finger

### Idiopathic hypoparathyroidism

This disease is caused by underfunctioning of the parathyroid gland and occasionally occurs as a familial X-linked recessive inherited condition. The principal *signs and symptoms* are tetany, laryngism, exhaustion, mental retardation, dry skin, brittle nails, premature tooth loss and cataracts. *Laboratory tests* reveal a decreased serum calcium content and an elevated serum phosphorus level. *Radiologically* the skeleton is usually normal, although with increased soft tissue calcification. The most important *differential diagnosis* is the (more common) pseudohypoparathyroidism. Treatment involves the administration of vitamin D and parathyroid hormone, and calcium infusions are administered to correct any tetany.

### Pseudohypoparathyroidism

In clinical and radiological respects, this disease resembles hypoparathyroidism. However, it does not primarily involve the parathyroid gland, but rather an inadequate response of the end organs to the parathyroid hormone. This can be caused either by the abnormal tubular absorption of phosphate or the excessive production of thyrocalcitonin. The condition is inherited and there are *three forms*: An X-linked dominant form (female:male = 2:1), an autosomal-dominant form and an autosomal-recessive form. The disease is occasionally associated with hypothyroidism, Turner syndrome and diabetes mellitus. Typical skeletal abnormalities are: stunted growth, short and crude hands with brachymetacarpia and cone epiphyses. Soft tissue calcification are occasionally observed. The treatment consists of the administration of high doses of parathyroid hormone.

#### 4.6.2.26 Increased bone density without modification of shape

##### Melorheostosis

Melorheostosis is a very rare disorder with usually *unilateral, hyperostotic changes in the bone that resemble dripping candle wax*. The name is derived from the Greek words *melos* = limb, extremity, and *rhein* = flow.

- The prevalence is less than 1/million [115].
- No genetic pattern has been detected. Since melorheostosis has also been observed in association with osteopoikilosis, it is thought that changes in the same gene are responsible for both disorders [79].
- The bone changes are limited to dermatomes. The long bones are usually affected, while the skull, spine and ribs are rarely altered. The disease can also occur in a monostotic form. *Histological examination* reveals osteosclerosis and osteofibrosis. Fibrosing of the skin and subcutaneous tissue is also common, while the muscles show edematous changes. From the *clinical* standpoint, pain is often present in the affected limb. Joint contractures can occur, principally in the hip and

knee. Shortening and thickening of the palmar or plantar fascia is occasionally observed. The *x-ray* shows the typical »dripping candle wax« appearance (■ Fig. 4.80).

- The main conditions to consider in the *differential diagnosis* are *osteomyelitis*, *osteopetrosis* and *osteopoikilosis*. In view of the joint contractures, *arthrogryposis* must be ruled out. Melorheostosis can also be confused with *scleroderma*.
- The treatment is restricted to the *correction of contractures* using casts or splints. If very pronounced contractures have become established, osteotomies and tendon lengthenings can be used to resolve the problem. Since the soft tissue situation often proves problematic, the use of the Ilizarov apparatus is not very appropriate, and amputation is even required occasionally.
- Osteopoikilosis involves the formation of *oval, plump, radiopaque spots in the cancellous bone*. These may be present even at birth or only become visible later on. The changes usually affect the *metaphyses* and *epiphyses*, and rarely the *diaphyses* (■ Fig. 4.81).
- The *sites* most commonly affected are the carpal and tarsal bones, although the long bones and pelvis can also be involved. The skull and spine are rarely affected.
- This *autosomal-dominant inherited disorder* has a prevalence of less than 0.1/million and boys are slightly more frequently affected than girls [5].
- *Histologically* the nodules consist of laminar bone. The bone changes generally produce no clinical signs or symptoms, although *skin lesions* occasionally occur (*dermatofibrosis lenticularis disseminata*). *Bone tumors* have been observed in isolated cases [30].

### Osteopetrosis

#### ► Definition

Osteopetrosis is a metabolic disorder characterized by a systemic increase in skeletal mass. It results from a failure of the osteoclasts [51]. The primitive cartilaginous bone persists, and the abnormal bone density is apparent on x-rays.

- *Synonyms*: Marble bone disease, Albers-Schönberg disease

#### Historical background, classification, etiology, occurrence

The condition was first described in 1904, shortly after the discovery of x-rays, by Heinrich Albers-Schönberg [1]. Nowadays, a variety of forms are distinguished [51] (■ Table 4.24).

- The *etiology* involves a failure on the part of the osteoclasts [51]. Osteopetrosis also occurs in animals (mice, cattle, poultry). Differing enzyme defects prevail depending on the type of osteopetrosis involved



## 4.6.2 · Skeletal dysplasias



■ **Fig. 4.80.** X-ray of the right lower leg of a 13-year old boy with *melorheostosis*. The »dripping candle-wax« appearance is typical. The disease also involves the epiphyses. a) AP x-ray of the knee b) AP view of the ankle joint. c) Lateral view of the hindfoot. The hindfoot is in a fixed calcaneus position. d) AP view of the midfoot.

[51]. Osteomalacia is also usually present, i.e. the inability to maintain a normal calcium/phosphorus balance [42]. This leads to the paradoxical situation in which increased bone mass and bone softening are present at the same time, which explains the increased susceptibility to fractures.

- One epidemiological study in Great Britain calculated a prevalence of 2.6/million for all forms of the disease [115]. Another investigation found a prevalence of 50/million inhabitants, but the great majority of these cases involved the mild autosomal-dominant form [15].



■ **Fig. 4.81.** AP x-ray of the left hand of a 15-year old girl with *osteopoi-kilosis*. The oval, plump, radiopaque spots are characteristic

■ **Table 4.24.** The various types of osteoporosis. (After [51])

Type	Age of manifestation	Inheritance	Gene locus	Signs and symptoms
Infantile malignant	1st year of life	Autosomal-recessive	11q13.4-q13.5	Small stature, fractures, hypocalcemia, cranial nerve compression, severely restricted life expectancy
Intermediate form	1st–2nd year of life	Autosomal-recessive	16p13	Small stature, fractures, hypocalcemia, cranial nerve compression
Intermediate form with tubular acidosis	1st–2nd year of life	Autosomal-recessive	8q22	Small stature, fractures, hypocalcemia, cranial nerve compression, extramedullary hemopoiesis, mental retardation, renal tubular acidosis
Pyknodysostosis	1st–3rd year of life	Autosomal-recessive	1q21	Small stature, fractures, hypoplasia of the phalanges, clavicular dysplasia, hypodontia
Benign form (Albers-Schönberg)	10th–40th year of life	Autosomal-dominant	?	Fractures, sandwich vertebrae, scoliosis, coxarthrosis, osteomyelitis



### Clinical features, diagnosis

- The following signs and symptoms are present in the *malignant form* [50]: In over 50 percent of cases optic nerve atrophy, splenomegaly and hepatomegaly are observed. In less than half of cases, delayed growth, fractures, deafness, osteomyelitis of the jaw, genu valgum or varum and chest wall deformities are present.
- In the *late-onset form* half of the patients remain asymptomatic [50]. Around 40% suffer from spontaneous fractures, while osteomyelitis of the jaw has been observed in 10% of cases, spontaneous bone pain in 20% and deficits of individual cranial nerves in 22% of patients.

❗ **The following problems are of relevance to the orthopaedist: bone pain, spontaneous fractures with poor healing, coxa vara, possibly genua vara or valga, slightly stunted growth, spondylolysis, possibly scoliosis and kyphosis, rarely spinal stenosis, osteomyelitis (spontaneously or after surgical treatments) and osteoarthritis (osteoarthritis of the hip or knee).**

- Particularly striking *radiological* signs include the greatly increased bone density and medullary obliteration. The metaphyses of the long bones frequently appear coarsened, with closely-packed transverse bands, while longitudinal striae can be seen at the diaphyses (■ Fig. 4.82). Bands of increased bone density also arise in the vertebral bodies. The increased density and resulting lack of air spaces are also typically seen in the roof and base of the skull.
- While the *laboratory findings* are usually normal, an abnormal calcium/phosphorus balance may be present [42]. The fractures show poor healing with abnormal callus formation. Where possible, fractures should be treated conservatively [15].
- *Differential diagnosis*: Increased bone density is also observed in the following diseases: *pyknodysostosis* (presumed to be the condition suffered by Henri de Toulouse-Lautrec), *melorheostosis*, *sclerosteosis*, *pro-*



■ Fig. 4.82. AP x-ray of the pelvis of a 20-year old female patient with *osteopetrosis*, i.e. greatly increased bone density

*gressive diaphyseal dysplasia (Engelmann disease)* and *metaphyseal dysplasia (Pyle's disease)*.

### Prognosis, treatment

- *Prognosis*: Patients with the *congenital malignant form* rarely reach adulthood, whereas those with the *late-onset form* have a normal life expectancy. Around half of the cases progress asymptotically, in which case the condition is diagnosed as a chance finding. The main problem in advanced age are the cases of premature osteoarthritis.
- *Treatment of the underlying disease*: Partial success has been obtained in severe cases with the administration of prednisone, calcitriol, parathyroid hormone and interferon [48]. The transplantation of allogeneic hematopoietic stem cells seems to be a promising approach [107].
- *Orthopaedic treatment*: In young patients, the treatment of fractures is the prime concern. Since the bones heal very poorly and cases of postoperative osteomyelitis are common, the fractures should be treated conservatively if possible. One effective therapeutic method is *medullary nailing*, although the surgeon must also ensure the greatest possible stability. *Osteotomies* may be needed for the correction of severe deformities. The principal orthopaedic problem, however, is the early onset of osteoarthritis of the hip and knee, which must be managed with corresponding *total prosthetic replacements*.

#### 4.6.2.27 Increased bone density with diaphyseal involvement

##### *Infantile cortical hyperostosis (Caffey disease)*

- This is a very rare, self-limiting condition of early childhood involving *swelling of the soft tissues, thickening of the cortical bone and local cutaneous hypersensitivity*.
- *Etiology*: The mode of inheritance can be autosomal-recessive (type I) or autosomal-dominant (type II). The gene locus is 1q41-q42. The enzyme defect is not yet known. A congenital defect in the arterioles of the periosteum may be involved.
- *Histopathological examination* initially reveals changes in the periosteum followed, by way of reaction, by hyperostosis. The jaw is most commonly affected, followed by the ulna. The tibia and clavicles are less frequently affected, while there are only isolated reports of other bones being affected by this condition.
- *Clinical features*: The condition generally starts around the third month of life, and an onset later than the fifth month is very rare. Initially, there is hypersensitivity of the skin over the jaw. The skin is very rough but not inflamed. A slight fever may also be present, and the alkaline phosphatase level may be slightly elevated.
- The *x-rays* show pronounced thickening of the cortical bone as a result of periosteal deposition. The con-

#### 4.6.2 · Skeletal dysplasias

dition lasts for several months, possibly extending to a few years in some cases.

- The main conditions to consider in the *differential diagnosis* are *osteomyelitis*, *hypervitaminosis A* and tumors such as *Ewing sarcoma*. But since the skin changes and the patient's age usually allow an unambiguous diagnosis to be made, biopsy is not necessary.
- *Treatment* involves the administration of *corticosteroids* to accelerate healing somewhat. Orthopaedic measures are not necessary, although the orthopaedist must be aware of the condition given the impressive radiological changes.
- Other genetically differentiated diseases in this group include Camurati-Engelmann diaphyseal dysplasia, craniodiaphyseal dysplasia, Lenz-Majewski dysplasia and osteoectasia with hyperphosphatemia (juvenile Paget disease). These conditions are all extremely rare.

#### 4.6.2.28 Increased bone density with metaphyseal involvement

##### *Metaphyseal dysplasia (Pyle disease)*

Metaphyseal dysplasia, also known as Pyle's disease, is a rare hereditary disease in which the metaphyses of the long bones are deformed. The deformation has been compared to an »Erlenmeyer flask«. In addition to the characteristic bulging of the metaphyses of the long bone (■ Fig. 4.83), thickening occurs at the medial ends of the clavicles, the pubis and the ischium. Sclerosis is observed at the base of the skull. This autosomal-recessive disorder does not cause any major clinical problems apart from the occasional case of bowleg. Increased bone brittleness may also be present in the area of the metaphyses, which can lead to problems in differentiating this condition from child abuse. The *prognosis* for metaphyseal dysplasia is very good. Apart from the occasional osteotomy to correct a bowleg, orthopaedic treatments are rarely required.

#### 4.6.2.29 Craniotubular digital dysplasia

This group includes frontometaphyseal dysplasia, osteodysplasty according to Melnick-Needles and the otopalatodigital syndrome. The metaphyseal bone changes seen in fronto- or craniometaphyseal dysplasia resemble those in Pyle's disease (► Chapter 4.6.28) and rarely require corrective orthopaedic surgery. The dome of the skull is sometimes thickened so considerably as to produce cranial nerve disorders.

#### 4.6.2.30 Neonatal severe osteosclerotic dysplasia

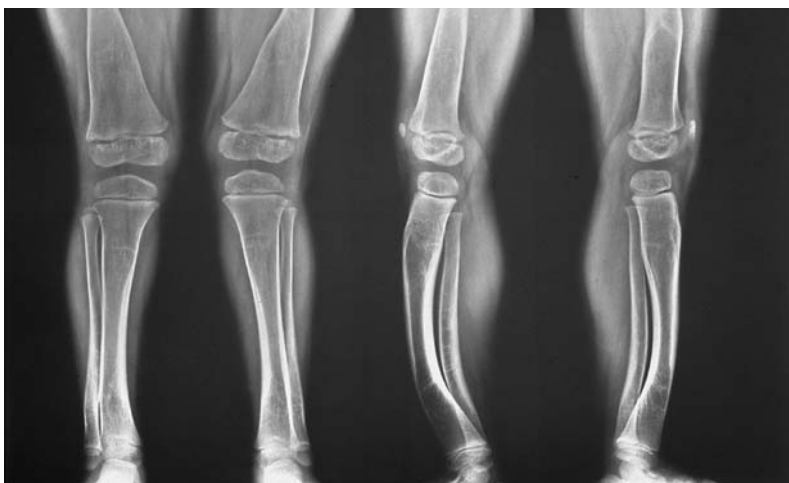
Since the syndromes belonging to this group (Blomstrand dysplasia, Raine dysplasia, prenatal form of cortical hyperostosis according to Caffey, Astley-Kendall dysplasia) are all lethal forms, orthopaedic treatments are superfluous.

#### 4.6.2.31 Disorganized development of cartilaginous and fibrous components of the skeleton

##### *Dysplasia epiphysealis hemimelica* (»Trevor's disease«)

Dysplasia epiphysealis hemimelica is characterized by abnormal osteocartilaginous exostoses in the epiphyses of the long bones and in the carpal or tarsal bones of the medial or lateral half of an extremity. The disease was first described in 1926 by Mouchet and B elot [75] under the name of tarsomegaly. However the name »Trevor's disease« has gained widespread acceptance in English-speaking countries following a case report by this author in 1950 [113]. The name of dysplasia epiphysealis hemimelica originates from Fairbank (1956) [24]. Just over 100 cases have been described in the literature to date.

The mode of inheritance is not known. In histological respects the changes in this condition are indistinguishable from osteochondromas (see below). Since the latter never occur in the epiphyses, but always in the metaphy-



■ Fig. 4.83. AP and lateral x-rays of both legs of a 4-year old boy with *metaphyseal dysplasia* with characteristic bowing of the bones around the metaphyses

ses, the presence of a corresponding epiphyseal change very probably indicates the presence of a dysplasia epiphysealis hemimelica. The disease occurs in association with enchondromatosis (see below this chapter) [82].

The diagnosis is usually made in early childhood. The most common sites are the tarsal bones, the distal femoral epiphysis and the proximal tibial epiphysis (■ Fig. 3.306, ► Chapter 3.3.6). The changes lead to joint incongruity and deformity with genu valgum or varum. Pain is not a primary concern.

In terms of treatment very disruptive lesions should be surgically removed. However, since these rapidly regrow repeated excisions may be required. The specific treatment indicated must be decided with care, since articular cartilage is inevitably damaged as a consequence of any operative procedure. Axial corrections are occasionally indicated. The situation usually consolidates on completion of growth, although continued growth during adulthood has occasionally been observed. Serious incongruity involves the risk of premature osteoarthritis.

### **Multiple osteochondromas (also known as cartilaginous exostoses)**

#### ► Definition

Autosomal-dominant hereditary disease with the occurrence of multiple osteochondromas in the area of the metaphyses of the long bones with moderately stunted growth, and frequently local growth disturbances as well (particularly on the forearm) and a tendency to affect males. The commonly used term of »multiple cartilaginous exostoses« is not correct since the osteochondromas are formed from hyaline cartilage and do not constitute »cartilaginous« exostoses covered with fibrous cartilage (► Chapter 4.5.2).

#### **Etiology, pathogenesis, occurrence**

This is an autosomal-dominant condition involving a defect on one of three different chromosomes (8q23-q24.1, 11p12-p11, 19p). The condition is more prevalent in males than in females. The histopathological picture corresponds to that seen in a solitary osteochondroma. The cartilaginous cap is usually relatively thick, the thickness depending on the age of the patient: the younger the patient the thicker the cartilaginous cap. The osteochondromas originate from incorrectly differentiated cartilaginous tissue from the growth plate. As growth continues, the incorrectly differentiated tissue remains at subperiosteal level, where it starts to proliferate at right angles to the original orientation of the growth plate. Multiple hereditary osteochondromas are rare. One British study reported a prevalence of 0.9/million inhabitants [115], while another study calculated an incidence of 2/100,000 [106]. As they appear only in later childhood and often do not need treatment, their prevalence is probably mark-

edly underestimated. In my experience the syndrome of multiple osteochondromas is one of the commonest hereditary skeletal dysplasias, occurring more frequently than achondroplasia or osteogenesis imperfecta.

#### **Clinical features, diagnosis**

Firm, well-defined bony bulges appear in various parts of the body, primarily in the vicinity of the knee and shoulders, around the age of two. In principle, the osteochondromas can occur on any bone with cartilage, but not on membranous bone [44]. The extremities are always more severely affected than the trunk, and osteochondromas tend to form particularly at those sites with rapidly growing metaphyses. The exostoses are *always* located at the metaphyses, never at the epiphyses. Deformities and joint problems occur secondarily. Typical deformities are a radial head dislocation (approx. 20%), ulnar deviation of the hand (approx. 30%), tibia valga (approx. 30%) and pes valgus (approx. 50%) [106].

The forearm and lower leg deformities are especially striking (■ Fig. 3.449, ► Chapter 3.4.13). Those on the *forearm* are attributable to the differing growth rates of the radius and ulna [44]. The deformities associated with osteochondromas on the forearm are discussed in ► Chapter 3.5.7. The most common deformities on the *legs* are tibia valga and pes valgus (■ Fig. 4.84).



■ Fig. 4.84. X-rays of both legs of an 8-year old girl with *multiple osteochondromas* on all the metaphyses of the long bones in the legs

#### 4.6.2 · Skeletal dysplasias

These deformities result from an excessively short fibula, which forces the tibia to grow abnormally via the clamping effect of the interosseous membrane (■ Fig. 3.449, ► Chapter 3.4.13) [78]. The deformities are at the metaphyseal level, while the growth plate remains horizontal. The change in the spatial orientation of the lateral tibial surface at the ankle facilitates the subluxation of the talus. The pes valgus results from the elevated position of this lateral surface. Leg length discrepancies are also not infrequent. Moderately stunted growth is present.

- **Radiographic findings:** The exostoses can have a wide or narrow base with short or long stalks. The morphology is highly varied. The exostosis is always oriented towards the diaphysis. As growth proceeds the base of the exostosis migrates towards the center of the shaft. The metaphyses are widened with an abnormal bottle- or flask-like shape. The exostosis is always covered by a cartilaginous cap of varying thickness. The thickness of the cartilage can be established by means of an ultrasound or MRI scan [45]. The extent of this cartilaginous covering is important for the prognosis in terms of malignant degeneration. As with all cartilaginous tumors, calcifications also often occur. The extent of the calcifications has no prognostic significance. Whether new enchondral bone formation is taking place or not can be determined by means of a bone scan.
- The most important condition to consider in the *differential diagnosis* is *metachondromatosis* (see below). This also affects the flat bones and the spine and involves characteristic changes of the femoral head (see below). The exostoses are oriented towards the epiphysis. If the exostoses are located in the epiphysis itself, the possibility of *dysplasia hemimelica epiphysealis* should be considered.

#### Prognosis

The probability of *malignant change* is of particular prognostic significance in hereditary multiple osteochondromas. The figures in the literature vary widely, ranging from 1% [88, 106] to 10–20% [7]. It must be assumed that negative selection was involved in those patient samples with high percentages. The actual rate of malignant change is probably less than 2% [88]. Special risk factors for malignant transformation are large exostoses close to the trunk, exposed to mechanical irritation and with a particularly thick cartilaginous cap.

The osteochondromas usually transform into a chondrosarcoma, although osteosarcomas and fibrosarcomas also occur [45]. The transformation generally occurs only above the age of 20. Since it usually occurs at the base of the tumor it must be resected well into the bone if the risk of degeneration is to be minimized. Much greater than the risk of degeneration is the risk of osteoarthritis as a result of deformities close to joints [81].

#### Treatment

The surgeon must be very cautious in deciding whether a surgical intervention is indicated. Excision is only indicated if the cartilaginous exostosis is painful, interferes with joint or muscle function or leads to nerve deficits or joint deformities. It is important to be aware of the fact that recurrences are possible during growth, and can only be ruled out after the child has stopped growing. Details about the specific treatment for osteochondromas on the forearm are provided in ► Chapter 3.5.7. The tibia valga and valgus deformity of the ankle must be corrected by an osteotomy (► Chapter 4.2.1).

#### Metachondromatosis

In this disease the multiple osteochondromas occur on the fingers and long bones. In contrast with classical multiple osteochondromatosis they are oriented towards the epiphyses and can regress spontaneously.

This very rare autosomal-dominant condition was described by Maroteaux in 1971 [66]. The metaphyseal changes in the area of the femoral neck are particularly striking, with flattening of the femoral head and cases of femoral head necrosis. The changes in metachondromatosis (■ Fig. 4.85) also occur in flat bones. Since the osteochondromas in metachondromatosis do not cause shortening, growth is not stunted. Since axial deviations do not occur, the prognosis is favorable and resection is



■ **Fig. 4.85.** X-ray of the left hand of an 8-year old boy with *metachondromatosis*. In contrast with multiple osteochondrosis, the metaphyseal osteochondromas are oriented towards the epiphyses rather than the diaphyses



rarely required, particularly as the osteochondromas can regress.

### Multiple enchondromatosis (Ollier syndrome)

#### ➤ Definition

Disease with, generally unilateral, multiple enchondromas, usually in the long bones, pelvis and, less commonly, the spine. The facial skull and cranium are hardly ever affected. The disease does not appear to be hereditary.

— **Synonym:** Dyschondroplasia

#### Historical background, Etiology, pathogenesis, occurrence

This autosomal-dominant condition was described in 1899 by M. Ollier [83]. The gene locus is 3p22-p21.1. The enchondromatosis involves a hamartomatous proliferation of chondrocytes derived both from the bone itself and the periosteum. Histological investigations have shown that aberrant cartilaginous tissue persists in the metaphyses of the long bones, where it starts to proliferate. Cases occur sporadically and are rare, although fairly large series with approx. 20–50 patients in each have been described. There is no difference between the sexes, and the condition does not affect one side more than the other.

#### Clinical features, diagnosis

The patients often have a striking appearance even in early childhood as a result of the bowing and shortening of the bones. The most common deformity is the varus deviation of the distal femur. The x-rays then show multiple, irregularly defined enchondromas, primarily in the metaphyses, but also in the diaphyses in severe cases. Any bone can be affected (■ Fig. 4.86), although the skull is rarely involved. The lesions are strictly unilateral. To date, only sporadic cases with generalized bilateral enchondromatosis have been described [72]. The most important clinical problems are the progressive shortening of the bowed extremity and, occasionally, pathological fractures. Joint deformities occur as a result of the bowing (■ Fig. 4.86).

#### Prognosis, treatment

The most important prognostic factor is malignant degeneration, for which the risk appears to be much greater than for multiple osteochondromas.

When degeneration occurs, the enchondromas almost always transform into a chondrosarcoma, although osteosarcomas and dedifferentiated chondrosarcomas can also occur. A risk of approx. 30% has been calculated in the literature for the degeneration rate [28]. This particularly applies to large enchondromas and those close to the trunk.



■ Fig. 4.86a–d. X-rays of the pelvis (a), left forearm (b) and left upper

(c) and lower leg (d) of an 8-year old boy with *enchondromatosis* (Ollier disease). The arrows indicate the multiple enchondromas, which are all located in the left half of the body



Treatment mainly involves the management of the fractures, correction of the axial deformities and leg lengthening procedures (► Chapter 4.2.1 and 4.2.2). The treatment is often difficult as the enchondromas weaken the bone considerable and thus often pose a problem in terms of stability. Where deformities are treated surgically, care is also required in avoiding excessive contamination of the healthy tissue with tumor cells. In contrast with fibrous dysplasia, there is a probable risk of tumor propagation.

### Maffucci syndrome

The Maffucci syndrome is a condition with unilaterally occurring enchondromas (as in Ollier disease), combined with multiple hemangiomas or spindle-cell hemangioendotheliomas.

The disease was described by Maffucci in 1881 [62]. It occurs sporadically as a spontaneous mutation at the same gene locus (3p22-p21.1) as Ollier's syndrome. Approx. 250 cases have been described in total. Both sexes are equally affected. Whereas hemangiomas always used to be observed in the past, several cases involving spindle-cell hemangioendotheliomas have recently been reported [26]. The hemangiomas are already present at birth and occur principally at subcutaneous level, enabling a diagnosis to be made in infancy. A recent investigation has provided evidence to indicate the presence of numerous nerve fibers and the secretion of large quantities of mitogenic neurotransmitters in the vicinity of the hemangiomas. These neurotransmitters play a role in the pathogenesis of the disease [99]. The enchondromas in Maffucci syndrome, and thus the orthopaedic problems as well, are similar to those in Ollier disease, as is the risk of their malignant degeneration, for which a figure of 23% has been calculated. By contrast, the risk of degeneration of the hemangiomas or hemangioendotheliomas is very low.

### (Polyostotic) fibrous dysplasia, Albright syndrome

#### ► Definition

Disease with congenital intraosseous, hamartomatous changes occurring simultaneously in different bones. These lesions increase in size as the patient grows. The condition can affect a single or multiple bones and, very rarely, is associated with endocrine abnormalities such as precocious puberty, premature physeal closure and hyperthyroidism. When associated with hormonal disorders the condition is known as *Albright syndrome*.

- *Synonyms:* Osteofibrosis deformans juvenilis, osteodystrophia fibrosa, osteitis fibrosa disseminata, McCune-Albright syndrome, Jaffé-Lichtenstein disease

#### Historical background

Polyostotic fibrous dysplasia was first described by Lichtenstein in 1938 [57]. Subsequently, Lichtenstein and Jaffé [58] merged the polyostotic and monostotic forms under

the umbrella term of »fibrous dysplasia«. The Albright syndrome was described by McCune in 1936 [70] and by Albright et al. in 1937 [2].

#### Etiology, pathogenesis

The disease appears to involve an abnormality of the osteogenic mesenchyme. The primitive fibrous tissue proliferates in the medulla of bone and also attacks the cortical bone from the center. Histologically the polyostotic and monostotic forms are identical (► Chapter 4.5.2). The polyostotic form usually affects one side only. The condition involves mosaic change in the genetic structure with differing degrees of penetrance. The gene locus is 20q13. The mutation is in the gene that codes for a membrane-bound signal protein (GS- $\alpha$ ). The manifestation of the clinical picture (McCune-Albright syndrome, polyostotic or monostotic fibrous dysplasia) depends on the time at which the mutation occurs. The disease cannot be inherited, because if all cells are affected by the genetic change the embryo does not survive.

#### Classification, occurrence

The following are distinguished:

- monostotic form (► Chapter 4.5.3),
- polyostotic form,
- McCune-Albright syndrome (polyostotic fibrous dysplasia, pigmentations of the skin, hormonal disorders with precocious puberty).

While the various forms of fibrous dysplasia are rare, the *monostotic type* in particular often involves no clinical signs and symptoms. As a result, a relatively large number of cases remain unreported. A prevalence of 2.6/million inhabitants in Great Britain has been calculated for the *polyostotic form* [115]. The *monostotic form* is twice as common as the polyostotic form. Females are slightly more frequently affected than males. The *McCune-Albright syndrome* is extremely rare (less than 5% of all cases of fibrous dysplasia).

#### Clinical features, diagnosis

The congenital developmental disorder progresses slowly as the child grows and normally comes to a halt on completion of growth. The condition is usually diagnosed during the first decade, although often not until the second decade. The fibrous changes can occur in any bone, but most commonly affect the proximal metaphysis of the femur and the tibia in the vicinity of the knee (approx. 50% of all cases) [22]. Several lesions can also occur simultaneously in one bone. The condition progresses asymptotically unless a pathological fracture occurs or bowing is outwardly visible (■ Fig. 3.111, ► Chapter 3.1.11). Bowing particularly affects the proximal femur, where the soft bone can bend into the shape of a »shepherd's crook« (■ Fig. 3.245). Clinically relevant leg length discrepancies can also occur.

The *radiological changes* are characteristic. There are osteolytic areas, the cortex is thin and bulges out, usually the whole bone is widened and the basic structure shows a ground glass opacity in the osteolytic zones. This ground glass pattern is attributable to the formation of new bone. Osteolytic and sclerotic components appear next to each other. The cortical bone is eroded and the bone is widened as a result of new periosteal bone formation. Microfractures also occur and can lead to painful episodes. The bone scan shows moderate uptake particularly in the active zones. On the MRI scan, the fibrous tissue is relatively signal-rich in both the T1- and T2-weighted images, indicating the presence of a high fluid content.

In the *McCune-Albright syndrome* the *polyostotic fibrous dysplasia* is accompanied by abnormal *skin pigmentations* that resemble the *café-au-lait spots* in neurofibromatosis. Girls experience a *precocious puberty*, resulting in a *small stature* as a result of the premature epiphyseal closure. Other hormonal disorders such as hyperthyroidism can occur, and cortisone metabolism may also be disrupted.

#### Differential diagnosis

Individual foci are not always easy to differentiate radiologically from *solitary bone cysts*, since the latter also show swelling of the bone with an osteolytic lesion and intervening bone trabeculae. However, the ground glass opacity is lacking. Note that both lesions can show a strong signal on the MRI scan because of the fluid content.

Another important differential diagnosis to consider is *osteofibrous dysplasia according to Campanacci*. This condition occurs almost exclusively on the tibia and is described in ► Chapter 4.5.3. If a *polyostotic form* of fibrous dysplasia is present, however, there is little possibility of confusion. *Enchondromatosis* and *histiocytosis*, which also affect multiple bones, usually differ markedly in their appearance on the x-ray. Enchondromas only produce minimal swelling of the bone and tend to form calcifications. While the appearance of histiocytosis varies considerably, it lacks both the bone swelling and ground glass opacity.

#### Prognosis

The prognosis for the disease is generally good and life expectancy is not reduced to any great extent. On the other hand, some cases of malignant degeneration have been described in the literature [47, 102]. Fibrous dysplasia can undergo cystic degeneration and thus break out of the bone, a finding that can be mistaken for malignant degeneration. In a large-scale study with over 1000 cases the risk of malignant change was calculated at 3% [102], i.e. much smaller than for enchondromatosis for example (up to 30%).

#### Treatment

No known treatment exists for the underlying condition. In severe cases, a certain amount of success has

been achieved with bisphosphonates (Aredia) [53]. The individual lesions are generally asymptomatic and do not require treatment. Only if symptoms, fractures or pronounced bowing occurs are therapeutic measures indicated. This particularly applies to the femur. A suitable solution for stabilization is an intramedullary load-bearing implant in the area of the femoral neck, or so-called »Y nail« or Gamma-nail (also known as a »Zickel nail«) fitted with an angularly stable blade or screw in the femoral neck. The blade must be adequately anchored in the femoral head. In children with open epiphyseal plates the new telescopic Gamma-nail can be used (■ Fig. 4.87, see also in osteogenesis imperfecta, ► Chapter 4.6.2.24). If doubt exists, the diagnosis must be confirmed by biopsy before such a radical measure is undertaken. Since fracture and osteotomy healing is not usually impaired, surgical measures are only rarely indicated. A biopsy is only indicated prior to surgical procedures. If the patients are asymptomatic, the current imaging procedures are sufficient for establishing the diagnosis.

#### *Fibrodysplasia ossificans progressiva*

##### ➤ Definition

This is an autosomal-dominant inherited disorder (gene locus 4q27–31) characterized by progressively increasing calcification and ossification of the fasciae, aponeuroses, tendons and ligaments and shortening of the great toes.

- The *etiology* involves the abnormal induction of enchondral osteogenesis in connective tissue [43]. The bone morphogenic protein content (BMP) of the cells is increased [84]. The calcifications – and subsequently ossifications – occur primarily in the interstitial connective tissue and in the tendons and ligaments, but not in the actual muscles. The use of the term »myositis« in this context is therefore misleading.
- *Occurrence*: The disease is very rare with only a few isolated cases in the literature. A total of 44 cases were described in a meta-analysis [13].
- *Clinical findings* include widely varying abnormalities of the great toes (usually shortening) at birth, and the thumbs may also be shortened. Since the ossifications only start appearing between the ages of 5 and 10, the diagnosis is not usually confirmed before this time. The ossifications generally appear initially in the neck and then gradually spread out over the whole body. The mobility of the joints is increasingly restricted secondarily as a result of the ossifications, until they eventually ankylose. As a rule, the calcifications and ossifications spread from cranial to caudal and central to peripheral directions [13]. An alopecia areata is also often observed.
- An *x-ray* of the cervical spine shows strikingly small vertebral bodies, whereas the pedicles are fairly large. Pathological fractures occasionally occur.

## 4.6.2 · Skeletal dysplasias

- In terms of *differential diagnosis* it is not always easy to distinguish this form of fibrodysplasia from an extraskeletal osteosarcoma. Confusion with a post-traumatic myositis ossificans is possible.
- *Prognosis*: The condition progresses even after the completion of growth and life expectancy is reduced, although the oldest known patient to date has reached the age of 70 [13].
- *Treatment*: The precise genetic and enzyme defects are not known and no specific treatment is currently available.

❗ **The calcifications and ossifications are exacerbated by trauma, particularly surgical trauma. The ossifications can increase even after a biopsy.**

- Since a reliable *diagnosis* can usually be made on the basis of the clinical findings, a biopsy is not necessary.
- *Surgical measures* will not produce any improvement in the calcifications and shortening. If an operation is required for other reasons, it should be performed under bisphosphonate protection and – in full-grown patients – postoperative radiotherapy in order to prevent subsequent ossifications. Surgery is indicated only in extremely restricted circumstances. The transplantation of allogeneic *hematopoietic stem cells* appears to offer the potential of influencing the disease positively [21].

**Cherubism**

In this condition, the mandible and thus the lower half of the face is deformed. The term cherub refers to an angel or celestial attendant. During the Renaissance the faces of such beings were painted in a particular style that is reminiscent of patients with cherubism. Orthopaedic problems are not a characteristic feature of this condition.

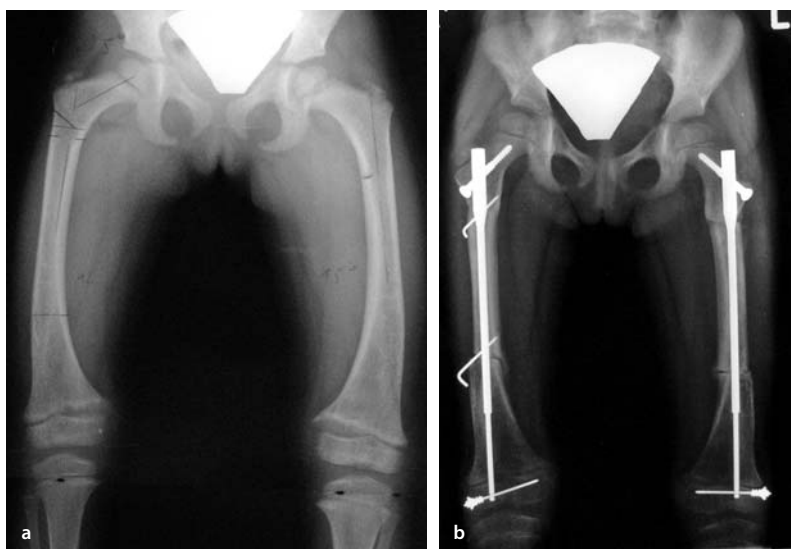
**4.6.2.32 Osteolyses**

Familial *multicentric carpal/tarsal osteolysis* is very rare and can be inherited as a dominant, recessive or sporadic disorder. It involves the progressive onset of osteolytic defects in the hands and feet. More generalized forms also exist, for example *Torg syndrome* or *familial expansile osteolysis*, in which other parts of the skeleton are affected. The *Gorham syndrome* (also known as »vanishing bone disease«) may also belong to this group, although it has not been clearly established whether this is a hereditary disorder. There is evidence to suggest that a local angiomatosis leads to bone loss [73]. Of the 200 or so cases described in the literature, most had achieved advanced adulthood by the time the disease manifested itself.

**4.6.2.33 Patella dysplasias*****Nail-patella syndrome (onycho-osteodysplasia)***

This condition was described as early as 1820 by Chatelain, who described changes *in the nails, together with abnormalities of the elbow and knees*.

- The prevalence has been cited as 1/million [115]. The gene locus for this autosomal-dominant condition is 9q34.1. The disorder affects the tissues of the ectoderm and mesoderm.
- *Clinical examination* reveals *dystrophy of the fingernails* (particularly toward the thumb side). The nails may be completely absent. At the same time the *patella is hypoplastic*, possibly with several ossification centers and a lateralized position. The latter can lead to lateral dislocation of the *patella*, particularly if the lateral femoral condyle is also hypoplastic. A *cubitus valgus* and posterior *subluxation of the radial head* may also be present.
- The *x-rays* of the pelvis show horn-like protrusions on the ilium.



❑ **Fig. 4.87.** AP x-rays of both femurs of a 4-year old girl with *vitamin-D-resistant rickets* and bilateral varus deformity of the femoral neck and shaft. **a.** Preoperative x-ray. **b.** X-ray after correction with subtrochanteric valgus osteotomies, diaphyseal osteotomies and stabilization with telescopic Gamma-nails, which have been inserted from the greater trochanter and locked with a screw in the distal epiphysis. The screw in the femoral neck ensures preservation of the valgus angle of the femoral neck.

- The prognosis is good.
- Orthopaedic problems are minimal, apart from the possible dislocation of the patella, which may require a recentring operation (► Chapter 3.3.5), and we have performed such a procedure in isolated cases. The surgeon should be cautious in deciding whether correction of the elbow deformity is indicated. A radial head resection can prove successful if pain or a troublesome bulge is present, but the range of motion of the elbow cannot be improved. Attempts to reduce the radial head are not successful, as the head does not fit into the joint.

This group of diseases also includes the *Meier-Gorlin syndrome* («ear patella short stature syndrome»). The ears and patellae are usually completely absent in this extremely rare autosomal-recessive inherited disorder.

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### 4.6.3 Dysostoses (localized hereditary skeletal deformities)

#### Definition

Dysostoses are congenital disorders in which individual regions of the body are affected rather than a whole tissue system. Dysostoses were classified according to the »International Nosology and Classification of Constitutional Disorders of Bone« in 2001 [5]. This classification is used here and, in contrast with that for skeletal dysplasias, uses letters to group the dysostoses (A, B, C).

#### 4.6.3.1 Dysostoses with predominant cranial and facial involvement (A)

This group includes a range of types of acrocephalosyndactyly with differing genetic characteristics. While the Pfeiffer, Crouzon, Jackson-Weiss and Seathre-Chatzen syndromes are extremely rare, the more common Apert syndrome is of greater orthopaedic relevance.

#### Apert syndrome

#### Definition

Congenital disorder with synostoses of the cranial sutures, syndactylies and synostoses on the hands and feet, contractures of various joints and frequently also ophthalmologic problems, segmentation defects of the cervical spine and sleep apnea.

— *Synonym: Acrocephalosyndactyly*

#### Historical background, etiology, pathogenesis, occurrence

This syndrome was first described in 1906 by E. Apert [1]. The mode of inheritance is autosomal-dominant, although most cases occur sporadically. The gene locus is 10q25-q26. It involves a defect in the mesenchymal tissue that results in a failure to isolate the ossification centers and consequent ossification even in non-osteogenic tissues. It is a rare condition, with a reported prevalence of 0.9/million [12]. A more recent report from California calculated an incidence of 1.24/100,000 live births [11].

#### Classification

The synostoses on hands (► Chapter 3.5.3) and feet (► Chapter 3.4.5) occur in varying degrees, and ■ Table 4.25 presents a suggested classification [2].

#### Clinical features, diagnosis

The most striking features in acrocephalosyndactyly are the shape of the head, the face and the synostoses on the hands and feet. The head is broad, as is the forehead, and the eyes are wide apart. The skull generally is slightly larger than normal. Ventriculomegaly is present in 76% of cases [10]. The hands and feet show bilateral,

**Table 4.25.** Classification of synostoses of the hands and feet in Apert syndrome

Severity	Synostoses in Apert syndrome
I	Synostosis between rays II–IV, rays I and V separate
II	Synostoses between rays II–V, only ray I separate
III	Synostoses between all rays



**Fig. 4.88.** AP x-ray of the hand of a 3-year old boy with *Apert syndrome* and several synostoses between the metacarpals and phalanges

usually symmetrical syndactylies with hyperextension of the great toes. There are intermetatarsal and intermetacarpal synostoses (■ Fig. 4.88), particularly between the 1st and 2nd rays. Multiple synostoses are present in the tarsal and carpal areas. In the more severe forms, the hands and feet form a single plate with almost no independent movement and on which the individual fingers and toes are also unable to move separately (■ Fig. 3.403, ► Chapter 3.4.5).

In addition to these outwardly striking features, movement is also often restricted at the *elbow* and *shoulder* [3, 6]. Shoulder mobility is never completely normal and the glenoid is dysplastic. Elbow mobility is also usually restricted to a greater or lesser extent. A certain stiffness is usually observed in the *hips* and *knees*, although the contractures in these joints are rarely severe enough to require treatment.

The *craniosynostoses* impair cranial growth and lead to increased intracranial pressure. This, in turn, leads to *psychomotor retardation* and problems with the ophthalmic nerve and muscles. Failure to provide treatment at

an early stage can result in blindness. There may also be obstruction of the upper *airways* with sleep apnea [8].

The x-rays of the *cervical spine* regularly show synostoses between the individual vertebral bodies. In two-thirds of cases, block vertebrae with restricted mobility of the cervical spine are observed [7].

#### Prognosis, treatment

Life expectancy is not significantly restricted in patients with Apert syndrome. *Treatment* should always be provided on a multidisciplinary basis. *Osteotomies* are performed at a relatively early stage on the *cranial* synostoses to prevent any impairment of brain growth.

The syndactylies on the *hands* should be separated at the age of 1–2 years in order to avoid any additional interference with the length growth of the fingers (► Chapter 3.5.4). Depending on the position of the fingers, osteotomies may be required, but amputations are obsolete [4]. Since the fingers are very rigid, deformities are poorly tolerated since an immobile projecting finger can prove very troublesome. The same applies to the feet. The hyperextension of the great toe can often lead to difficulties when putting on shoes. Osteotomies are often indicated here (► Chapter 3.4.5.11). Toes that deviate markedly towards the plantar side can hinder the heel-to-toe roll. An extending osteotomy can help alleviate this situation.

The management of children with acrocephalosyndactyly requires close collaboration between neurosurgeons, plastic surgeons, hand surgeons and orthopaedists. Surgical treatment is relatively rarely required for contractures that affect the major joints (shoulder, elbow, hip and knee), although regular physical therapy is indicated in order to improve mobility.

#### 4.6.3.2 Dysostoses with predominant axial involvement (B)

##### *Spondylocostal dysplasia (Jarcho-Levin syndrome)*

This is a hereditary condition with multiple deformities of the spine and synostoses of the ribs, usually on both sides. The mode of inheritance can be autosomal-recessive or autosomal-dominant, and the gene locus is 19q13 in both forms. Details of the clinical features and treatment are provided in ► Chapter 4.1.7. The use of the »vertical expandable prosthetic titanium ribs« (VEPTR) offers new possibilities for improving lung function.

#### 4.6.3.3 Dysostoses with predominant involvement of the extremities (C)

This category includes the various groups of the Fanconi syndrome (with renally related osteomalacia), the Coffin-Siris syndrome (brachydactyly, abnormalities of the nails, clinodactyly, facial abnormalities), symphalangism and multiple synostoses. Aplasia of the radius and thumb

deformities are typical in Fanconi syndrome. The Coffin-Siris syndrome is characterized by an absent nail and terminal phalanx of the fifth finger. These syndromes are all either extremely rare or are of little orthopaedic relevance.

### Rubinstein-Taybi syndrome

This autosomal-dominant symptom complex (gene loci 22q13, 16p13.3) is characterized by severe mental retardation combined with typical changes in the thumbs and great toes. The thumb is deviated toward the radius («hitchhiker thumb»; Fig. 4.89a). The great toes are widened and shortened (Fig. 4.89b). The nose is disproportionately large, and the philtrum between the nose and the upper lip ends beneath the alae.

Although this disorder is rare and only occurs sporadically, one study has managed to investigate a total of 732 cases [9]. Instability of the patella was present in 3.4% of the patients. A particular problem is occasionally



Fig. 4.89a, b. 12-year old girl with *Rubinstein-Taybi syndrome*. **a** Hand with radial deviation of the thumb. **b** Feet of the same girl with widening and shortening of the great toes

posed by the presence of cervical spondylolisthesis since it can lead to tetraplegia. The thumb deformity frequently required surgical correction, since the deviation prevents normal opposition, making a pinch grip impossible or at least inadequate. A wedge osteotomy combined with a Z-plasty, and occasionally a rotation osteotomy, is usually required. If possible, the operation should be undertaken during the first two years of life so that hand-eye coordination can then develop normally.

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## 4.6.4 Chromosomal abnormalities

### Definition

This chapter deals with congenital diseases which are not attributable to individual defective genes, but in which whole chromosomes are abnormally formed, either by duplication (trisomy) or by the absence of a chromosome or chromosomal section. Autosomes or sex chromosomes may be involved.

#### 4.6.4.1 Down syndrome

### Definition

In Down syndrome, half of chromosome pair 21 is duplicated. Also known as Down syndrome, this is the commonest hereditary disease. It is characterized by diminished intelligence, mongoloid epicanthal fold, general hypotonia, short hands with a typical palmar

crease, internal rotation of the little finger, widened gap between the great toe and second toe, broad iliac wing and general ligament laxity. Heart defects and gastrointestinal anomalies are also often present. The (not especially frequent) orthopaedic problems are connected with the ligament laxity: atlantoaxial instabilities, habitual patellar dislocations, hip dislocation and flexible flatfeet.

— *Synonym: Trisomy 21, mongolism (no longer used as this term is politically incorrect)*

#### Etiology, pathogenesis, occurrence

Down syndrome involves duplication of chromosome 21, resulting in 47 instead of 46 chromosomes in the fertilized ovum. The defect usually occurs as a result of the failure of the chromosome to divide during mitosis. More rarely, the additional chromosome is translocated to another one, in which case the patient again has 46 chromosomes. This possibility is important to bear in mind in respect of genetic counseling. A mother with a translocated chromosome 21 has a risk of 1 in 3 that her next child will have Down syndrome, whereas the risk associated with the usual form is just 1 in 50.

The earliest evidence of the syndrome was found in a terracotta head dating from approximately 500 AD and associated with the Tolteca culture of Mexico, in which it is easy to identify the short palpebral fissures, oblique eyes, midface hypoplasia, and open mouth with macroglossia [7]. In the 1980's the incidence of trisomy 21 in England was calculated at 140/100,000 live births [4]. Thanks to prenatal diagnosis however (particularly ultrasound),

the tendency is sharply declining [6]. Down syndrome remains the commonest hereditary disease, followed by neurofibromatosis (► Chapters 4.6.1 and 4.6.6). The female:male sex ratio = 3:1. The risk of a child contracting Down syndrome increases with the age of the parents. The risk is 1/1,000 for a 20-year old mother, compared to 1/40 for a 45-year old woman [1].

#### Clinical features, diagnosis

The typical features are apparent even at birth: the mongoloid epicanthal fold, the general hypotonia and ligament laxity, the abnormal palmar crease, the inward-turning little fingers and the wide gap between the great and second toes. A possible heart defect and any gastrointestinal abnormalities may necessitate early surgical treatment. The children subsequently show psychomotor retardation.

— *Orthopaedic problems* mainly arise from habitual patellar dislocations, flexible flatfeet, refractory congenital or, later on, voluntary hip dislocations and atlantoaxial instability, all of which are attributed to the pronounced general ligament laxity. Some 8% of all children with Down syndrome experience *hip problems* even in childhood [8, 14] (► Chapter 3.2.7.2), compared to 22% in adulthood [5]. Although congenital hip dislocations are not especially common, they are difficult to treat as the *ligament laxity* obstructs attempts to achieve stable centering. As a result, secondary and voluntary dislocations occur (■ Fig. 4.90). *Femoral head necrosis* and *slipped capital femoral epiphysis* are frequent in Down syndrome [14].



■ Fig. 4.90a, b. Image converter x-rays of the left hip of a 7-year old girl with *Down syndrome*. Normal position (a), after *voluntary hip dislocation* (b)



Another frequently observed problem is *habitual* or *chronic patellar dislocation*, which generally occur bilaterally. Occasionally, the dislocations can also be produced at will. The same applies for the multidirectional instability of the *shoulders*, which is more commonly observed in trisomy-21 patients. *Flexible flatfeet*, with features similar to those of the idiopathic form, are also very often found in children with Down syndrome. Isolated cases of clubfeet have also been described for trisomy 21 [9].

Special attention must be paid to any *atlantoaxial instability* or *occipitoatlantal hypermobility* (► Chapter 3.1.11.14). The possibility of an atlantoaxial instability should be considered if the child has neck pain, torticollis, motor weakness or gait or micturition abnormalities. Functional x-rays of the cervical spine should be arranged if such signs and symptoms are present. These are also essential before operations or if the child wishes to take part in sports [11]. One large-scale study found that atlantoaxial instability was present in 8.5% of trisomy-21 patients. Of these two-thirds showed neurological symptoms [10]. It is particularly important that the anesthetist should be aware of any atlantoaxial instability before inducing anesthesia. Since inept manipulations can trigger neurological signs and symptoms, functional x-rays in maximum inclination and reclination are essential prior to surgical procedures.

**Radiographic findings:** The pelvic changes are characteristic (■ Fig. 3.213, ► Chapter 3.2.7). The ilium is broad and shaped like an elephant's ear (since it is rotated towards the frontal plane), the acetabulum is flat and the ischium is narrow. This configuration is also called »cordate pelvis«. X-rays of the lumbar spine often show that the vertebral bodies are disproportionately high in relation to their sagittal diameter.

- **Differential diagnosis:** Since the features of trisomy 21 are usually very obvious, the diagnosis is not difficult. It is confirmed definitively by chromosome analysis.

#### Prognosis, treatment

Whereas the life expectancy of trisomy-21 patients used to be significantly restricted, this is much less the case nowadays. Particularly since heart defects can now be treated surgically, most patients with Down syndrome reach adulthood and can live to the age of 70 [5]. As a result, osteoarthritis of the hip and knees is fairly common.

- The *treatment* of the heart defects and gastrointestinal abnormalities will not be discussed here. No treatment exists for the underlying disorder, nor is one likely ever to be developed. Prenatal diagnosis and genetic counseling are important, particularly if the parents are relatively old.

- **Hip dislocation:** We make a distinction between primary »congenital« dislocations and secondary, spontaneous dislocations. The latter do not appear until the age of 2–4 years and are attributable to the extreme ligament laxity. Dislocations can sometimes not appear until adulthood [5]. The treatment is very difficult, and conservative management usually proves unsuccessful. Surgical measures with capsular shrinkage and longer-lasting fixation are usually required (► Chapter 3.2.4).
- **Habitual patellar dislocation:** In this case physiotherapy should be administered with the aim of strengthening the quadriceps muscle. This sometimes produces the desired outcome, particularly if the vastus medialis muscle can be strengthened. Occasionally, however, surgical measures are also required (► Chapter 3.3.6).
- **Atlantoaxial instability:** Since neurological symptoms occur in 66% of patients with instability of the upper cervical spine [10], surgical stabilization is sometimes (but very rarely) unavoidable and is occasionally required even during childhood [3, 13]. The treatment involves atlantoaxial screw fixation or occipitocervical fusion, in which a spondylodesis is performed between the occiput and C2. The result is fixed either with wires or plates. Additional external stabilization either with a Minerva jacket or a halo is also required for a fairly prolonged period (► Chapter 3.1.10).

#### 4.6.4.2 Trisomy 8

Various abnormalities of the skeletal system are present in trisomy 8: Thus, the patient may have 13 ribs and the vertebral bodies are often wedge-shaped. The head is unusually large and the neck rather short. Contractures of various joints may be observed, and clubfeet and camp-todactylies and clinodactylies are usually also present. Trisomy 8 is very rare and occurs primarily as a mosaic form. The most important condition to consider in the *differential diagnosis* is *arthrogryposis*. In a few patients, however, no major abnormalities are apparent, in which case the diagnosis is often made by chance during the investigation of repeated abortions.

#### 4.6.4.3 Trisomy 5 (cri-du-chat syndrome)

This involves a defect of chromosome 5 in which one arm of the chromosome is missing. The name derives from the catlike whine emitted by the patients. Typical *orthopaedic problems* are clinodactyly, shortening of the metacarpals, congenital dislocation of the radial head and deformities of the spine. The children show severe mental retardation.



## 4.6.5 · Syndromes with neuromuscular abnormalities

### 4.6.4.4 Trisomy 18

Various abnormalities are observed in trisomy 18: A characteristic feature is an excessively long index finger, which is longer than the middle finger. The little finger shows a flexion contracture of the proximal interphalangeal joint. Clubfoot is also present in most cases. Both the pelvis and thorax appear narrow on *x-rays*. The skull is elongated and congenital abnormalities of the vertebral bodies are frequently observed. Patients with trisomy 18 have a poor prognosis and often die in early childhood. Only a few survive until adulthood, and such individuals usually develop a scoliosis that is relatively difficult to treat [12].

### 4.6.4.5 Turner syndrome

In the Turner syndrome one of the two X chromosomes is missing (monosomy X). The children have a female phenotype, but since the ovaries are missing they are amenorrhoeal. At birth the infant shows a short neck, cubitus valgus and a shortening of the 4th metacarpal. Various other abnormalities are also frequently observed (heart defects, renal deformities and deformities of the spine). These defects also determine the prognosis. While children with Turner syndrome can grow into adults, they remain mentally retarded.

### 4.6.4.6 Klinefelter syndrome (XXY syndrome)

This syndrome involves duplication of the X chromosome with the simultaneous presence of a Y-chromosome (XXY). The patients are phenotypically male, but with very small testes and no sperm production. The patients are unusually tall and the extremities, in particular, are very long. Patients show an increased incidence of radioulnar synostoses. A rickets-like osteopenia is also typical [15].

### 4.6.4.7 Fragile X syndrome

Fragile X syndrome is one of the commonest causes of mental retardation in boys. One study investigated 150 patients [2]: 57% showed abnormal ligament laxity, 7% scoliosis and 20% had flexible flatfeet.

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## 4.6.5 Syndromes with neuromuscular abnormalities

R. Brunner

### 4.6.5.1 Arthrogryposis

#### ➤ Definition

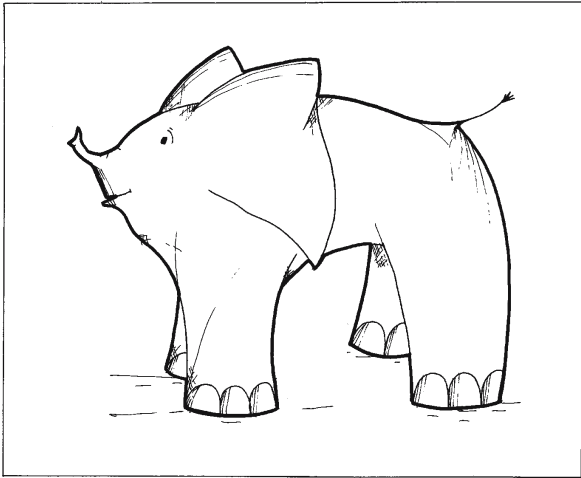
Arthrogryposis multiplex congenita is not a uniform clinical entity but rather a complex of symptoms resulting from the defective development of skeletal muscles. The complex is characterized by multiple congenital joint contractures. The extremities typically show symmetrical involvement. While the arthrogryposis may be neurogenic or myogenic in origin, the etiology often remains unknown.

#### Historical background

The condition was first described by Otto in 1841 (monstrum humanum extremitatibus incurvatus) [28]. Stern coined the term arthrogryposis multiplex congenita in 1923 [38].

#### Etiology, pathogenesis, occurrence

Arthrogryposis is characterized by the presence of multiple congenital, generally symmetrical contractures. Similar findings are seen in animals after viral disorders or contact with toxins (e.g. d-tubocurarine, botulinum toxin,



If the muscles didn't grow independently (by means of a separate growth organ), but were only passively »stretched« by the lengthening skeleton, the elephant would have a tiny trunk...

insecticides). In terms of etiology, there is no underlying uniform clinical entity. The complex of symptoms can also occur in association with various known disorders and syndromes (e.g. Möbius syndrome, Kniest syndrome, Pierre-Robin syndrome, myelomeningoceles, congenital muscle disorders). A hereditary component is not involved in most cases. Toxins and viruses (e.g. cytomegaly, rubella or herpes simplex) are responsible for most cases of arthrogyryposis in humans.

A striking increase observed in the incidence of arthrogyryposis in English-speaking countries in the 1960's may have been triggered by a virus. Either defective formation or the destruction or inhibition of central nervous structures involved in muscle control during embryogenesis can prevent the normal development of muscle tissue. A similar clinical picture can arise if the motor endplates fail to form or if disorders interfere with endplate function or the muscles themselves. This results in a lack of movement just at the time in embryogenesis when the joints are developing.

Although the causes of the lack of movement are varied, the response of the body is uniform: the formation of extra connective tissue in the muscles and joint capsules. In one study involving 74 children with arthrogyryposis, a neurogenic cause was found in 93% of cases, while the cause was myogenic in just 7% of cases [3]. Hereditary causes are rare, although isolated hereditary forms do exist [40].

Normal muscles appear to possess a kind of growth zone at the aponeurosis where the muscle fibers are attached. Here the muscle grows, mainly in length, in response to stretching stimuli by the deposition of contractile elements [25]. In arthrogyryposis, parts of the muscle (or whole muscles) develop as fibrous bands that lack the

ability to grow in length. The amount of connective tissue in the joint capsules also increases, which explains the substantial failure of measures aimed at improving joint mobility during growth. The *incidence* of arthrogyryposis is approx. 3/100,000 births.

#### Clinical features and diagnosis

Children with severe arthrogyryposis (with additional CNS abnormalities) are often incapable of survival. A lack of movement by the infant during pregnancy may be noticed. On clinical examination the pattern of joint involvement is symmetrical, and the movement of the joints is restricted generally. Flexion or extension contractures or combinations of the two may be present. The extremities appear cylindrical as a result of the defective muscle formation, giving the infant the appearance of a stuffed doll. The normal skin wrinkles are missing, although sensation remains intact. All four extremities or the lower limbs only are affected in just under half of the patients in each case, while the upper limbs only are affected in approx. 10% of cases. Peripheral (distal) involvement is common, and these patients with poorly functioning hands and feet generally have good trunk muscles. Complex foot deformities are usually present, producing a picture reminiscent of clubfoot (■ Fig. 4.91). Tarsal coalitions may also be present concurrently [37]. Flexion and extension contractures are observed at knee level.

Patients with proximally accented forms have effectively functioning hands but poor or completely lacking muscle activity in the trunk. A muscle, and possible nerve biopsy can be useful in identifying the primarily damaged organ. Further investigations (e.g. a chromosome analysis or MRI scan) may then be needed depending on the result. From the orthopaedic standpoint, the skeletal configurations in the affected parts of the body are of primary interest. The hips are very often affected (in 80% of cases) [1, 44], the knees [36] and feet [22, 37] to a lesser extent. Investigation of the hips, by ultrasound at birth and subsequently by radiographic methods or MRI, is indicated in every case.

If clinically visible deformities of the knees are present (and to a lesser extent the feet) imaging procedures can also reveal the pathological anatomy and produce a basis for the treatment plan. Fractures or epiphyseal separations often occur at birth, most often in the vicinity of the femur, as a result of the rigid joints.

#### Treatment, prognosis

The progression depends on the underlying disorders. In most cases only the effects of the harmful agent are apparent in the form of the prevailing signs and symptoms, while the cause remains unknown. Most underlying disorders that have resulted in arthrogyryposis are not progressive. The coexistence of normal and abnormally



■ Fig. 4.91. Clubfoot in arthrogyposis

formed muscles can lead to problems. Only healthy muscles behave normally in respect of growth and trainability. Pathologically altered muscles on the other hand are difficult to stretch and experience restricted growth.

Certain functionally relevant improvements can be achieved with *conservative measures* (physiotherapy, orthotic provision). In stubborn cases, the deterioration of the joint contractures and the necessary surgical correction can at least be delayed. Orthoses are needed to secure deformed feet sufficiently to enable the patient to walk or to protect hand deformities from severe progression. It is a good idea to delay surgical measures for as long as possible, since recurrences during growth tend to be the rule. Definitive surgical corrections are only possible after the completion of growth.

*Surgical measures* must take account of the patient's already diminished muscle power. Muscles that are still active may be lengthened but the associated power loss should be kept to a minimum. Only if the muscles are completely degenerated can muscle power be disregarded, in which case simple tenotomies may be appropriate. Since the muscle contractures are usually accompanied by capsular contractures however, simple tendon lengthening procedures on their own are often not sufficient. In light of the foregoing, external fixators are particularly suitable for correcting joint deformities, since they allow all of the sections that are contributing to the contracture to be stretched at the same time [8, 16, 29]. They also offer corrective options in all dimensions (■ Fig. 3.347–3.349, ► Chapter 3.3.13).

Unfortunately they are only suitable for knees and feet. More problematic is the treatment of *hip flexion contractures*. We have not managed to confirm reports

in the literature of good results obtained with purely conservative methods. This problem can be resolved to some extent satisfactorily only through carefully performed tendon lengthening procedures, although these always result in a loss of power [7]. Dispute exists about the correct approach for bilateral *hip dislocations* (as is the case with most disabled patients) [1, 40]. Studies have shown, however, that the functional results for patients who are able to walk are better if the hips are reduced (► Chapter 3.2.8).

*Knee contractures* respond poorly to physical therapy on its own, and conservative appliances (corrective casts, knee supports) may be required. Surgical measures are successful in the short term, but recurrences are common in the long term [8, 24]. Almost any contracture can be stretched with external fixators [8, 10] (► Chapter 3.3.13). The tendency for recurrences to occur declines from the age of ten. Definitive corrections (particularly foot corrections by means of arthrodesis) should be implemented only after growth is complete. By contrast, dislocated joints (particularly of the hip and knee) should be reduced as soon as possible after birth. The recurrence tendency also applies after corrections of *foot deformities*. Only a talectomy, which can be performed in older children, produces better results (■ Fig. 4.92).

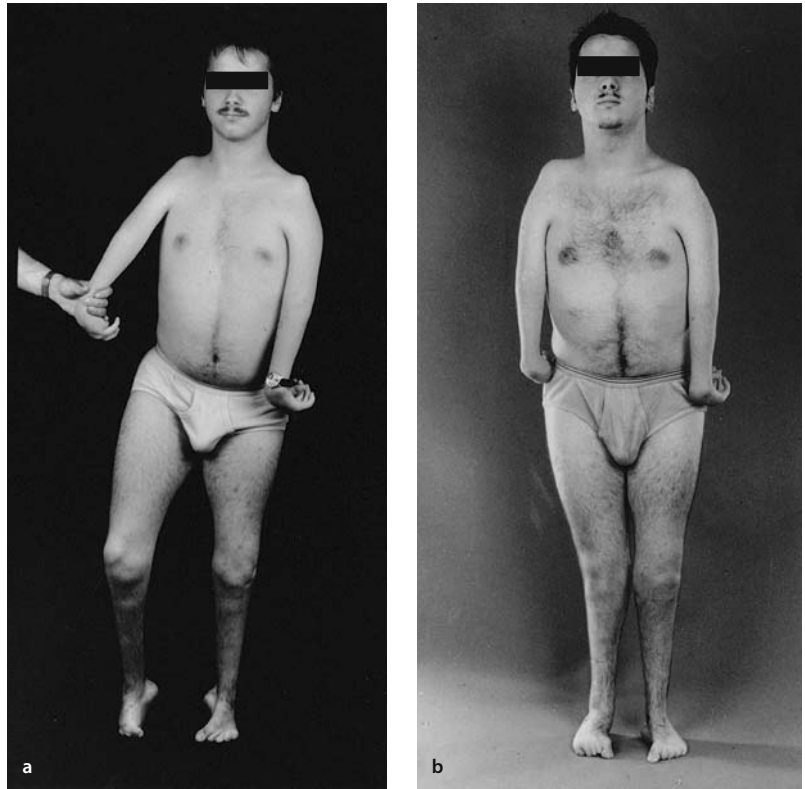
The *upper extremities* are likewise generally affected by symmetrical contractures, although patients tend to adapt to these amazingly well in everyday life. The main handicap arises from the loss of power, which is not amenable to improvement, although the range of motion of the joints can be enhanced or transferred by orthopaedic methods (conservative and surgical). The primary goal of treatment must be to optimize the functioning of the upper limbs. The patients should be able to eat independently, take care of their own personal hygiene and, if possible, be able to write. Any surgical corrections should always include the whole extremity (shoulder, elbow and hand) [2].

#### 4.6.5.2 Pterygium syndromes

Pterygium syndrome involves a group of disorders characterized by congenital webbing of the skin across joints. These are classified according to the location of the pterygia and the life expectancy. A total of six lethal and seven survivable forms are distinguished. Three forms that commonly require orthopaedic treatment are mentioned below:

##### — Popliteal pterygium syndrome:

This syndrome probably has an autosomal-dominant mode of inheritance [9], the gene map locus is 1q32-q41. Popliteal pterygia are characteristic of the syndrome. Other typical features include a cleft lip and palate, lower lip fistulas, cutaneous syndactyly of the toes, nail abnormalities and anomalies of the external



**Fig. 4.92a, b.** 16-year old patient with *arthrogryposis*. **a** Preoperatively he had been unable to stand freely, and orthoses could no longer be fitted satisfactorily. A leg length discrepancy of 4.5 cm was also present. **b** 3 years after corrective osteotomy of both feet at ankle joint level and concurrent leg length equalization with the aid of external ring fixators. The patient is able to stand freely and wear normal shoes with inserts. The result has been preserved unchanged

genitalia. Hypoplasia of the toes and lower legs, patellar agenesis or scoliosis with vertebral deformities may also be present. Intelligence is normal. Since the sciatic nerve passes through the pterygium, the options for correction are limited.

— *Progressive form of multiple pterygium syndrome:*

This syndrome occurs sporadically and may involve autosomal-dominant new mutations [9]. Characteristic features include contractures of the toes (camptodactyly) and hips, clubfeet, muscle hypoplasia, scoliosis, keeled chest, facial dysmorphias, small stature with reduced weight and recurrent respiratory infections. The pterygia occur secondarily on the neck and shoulder.

— *Recessive form of multiple pterygium syndrome:*

This is an autosomal-recessive inherited disorder [9], the gene map locus is 2q33-q34. It is also known as Escobar syndrome. The differential diagnosis must rule out other syndromes (Turner, Noonan, Leopard). The first diagnostic sign is that of reduced fetal movement detected by ultrasound or reported by the mother [17]. Pterygia in the popliteal fossa are the main feature, although they may also occur on the neck, elbow or the axilla. The patients also suffer from scoliosis with vertebral and finger deformities [9, 33].

Extension contractures of the hips, clubfeet or metatarsus adductus, facial dysmorphias with cleft lip and

palate and retardation of the secondary sex characteristics with cryptorchism and small stature may also be present. The patient's intelligence is normal. The pterygia become more pronounced with age and lead to contractures. Since the sciatic nerve remains in its normal anatomical position correction is minimally restricted.

From the orthopaedic standpoint, the pterygia in the popliteal fossa are the prime concern. These extend from the ischial tuberosity to the calcaneus and, anatomically, form a ligament-like structure consisting of muscle or connective tissue immediately beneath the subcutis. They produce a flexion contracture at the knee and an equinus foot. The position of the sciatic nerve is important as it can pass through the pterygium. A preoperative MRI scan can clarify the pathoanatomical situation. The anatomy of the blood vessels remains normal in all forms of the pterygium syndrome.

The following *therapeutic* options are available:

- Excision of the connective tissue/muscular band. The skin is lengthened by a plastic surgery procedure (Z-plasty or with interposed tissue). The knee is then stretched continuously by conservative means (cast or orthoses).
- Shortening or extension osteotomy of the distal femur [26].



- Joint-bridging external fixators [10] (► Chapter 3.3.13). The pathological subcutaneous band should be resected to prevent recurrences. Stretching of the knee may need to be supplemented by Achilles tendon lengthening. The result after surgical correction is difficult to predict and the condition has a strong tendency to recur [33]. Intensive physiotherapy is needed to preserve major joint function.

#### 4.6.5.3 Goldenhar syndrome

The prevalence of this syndrome, also known as oculo-auriculovertebral syndrome or hemifacial microsomia, is 0.9–2.1 patients/million inhabitants [43, 44]. The gene map locus is 14q32. Various inheritance patterns, including autosomal-dominant or -recessive, are discussed in the literature [9, 38, 43], as is the possibility of its development during embryogenesis [19]. The syndrome involves the impaired development of the derivatives of the 1st and 2nd branchial arches [9]. Patients with Goldenhar syndrome show facial asymmetry, one-sided mandibular hypoplasia, cleft lip and palate, auricular hypoplasia, eye abnormalities, deformities of the cervical spine and cardiac anomalies. Ipsilateral renal agenesis and uterine deformities and mental retardation are possible [9, 43]. Spinal abnormalities, clubfeet, congenital hip dislocations, Sprengel deformity, radial defects and basilar impression may require orthopaedic treatment [12, 15].

#### 4.6.5.4 Moebius syndrome

The syndrome was described by Möbius in 1888 [23]. The basic defect is unknown [43] and the disorder only occurs sporadically (approx. 300 described cases). An autosomal-dominant mode of inheritance is suspected, the gene map locus is 13q12.2–q13. It can be difficult to differentiate Möbius syndrome from Poland syndrome (► Chapter 4.6.6.10).

Typical features are congenital cranial nerve pareses and bilateral facial palsy, bilateral abducent nerve paresis and/or a deficit of the vestibular nerve system and cranial nerve IX. The cranial nerve VII (facial nerve) may be absent [41]. The tongue atrophies resulting in weak mastication [9]. There may also be mandibular hypoplasia, laryngeal stridor, epicanthus, syndactylies and other limb deformities (e.g. clubfoot). Intelligence is restricted in a minority of cases, although life expectancy is reduced. The orthopaedic problems arise from the hand and foot deformities.

#### 4.6.5.5 Cornelia-de-Lange syndrome

The incidence of this autosomal-recessive disorder is approx. 0.7/100,000. The syndrome is probably caused by a change at 5p13.1, although a lesion in chromosome 3

has also been discussed. This rare condition (also known as Brachmann-de-Lange syndrome) is characterized by stunted growth, microcephaly, bushy eyebrows (which join over the base of the nose), epicanthus, diminished growth, limb abnormalities (small hands and feet, proximally displaced thumb, clinodactyly of the little finger, ray defects on the ulnar and radial side, possibly with monodactyly), simian crease, hypertrichosis, abnormalities of the genitalia, internal deformities with heart defects and gastrointestinal or renal abnormalities and mental retardation [9]. Distal ray defects are also present on the extremities [9, 42].

Life expectancy is reduced [43]. Most children experience a permanently delayed development in their learning abilities, although the spectrum of intellectual skills is wider than used to be assumed some years ago. Early counseling and individual stimulation is very important for maximizing the development possibilities.

The *orthopaedic problems* arise from the combination of the limb deformities and psychomotor retardation. In our experience, conservative measures generally prove sufficient since the children – subject to the limitations imposed by their deformed limbs – cope well in accordance with their intellectual skills.

#### 4.6.5.6 Pierre-Robin syndrome

This syndrome was first described by Pierre Robin in 1923 [31]. The prevalence is 0.9 patients/million inhabitants [44]. The Pierre-Robin syndrome is comprised of abnormalities of the brain stem, jaw and palate (microgenia, glossoptosis and cleft palate) [9]. The small mandible, which is too far back, and the cleft lip and palate cause the weak tongue to fall back, potentially interfering with respiration. Heart defects and brain abnormalities with mental retardation occur in approx. 20% of patients respectively [9]. An associated occipito-atlantoaxial instability may also be present as a result of a defectively formed atlas.

The orthopaedic problems arise from the limb deformities, with constrictions, clubfeet, hip dislocations, syndactylies and sternal abnormalities [9]. Other orthopaedic problems are posed by excessively long and enlarged thumbs and great toes, which can also deviate progressively in a medial or radial direction and require surgical correction [5].

#### 4.6.5.7 Williams-Beuren syndrome

This condition was first described, independently of each other, by Williams [42] and Beuren [6]. The incidence is approx. 3/100,000 [43]. This autosomal-recessive syndrome is inherited as a result of a deletion at 7q11.23, and involves an alteration of several proteins involved in elastin synthesis. The condition is charac-



terized by facial hypoplasia and supraaortic stenosis. Other signs and symptoms include diminished growth, characteristic facies, dental abnormalities, renal deformities, radioulnar synostoses, calcification of the intervertebral disks, scolioses and a low voice. The affected children are mentally retarded. The orthopaedic problems arise from the developmental retardation with coordination problems, radioulnar synostoses and any scoliosis [27].

#### 4.6.5.8 Prader-Willi-Labhard syndrome

This inherited disorder was first described in 1956 by Prader, Labhard and Willi [32]. A translocation or deletion on chromosome 15 has been described [9, 21, 42] (gene map locus 15q11-q13) and confirms the diagnosis. The incidence is approx. 4/100,000 [9]. The syndrome is characterized by muscle hypotonia, massive obesity, psychomotor retardation, delayed skeletal maturity, hypogonadism and ligament laxity. Small hands and feet and the subsequent development of insulin-resistant diabetes mellitus are additional general signs [9, 42]. The life expectancy is reduced to 20–30 years [9]. Spinal deformities represent the main orthopaedic problem in Prader-Willi syndrome [34]. Since these occur in 80% of cases, regular orthopaedic check-ups are indicated.

#### 4.6.5.9 Rett syndrome

Rett syndrome was first described in 1966 by Rett [35]. The incidence is approx. 10/100,000 births and the gene defect is on the sex chromosome (Xq28). The condition only affects females, probably because the defect is lethal when just one X chromosome is present (with a few exceptions [11]).

The basic defect responsible for the mainly extrapyramidal progressive disorders with atrophy of the cerebrum is not known [9, 42]. The patients' development initially appears normal, and they are able to maintain an upright posture, stand and walk and possibly achieve a certain degree of independence during the first 6–18 months of life. At birth the head circumference is normal, although head growth is subsequently delayed [9, 42]. Motor and cognitive functions, including the use of the hands, speech and the ability to walk, progressively disappear after the age of 6–18 months, resulting in apraxia of gait and trunk control with signs of ataxia. Other symptoms are respiratory problems, apnea attacks, spasticity, scoliosis and mental retardation. The patients show typical stereotypic hand movements [4, 13].

The scoliosis is the main orthopaedic problem in Rett syndrome, with a reported incidence of over 50% [18]. It develops as a typical neurogenic, long drawn-out C-shaped scoliosis, in some cases with hypokyphosis, that responds poorly to conservative treatment. A compensa-

tory countercurve forms while the patients are still able actively to straighten up. The scoliosis in Rett syndrome occurs during childhood and its progression depends on the advance of the underlying disease [14, 18, 20]. Surgical correction and stabilization of the spine are indicated by way of treatment if the scoliosis angle exceeds 40°. While corset treatments are possible, and also effective, they hinder the patient to a greater extent compared to a single surgical corrective procedure.

The *atactic gait pattern* hampers walking and standing. Physiotherapy exercises designed to improve body control can be helpful, as can stabilizing orthopaedic appliances for the feet (stabilizing shoes, possibly lower leg splints). Since the patients tend to suffer muscle contractures that interfere with their ability to walk stretching exercises are indicated. Severe muscle shortening may require surgical intervention. Hand braces are fitted to the patients in order to correct the *stereotypic hand movements*.

#### 4.6.5.10 Dandy-Walker syndrome

This syndrome involves the formation of cysts in the brain, usually in association with hydrocephalus. Pronounced mental handicap and a gait disorder with an atactic gait pattern are present in over half of the cases. Life expectancy is significantly restricted in the severe forms. Isolated milder cases are not diagnosed until adulthood. The clinical picture can also include severe scolioses and kyphoses. The gene map locus is 3q24, the inheritance is unclear. The treatment involves the insertion of a shunt in order to drain the hydrocephalus. Where appropriate, the scoliosis or kyphosis should be treated. However, the surgeon should be very careful in deciding whether an operation is indicated since, if a gait disorder is present, the reduction in the mobility of the spine may result in the loss of the ability to walk.

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#### 4.6.6 Various syndromes with orthopaedic relevance

This chapter deals with those congenital disorders that are not listed in the »International Nosology and Classification of Constitutional Disorders of Bone« 2001 [18], but which are nevertheless of orthopaedic significance. Most are hereditary conditions, while the mode of inheritance is not clear in others.

##### 4.6.6.1 Gaucher disease

This is an autosomal-recessive inherited disorder (gene locus 1q21) and is one of the storage diseases. An enzyme defect (glucocerebrosidase deficiency) results in the abnormal storage of glucocerebrosides. The condition was described by Gaucher in 1882 [12]. A similar enzyme defect has been identified in Niemann-Pick disease. Gaucher disease is a rare disorder that occurs more commonly in the Jewish population in Israel and in Ashkenazi Jews in eastern Europe.

Gaucher disease is classified in three forms:

- *Acute, infantile neuropathic form*, which proves fatal during the first few months of life.

- *Chronic, non-neuropathic form*, which often manifests itself during adolescence. The signs and symptoms are highly variable.
- *Subacute, neuropathic or juvenile form*. This resembles the chronic form, but is associated with additional neurological symptoms; the onset occurs during childhood.

Another classification distinguishes [31]:

- *Type I: noncerebral juvenile form*
- *Type II: infantile, cerebral form*
- *Type III: juvenile and adult neuropathic form* (also Norrbottnian type)

The disease manifests itself *clinically* either during the course of childhood (type III) or during adolescence (type II), with splenomegaly, skin pigmentations and enlarged lymph nodes. Type III is additionally characterized by muscle hypotonia with gait abnormalities, strabismus and psychomotor retardation. These are soon followed by anemia, bleeding tendency and repeated infections. The following skeletal changes are observed: Infiltration of the bone marrow, bone necroses, »bone crises« [26], pathological fractures, osteolysis and osteomyelitis [56]. All of these changes are attributable to the deposition of Gaucher cells in the bone marrow. The metaphysis and diaphysis of the affected bone are widened and the cortical bone becomes thinner (■ Fig. 4.93).

These changes often remain asymptomatic for a fairly long time. The storage of Gaucher cells can ultimately lead to impairment of the blood supply and thus to *necrosis*. This typically affects the femoral head, resulting in potential confusion with Legg-Calvé-Perthes disease, although the necrosis can also occur in the head of the humerus or in any metaphysis or diaphysis.

A particularly characteristic event in Gaucher disease is the »bone crisis« [26]. This is characterized by intense pain in the extremity with local tenderness, excessive warmth, redness and occasionally fever as well. During the crisis, an increased uptake is apparent on the technetium bone scan [26]. The crisis subsides after a few days and the pain disappears spontaneously. It is thought that the blood supply is temporarily disrupted during crises as a result of the infiltration with Gaucher cells.

*Pathological fractures* often occur in patients with Gaucher disease. These are often diagnosed at a belated stage and are associated with a poor healing tendency in view of the underlying bone necroses.

One condition that can sometimes be difficult to diagnose is *osteomyelitis*, which can also occur in association with local necrosis. Since the osteomyelitis is not infrequently misdiagnosed as a bone crisis, the possibility of this diagnosis should be borne in mind if the crisis lasts longer than a few days. A gallium bone scan can be helpful in establishing the existence of a bacterial infection.

**Treatment of the underlying disorder**

A breakthrough was made some years ago in the treatment of Gaucher disease, following the production of the defective enzyme by genetic engineering techniques. The missing enzyme can now be substituted with Ceredase [38]. The treatment is very expensive, but is capable of alleviating the frequent orthopaedic problems, which can often be difficult to treat.

#### Treatment of the underlying disorder

**Orthopaedic treatment**

The orthopaedic treatment depends on the nature of the complication. Children are particularly affected by pathological fractures and osteomyelitis. The fracture management is very difficult since the fractures are associated with underlying necrosis. Long periods of immobilization are frequently required. The osteomyelitis is treated by local clearance and the administration of antibiotics. Interventions on the spine are occasionally required, particularly if the vertebral bodies collapse, resulting in increasing kyphosing at the thoracolumbar junction (■ Fig. 4.94) [27]. The frequently occurring femoral head necrosis can result in premature osteoarthritis of the hip and the need for a total hip replacement [33].

#### Orthopaedic treatment

The orthopaedic treatment depends on the nature of the complication. Children are particularly affected by pathological fractures and osteomyelitis. The fracture management is very difficult since the fractures are associated with underlying necrosis. Long periods of immobilization are frequently required. The osteomyelitis is treated by local clearance and the administration of antibiotics. Interventions on the spine are occasionally required, particularly if the vertebral bodies collapse, resulting in increasing kyphosing at the thoracolumbar junction (■ Fig. 4.94) [27]. The frequently occurring femoral head necrosis can result in premature osteoarthritis of the hip and the need for a total hip replacement [33].

■ Fig. 4.93. AP x-ray of the pelvis in a 7-year old girl with *Gaucher disease* and an intertrochanteric fracture of the right femur





■ Fig. 4.94. X-ray of the spine of a 9-year old boy with *Gaucher disease* and a low thoracic kyphosis

#### 4.6.6.2 Neurofibromatosis

##### ► Definition

Common autosomal-dominant, multisystemic hereditary disorder with tumorous changes of the connective tissues associated with the central and peripheral nervous system in the skeleton, skin and soft tissues. Typical orthopaedic problems include leg lengthening, congenital tibial pseudarthroses and short-curved progressive scolioses, often with a kyphotic component.

■ *Synonyms:* von Recklinghausen disease

##### Historical background

The disorder acquired its name from a monograph written by F. von Recklinghausen in the year 1882 [44], although isolated cases had already been described before that date. In 1918 Gould mentioned the occurrence of scolioses in connection with neurofibromatosis. The condition became known to the public through Joseph Merrick, a man with a grotesquely deformed skull who was the subject of a play by Bernard Pomerance entitled »The Elephant Man«, which was later made into a successful film. However, more recent investigations of his skull have revealed that John Merrick had suffered not from neurofibromatosis, but rather from Proteus syndrome.

##### Etiology, pathogenesis

This is an autosomal-dominant disorder although most cases occur as new mutants. Two types are distinguished. The gene locus of the common type I is 17q11.1–11.2, while that for the rare type II is 22q12.1–12.3. The proliferating cells are derived either from the Schwann cells or accompanying cells. Viewed macroscopically the neurofibromas are bright, relatively compact nodules that are connected to the peripheral nerves. The affected nerves are usually thickened. Neurofibromas also occur in the central nervous system (brain, spinal cord).

##### Occurrence

After Down syndrome, neurofibromatosis is the most common hereditary disease. The incidence of type I in Finland is 27.4/100,000 births (with no difference between the sexes) and the prevalence 225/1,000,000 inhabitants [43]. In Israel, prevalence figures of 104/1,000,000 males and 119/1,000,000 females have been reported [13]. Type II neurofibromatosis is much less common (1/1,000,000 inhabitants).

##### *Neurofibromatosis type I*

##### Clinical features, diagnosis

At least two of the following clinical *characteristics* are required before a diagnosis of neurofibromatosis can be made [45]:

- *Café-au-lait spots:* Light brown, regular pigmented lesions that can cover large areas. More than five such spots are required to confirm the diagnosis. 99% of patients with neurofibromatosis show this sign.
- *Nodules (fibroma molluscum):* These are subcutaneous neurofibromas derived from the Schwann cells of the peripheral nerves. They are observed in 16% of patients.
- *Spinal changes:* Characteristic signs are scolioses and kyphoscolioses (► Chapter 3.1.11), which may be of the dystrophic or idiopathic type. *Dystrophic scoliosis* is short-curved and has a kyphotic component. The vertebral bodies show a curved indentation, the ribs are thinned on the concave side facing the spine and markedly rotated (■ Fig. 3.103, ► Chapter 3.1.11). The apex of the scoliosis is usually at the level of the lower thoracic spine [11]. However only 10% of patients show this typical scoliosis [2, 52]. Scolioses of the *idiopathic type* can also occur.
- *Leg length discrepancies:* These usually occur in connection with substantial accumulations of subcutaneous neurofibromas, which lead to growth stimulation and *lengthening* of the extremity. Such lengthening occurs in approx. 10% of patients.
- *Neurological lesions:* Discrete neurological changes, particularly sensory abnormalities, occur in 15% of cases. A characteristic symptom is hyperesthesia.



In addition to these classic signs and symptoms, of which at least two must be present, the following *additional changes* can occur:

- **Elephantiasis (*pachydermatocele*):** hypertrophy of the skin with subcutaneous neurofibromas (■ Fig. 4.95).
- **Congenital pseudarthrosis of the tibia:** 40% of patients with congenital tibial pseudarthrosis suffer from neurofibromatosis. Within the population of neurofibromatosis patients, however, tibial pseudarthrosis is rather rare (approx. 2%). A detailed description of congenital tibial pseudarthrosis can be found in ► Chapter 3.3.6.5.
- **Paravertebral soft tissue tumors:** These are usually associated with scoliosis. They are readily viewed on the MRI scan and must be differentiated from ganglioneuromas.
- **Axillary freckles:** These are hyperpigmented spots in the axillary area with a diameter of 1–3 mm.

The diagnosis of neurofibromatosis is confirmed clinically. The family history must also be noted. A positive history is present in only a minority of patients and most cases occur as new mutants. X-rays are needed for diagnostic purposes and to monitor the spinal changes and congenital tibial pseudarthrosis. The MRI scan can reveal paravertebral, intraspinal and intracerebral tumors.

#### Differential diagnosis

Neurofibromatosis can be confused with the very rare *Proteus syndrome* [4] (see above: »Historical background«). Nodular, and potentially very major, changes in the skin can also occur in this condition, and the patients may also show macrodactyly. But the nodules do not consist of neurofibromas, but rather of hamartomas derived from

fatty tissue. In addition to the neurofibromatosis *solitary neurofibromas* are also present in Proteus syndrome, although these do not usually affect children and adolescents, but patients aged between 20 and 40 years. These occur subcutaneously and constitute harmless changes.

#### Prognosis, treatment

Patients with neurofibromatosis have a largely normal life expectancy. Around 50% show diminished intelligence.

A very important prognostic indicator is the malignant degeneration of the neurofibromas. The corresponding risk is of the order of 5%. We have observed three such cases. Isolated reports of degeneration into malignant schwannomas or rhabdomyosarcomas are also described in the literature [20].

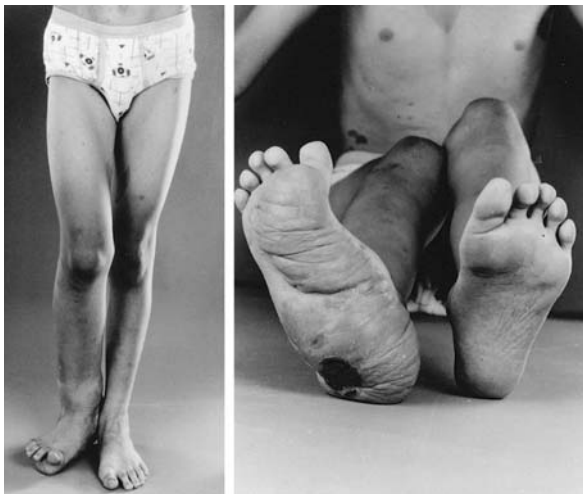
No known *treatment exists for the underlying disorder*. The *orthopaedic treatment* is primarily concerned with three areas: Spinal changes, leg length discrepancies and congenital tibial pseudarthrosis. The *spinal changes* must be diagnosed as soon as possible and then monitored very closely. The scolioses are usually strongly progressive and corset treatments are not very effective. As a result, early surgical treatment is often indicated (► Chapter 3.1.11).

*Leg length discrepancies* are associated with – usually severe – cutaneous changes, and also with sensory abnormalities. Collaboration with a plastic surgeon is often required in such cases. The thickened, flabby skin on the sole of the foot can lead to pronounced trophic problems and pressure sores. Removal of the excess skin by the plastic surgeon stabilizes the sole and can considerably alleviate the problem. From the orthopaedic standpoint, as soon as a discrepancy exceeds 2 cm, leg length equalization by means of epiphysiodesis should be attempted. Surgical lengthening of the healthy leg should be avoided, otherwise both legs will be impaired in the event of any complications. Shortening of the hypertrophic leg is problematic. In view of the prevailing soft tissue changes, the risk of vein thrombosis and other complications is very high. The surgical leg length equalization procedure is described in ► Chapter 4.2.2.

Since the course of *congenital tibial pseudarthrosis* in patients with neurofibromatosis is not different from that in other patients, the therapeutic principles are completely identical (► Chapter 3.3.6.5).

#### Neurofibromatosis type II

This type of neurofibromatosis is characterized by multiple tumors (schwannomas) of the cranial and spinal nerves. The initial symptom is central hearing loss, since cranial nerve VIII is usually affected. Poor eyesight, sensory abnormalities and weakness of the facial muscles can also occur. This type is also known as »bilateral acoustic neurofibromatosis« or »central type«. Malignant peripheral nerve sheath tumors can develop from the schwannomas (► Chapter 4.5.4.3). The prognosis is poor. The tumors



■ Fig. 4.95. 8-year old boy with *neurofibromatosis* and *elephantiasis* with hypertrophy of the skin and subcutaneous neurofibromas



can only be treated surgically, but usually leave residual functional deficits.

#### 4.6.6.3 Proteus syndrome

Neurofibromatosis can be confused with the very rare *Proteus syndrome* [51], of which approx. 100–200 cases worldwide have been described in the literature to date. The syndrome is characterized by macrosomia, which invariably affects individual structures and never the whole body. Thus, individual fingers or toes, a whole limb or even one half of the body may be enlarged. Tumorous thickening of the fatty or connective tissues can also occur. The skin may show areas of striated thickening or vascular markings (■ Fig. 4.96). Over time, such changes can lead to functional problems of the affected organs. These can be alleviated by early diagnosis and careful follow-up and, if specifically required, by surgical measures.

The syndrome is polymorphic by nature and manifests itself in highly individual ways – which explains why it was named for the Greek demigod Proteus, who was able to escape from his enemies by altering his outward appearance. While the *etiology* of Proteus syndrome is not fully understood, it is thought to be caused by a genetic change that occurs during the first few weeks of pregnancy and that affects only a few individual cells. Other conditions to be considered in the *differential diagnosis* are the Klippel-Trenaunay syndrome and neurofibromatosis. *Treatment* is limited to the surgical removal or the reduction in size of functionally or cosmetically troublesome areas of thickening.

#### 4.6.6.4 Klippel-Trenaunay syndrome

##### ► Definition

Rare congenital abnormality characterized by large hemangiomatous nevi, unilateral hypertrophy of the soft tissues and bones and venous varices. Anomalies of the finger and toes and spinal changes can also occur.

— *Synonym:* Klippel-Trenaunay-Weber syndrome

##### Historical background, Etiology, occurrence

The syndrome was described in 1900 by M. Klippel and P. Trenaunay [30]. It occurs very rarely. The disorder is thought to be caused by a mutation at 5q13.3 during fetal development with a mosaic gene anomaly, which would be lethal if it affected all cells. Other investigations have indicated the possibility of a paradominant inheritance [19].

##### Clinical features, diagnosis

The diagnosis can be confirmed even in infancy by clinical examination. The vascular nevi are usually very striking and extensive. They can cover large sections of an extremity or the trunk, are red-bluish in color and



■ Fig. 4.96. Clinical appearance of the feet of a 9-year old boy with *Proteus syndrome*. Note the hypertrophy of the right foot and the tumorous skin changes

the overlying skin is very sensitive. The vascular changes also affect the underlying soft tissues where the veins show varicose changes. The increased circulation leads to growth stimulation, potentially causing the affected extremity to become much too long (■ Fig. 4.97). The severity of the signs and symptoms is variable. The vascular nevi always affect a lower limb, usually along the whole of its length.

Abnormalities of the fingers and toes are also almost always present in the *classical form* of the disease: macrodactylies, syndactylies, metatarsus primus varus, clinodactylies, polydactylies, camptodactylies or stenosing tenosynovitis [21]. The »*forme fruste*« of the disorder is slightly rarer, and finger abnormalities only occur sporadically in this form. Changes in the spinal column are not infrequent: scolioses, kyphoses, hemivertebrae and intraspinal tumors [16].

The most difficult task in the *differential diagnosis* is to distinguish this condition from *Proteus syndrome*, which is also characterized by a hemihypertrophy, although without the vascular nevi. On the other hand, lipomatous hamartomas are found in the skin of patients with Proteus. Hypertrophy can also occur in *neurofibromatosis*,

although the skin changes clearly differ from those in Klippel-Trenaunay syndrome.

### Treatment

The most important orthopaedic problem is the *leg length discrepancy*. In Klippel-Trenaunay syndrome this can pose considerable difficulties since surgical lengthening of the other, healthy leg is out of the question. The limb affected by the disorder is usually altered so greatly, that the risk of damaging the health leg is unacceptable. A shortening osteotomy of the diseased extremity is also extremely problematic because of the varicose veins and the associated risk of vein thrombosis and pulmonary embolism.

For the above reasons, *leg length equalization* should always be attempted in the form of a timely *epiphysiodesis* on the diseased extremity (► Chapter 4.2.2). The morbidity associated with this procedure is very low. The correct timing of the operation is naturally difficult. In doubtful cases however, the epiphysiodesis should be performed too early rather than too late. In a worst-case scenario in which the healthy leg also threatens to grow excessively long, an epiphysiodesis would also have to be performed on this leg at a later date. Since the patients are not small in stature this is not especially problematic. The treat-

ment can be very difficult however, if epiphysiodesis is not performed in time. Even an amputation often fails to resolve the problem in these patients since the use of a prosthesis is rendered almost impossible as a result of the skin changes on the thigh and buttocks.

### 4.6.6.5 Marfan syndrome

#### Definition

Autosomal-dominant hereditary disorder with impaired collagen synthesis and characterized by excessively long extremities compared to the trunk and the increased occurrence of ligament laxity, ectopia lentis, aortic dilatation, scolioses, acetabular protrusion and foot deformities.

— **Synonyms:** Arachnodactyly («spider fingers»), dolichostenomelia (long, thin limbs)

#### Historical background

The condition was described by Marfan in 1896, although the case that he described involved the rare contractural form. It is now assumed, on the basis of photographs, that the former American President Abraham Lincoln suffered from Marfan syndrome, as did the virtuoso violinist and composer Niccolò Paganini.

#### Occurrence, etiology, pathogenesis

Among the orthopaedically relevant hereditary disorders, Marfan syndrome is relatively common, with an estimated prevalence in Great Britain of 11.2/million inhabitants [58]. Only isolated cases of the contractural form have been described [41]. The mode of inheritance is autosomal-dominant and the gene locus is 15q21.1 for the classical type and 3p25–24.1 for the contractural type (type II). The main problem is a structural deficit of collagen. The disease predominantly affects connective tissue, although all other tissue types formed from collagen can also show changes. The proteoglycan content of cartilage tissue is reduced compared to normal individuals (in contrast with the osteochondrodysplasias and mucopolysaccharidoses in which it is elevated).

#### Classification

The classical Marfan syndrome occurs in various forms. A *classical Marfan syndrome* exists if two or more primary symptoms are present. A *forme fruste* also exists in which secondary symptoms are accompanied by a maximum of one principal symptom [24]. A *contractural form* [41] is also distinguished. Ectopia lentis and the cardiac changes are lacking in this form, which is characterized by contractures in most of the joints, most markedly in the knee.

#### Clinical features, diagnosis

A Marfan phenotype is said to exist when spider fingers, an abnormally long arm span, increased ligament laxity and tall stature are all present at the same time. Although



■ **Fig. 4.97.** Clinical appearance of the legs of a 17-year old boy with Klippel-Trenaunay syndrome. Note the pronounced hypertrophy of the left leg and the large vascular nevi

typical, this habitus is not a confirmatory diagnostic factor. A reliable diagnosis of Marfan syndrome can be made only if at least two of the *primary symptoms* are present.

### Reliable diagnosis of Marfan syndrome:

#### Two primary symptoms

- ectopia lentis
- cardiac changes
- scoliosis

- **Habitus:** Patients with Marfan syndrome are unusually tall. Their striking appearance is attributable to the exceptionally long extremities combined with a normally sized, or even rather short, trunk. Full-grown patients are usually taller than 180 cm (5 ft 11 in). The ratio of trunk length (= height minus leg length) to leg length (= height of the symphysis) is much less than 1. The arm span is greater than the patient's height and, with the arms hanging down, the fingertips are below the middle of the upper leg. The fingers on the hands are abnormally long and thin, while the extremities as a whole are long-drawn out and appear rather delicate. All joints are excessively mobile. The thumb can be pressed against the forearm, and the finger, elbow and knee joints are hyperextensible. The face is long and thin with prognathism and a bulging forehead (dolichocephaly).
- **Primary symptoms:**
  - **Eyes:** A characteristic feature is the tremulousness of the lens (phacodonesis) of the ectopia lentis that results from the ligament laxity. This often leads to extreme myopia. Strabismus, cataract and retinal detachment can also occur.
  - **Cardiovascular:** Typical findings are aortic dilatation, mitral valve insufficiency, septal defects and aortic aneurysms. The aortic aneurysm is the commonest cause of death in Marfan syndrome.
  - **Scoliosis:** Scolioses occur in over 50% of patients with Marfan syndrome [24, 48]. These are often S-shaped, rigid and slightly kyphotic and progress relatively rapidly. In many cases they are associated with back pain.
- **Secondary symptoms:** A *funnel chest* is often seen in Marfan syndrome as a result of the abnormal growth in the length of the ribs. A *spondylolisthesis* may also occur. *Foot deformities* are often observed in the form of pronounced flat valgus feet, skewfeet or clubfeet [24]. The *acetabular protrusion* is present in a third of cases [9] and leads to premature osteoarthritis of the hip.
- **Radiographic signs:**
  - **Metacarpal index:** The ratio of the length of the 2nd–5th metacarpals to the width of the metacarpals at the level of the distal epiphyseal plate is increased.

- **Acetabular protrusion:** The CE angle according to Wiberg is greater than 40°.
- **Dural ectasia:** A very highly specific finding is the widening of the interpedicular distance in the lumbar spine, which is present in 95% of cases [1].

- **Differential diagnosis:** Marfan syndrome can only be diagnosed on the basis of the clinical findings, and there is no biochemical test as in *homocystinuria*, which is the most important condition to consider in the differential diagnosis since it can produce similar signs and symptoms (► Chapter 4.6.6.6). Marfan syndrome can also be confused with *Ehlers-Danlos syndrome* (► Chapter 4.6.6.7).

### Prognosis, treatment

Patients with Marfan syndrome are of normal intelligence. The life expectancy is shortened because of the cardiovascular complications and the commonest cause of death is rupture of the aorta.

**! No known treatment exists for the underlying disorder.**

#### Orthopaedic treatment:

- **Scoliosis:** Corset treatment is rarely successful for Marfan-related scolioses, but should nevertheless be tried in scolioses of more than 20°. While the corset may not be able to stop the progression, it may at least slow it down. Surgical correction is indicated if the scoliosis is 40° or more. If the pubertal growth spurt has not yet occurred (or is only just starting), the correction must be performed from both anterior and posterior approaches. If the peak of the pubertal growth spurt is passed, correction from the posterior or anterior side alone will be sufficient. The surgical technique is similar to that for idiopathic scolioses (► Chapter 3.1.11). Secondary curves should also be stiffened in case of doubt, otherwise they will readily decompensate [36]. It should be borne in mind that, while lordotic forms of scoliosis occur, a kyphotic component is often present. Surgery should not be delayed for too long since scolioses in patients with Marfan syndrome can very quickly become rigid and thus limit the options for correction. The complication rate associated with scoliosis operations is very high [23] (high blood loss 10%, infections 10%, injury to the dura 8%, implant pull-out 21%, pseudarthrosis 10%, decompensation 30%). Fatalities can also occur as a result of cardiac and aortic complications.
- **Clubfeet** are treated in the usual way. Even *severe cases of flat valgus foot* or *flexible flatfoot* are treated according to the same principles applicable to patients without Marfan syndrome.

- *Recurrent patellar dislocations* or *multidirectional shoulder instabilities* can prove particularly problematic. The therapeutic principles are identical to those applicable to other patients (► Chapter 3.3.5, 3.5.4).
  - In the *contractural form of arachnodactyly* physical therapy aimed at stretching the contractures is extremely important and successful.
- Knee contractures* can be corrected with the Ilizarov apparatus (► Chapter 3.3.13).

#### 4.6.6.6 Homocystinuria

This is an autosomal-recessive inherited disorder involving a cystathionine- $\beta$ -synthase deficiency (gene locus 21q22.3). As with Marfan syndrome, an ectopia lentis with myopia is often present, the extremities are long and the patient has spider fingers associated with tall stature. The ligament laxity, on the other hand, is not typical. If anything, joint mobility tends to be reduced. Intelligence is often diminished. Pes cavus, funnel chest and scoliosis are also often present. The diagnosis can be confirmed by measuring homocystine excretion.

#### 4.6.6.7 Ehlers-Danlos syndrome

This is a group of disorders involving impaired collagen synthesis and characterized by excessive general ligament laxity, skin changes such as scar formation – the skin may resemble »cigarette paper« –, fragility of the blood vessels and a slightly increased susceptibility to fractures. The condition was described by Ehlers in 1901 [10] and Danlos in 1908 [8]. Nine types with differing inheritance patterns are currently distinguished [49]: types I (gravis), II (mitis) and III (hypermobile type) are autosomal-dominant, type IV (vascular type) has a heterogeneous mode of inheritance, type V is X-linked, type VI (oculoscoliotic type) is autosomal-recessive, type VII (arthrochalis multiplex congenita) is heterogeneous, type VIII (periodontosis type) autosomal-dominant and type X (fibronectin defect) is autosomal-recessive.

The former classified type IX with abnormal ligament laxity is no longer described as Ehler-Danlos syndrome.

- The prevalence of the syndrome in Great Britain has been calculated at 26/1,000,000 inhabitants [58], although a large number of cases probably remains unreported since the condition (particularly its mild form) is often not diagnosed. A prevalence of 300/1,000,000 inhabitants is currently assumed for all types.
- The most striking *clinical* feature is the *hyperelasticity of the skin*. The skin can be pulled out from the cheeks to a highly abnormal extent. In older patients the eyelid falls over the eyelashes. The skin is very fragile and bruising readily occurs. In certain forms pseudotumors occur at particularly susceptible sites (elbow,

knees). The skin is hyperpigmented. Scar formation is abnormal, and the skin over the scar is very thin, resembling cigarette paper. The interdigital folds on the hands and feet are particularly good sites for testing the hyperelastic skin.

Most forms are associated with abnormal joint mobility with *general ligament laxity*: The thumb can be apposed to the forearm, while the fingers can be hyperextended to over 90° at the metacarpophalangeal joints. Hyperextensibility of the elbows and knees is also present. Flexible flatfeet are also very frequent.

Common problems are *habitual shoulder* and *patellar dislocations*. The shoulder dislocations are usually multidirectional. As well as being habitual, the dislocations can also be voluntary, i.e. the patients can actively contract their muscles to dislocate the head of the humerus on command. The same applies (though less frequently) to the patella. Shoulder and patellar dislocations are often bilateral.

Scolioses occur in approx. 50% of cases [40, 49]. These can develop at a very early stage, are strongly progressive and occasionally associated with kyphoses.

A particular problem is posed by *hip dislocations*. In the initial stages the dislocations cannot be distinguished from other forms of congenital hip dislocation associated with hip dysplasia. But hip dislocations in Ehlers-Danlos syndrome have a less favorable progression than in otherwise normal children. In particular, a well-reduced hip can deteriorate again after the child starts walking as a result of the abnormal ligament laxity. The femoral head may show a tendency to move off-center, adversely affecting the acetabular roof angle. Children with general ligament laxity require particularly close monitoring if hip dysplasia is also present.

- *Diagnosing* a case of Ehlers-Danlos syndrome is not always easy. The condition is hardly ever diagnosed before the age of four, and only then in patients with severe, clinically striking forms. It can often be difficult to make a clear diagnosis particularly for the mild forms (types II, III, VI). General ligament laxity is extremely common in children in any case, and the transition from normal to pathological is continuous. The possibility of Ehlers-Danlos syndrome should be considered in patients with voluntary bilateral multidirectional shoulder dislocations, bilateral habitual patellar dislocations or flexible flatfeet.
- *Differential diagnosis*: Type VII can be confused with *Larsen syndrome*, type IX must be differentiated from the contractural form of *Marfan syndrome*. Generally speaking, Marfan syndrome is the most difficult condition to distinguish from Ehlers-Danlos syndrome because of the hyperlaxity of the joints.
- *No specific treatment exists for Ehlers-Danlos syndrome*.



*Considerable restraint* is required in respect of *surgical orthopaedic treatment* for two key reasons: In the short and medium term there is a substantial risk of recurrence, particularly for soft tissue operations since all the tightened tissues will start to stretch again. In the long term, the prognosis is fairly good even without surgery since, as in all people, the ligaments tighten up with age in patients with Ehlers-Danlos syndrome.

**!** We would specifically advise against surgical treatment for multidirectional voluntary shoulder dislocation in Ehlers-Danlos syndrome, since no operation is capable of resolving this problem in children and adolescents (► Chapter 3.5.4).

*Conservative treatment* consists of a total ban on dislocating the shoulders voluntarily (which is often difficult for the patients in psychological respects since they can attract much attention with their ability to dislocate the shoulders voluntarily) and intensive and consistent muscle training exercises, e.g. according to the San Antonio program [7] (► Fig 3.493). With an intensive training program it is usually possible to gradually reduce the number of dislocations until the instability no longer represents a real problem. The dislocations pose less of a problem in adulthood in any case because of the general ligament contraction. Certain sports or occupations that place high demands on shoulder mobility should be avoided by these patients.

The *habitual patellar dislocations* (► Chapter 3.3.5) in patients Ehlers-Danlos syndrome should also be approached with caution. Although muscle training is less successful here than for the shoulder, it should be borne in mind that soft tissue operations are associated with a high relapse rate. The transfer of the tibial tuberosity is only possible in fully-grown patients and involves the risk of overcorrection and dislocation in the medial direction. Nevertheless, surgery is more frequently indicated for recurrent patellar dislocations than for those of the shoulder.

The indication for treatment must also be decided carefully in patients with flexible *flatfoot*. Since this is always associated with the increased ligament laxity, the treatment is no different than for the usual forms of flatfoot (► Chapter 3.4.9).

*Scolioses* and *kyphoses* are associated with a worse progression than the idiopathic forms. Corset treatment are not particularly successful and surgery is often required at an early stage [40]. It should also be noted that *atlantoaxial instability* can occur. On the one hand, this can represent a problem for any general anesthesia while, on the other, the instability can even be severe enough to require surgical correction (► Chapter 3.1.11).

#### 4.6.6.8 Silver-Russell syndrome

This rare syndrome is characterized by small stature, postnatal linear growth deficit, bodily asymmetry with hemihypotrophy, craniofacial dysmorphism with triangular facies, downturned corners of the mouth and clinodactyly of the fifth finger. A uniparental disomy 7 has been found as a genetic cause in some patients.

#### 4.6.6.9 Holt-Oram syndrome

This syndrome is an autosomal-dominant inherited disorder in which various skeletal malformations are present at the same time (e.g. shortening of the thumb, humerus, radius, poss. radioulnar synostosis, syndactyly, abnormal metacarpals) combined with heart defects. Abnormalities of the ribs may also be observed, and keeled chest and scoliosis can occur. Genetic studies have shown a defect of chromosome 12 (12q24.1)[5]. The heart defects can be severe enough to restrict the life expectancy of the patients considerably [32]. The syndrome is also known as »atriodigital dysplasia«. Any orthopaedic treatments must be evaluated in connection with the reduced life expectancy. Physical and occupational therapy are important [17].

#### 4.6.6.10 Poland syndrome

##### ► Definition

Disorder of unknown etiology (no hereditary component identified) characterized by the simultaneous occurrence of unilateral aplasia of the pectoral muscle and syndactyly with missing middle phalanges (symbrachydactyly). Other unilateral malformations are also occasionally present.

##### Historical background, etiology, pathogenesis, occurrence

The disorder was described in 1841 by A. Poland [42]. The etiology is uncertain, although it is probably caused by fetal oxygen deficiency. No hereditary component has been identified. The clearest indication that the disorder is not genetic was provided by a twin with the condition whose uniovular twin sister was not affected [50]. Poland syndrome is similar to Moebius syndrome (► Chapter 4.6.5.4). Both syndromes are based on damage to the embryo that occurs at the same stage of development (6th–8th week of pregnancy), but Moebius syndrome also involves the additional signs and symptoms of cerebral palsy. The incidence of Poland syndrome is approx. 3/100,000 births.

##### Clinical features, diagnosis

The most striking feature is the hypoplasia or aplasia of the *pectoralis major muscle* (► Fig. 4.98) and possibly the *pectoralis minor* as well. The nipple is hypoplastic or may



even be absent, hence the alternative description of the Poland syndrome as the *Amazon syndrome*. According to the legend, members of this ancient tribe of warrior women would cut off one breast so as to be better able to hunt with the bow. Occasionally the ribs are also hypoplastic. The *scapula* may be elevated and the shoulder mobility is often restricted. A *hand deformity* is almost invariably present (► Chapter 3.5.3). The right side is more often affected and the middle phalanges are usually missing, or else are hypoplastic.

The deformity of the upper extremity is classified according to its severity (■ Table 4.26).

### Prognosis, treatment

Patients with Poland syndrome have a normal life expectancy and do not usually suffer from any mental retardation. *Treatment* is based on functional criteria. Since the pectoral muscle is often not completely missing and the deficit can largely be compensated for by the pectoralis minor muscle, the functional restriction in this invariably

■ **Table 4.26.** Classification of deformities of the upper extremity in Poland syndrome

Severity	Clinical features
I	5 fingers present, poss. hypoplastic
II	Functional fingers on the ulnar and radial sides with absence of central fingers
III	Absence of functioning long fingers
IV	Radial ray defect with absence of fingers, incl. thumbs



■ **Fig. 4.98.** Clinical appearance of the upper body of a 13-year old boy with *Poland syndrome*. The pectoralis major muscle is missing on the right side, the right nipple is slightly higher than the left, the pectoralis minor muscle is present and is tensed

one-sided condition is usually not very serious. However, since the absence of the breast can be very upsetting for girls, plastic surgery is occasionally indicated.

The absence or hypoplasia of the central rays of the hand does not matter too much in functional respects provided opposition of the thumb and a sufficiently powerful pinch grip is possible. If this is not the case, surgical measures may be indicated. However, since the thumb tends to be missing only if the long fingers are also aplastic, pollicization is rarely possible. Lengthening operations on the forearm for cosmetic reasons are rarely indicated.

### 4.6.6.11 Hemophilia

#### Definition

Group of hereditary disorders involving impaired blood clotting. The commonest type, hemophilia A, has an X-linked mode of inheritance and exclusively affects male patients with factor VIII deficiency. The disorder is acquired through the mother, who acts as a carrier. The impaired blood clotting results in bleeding into the major joints (particularly the knees, elbows and ankles), which damages the synovial membrane, producing further bleeds and, ultimately, destruction of the joint.

— **Synonyms:** Factor VIII deficiency

#### Historical background

Evidence of the existence of hemophilia dates back to the biblical era and the Talmudic scriptures. In 19th century England, Queen Victoria was a carrier. She bequeathed hemophilia to the royal houses of Russia, Spain and Austria. The son (Alexei) of the last Tsar (Nicholas II) of Russia was a hemophiliac. The monk Rasputin managed to acquire great influence over the Tsarina, and thus accelerate the decline of the monarchy, thanks to his ability to stop the bleeds of the Tsarevich. The term »hemophilia« was coined by Hopff in 1828.

#### Occurrence, etiology, classification of the disease

The incidence is approx. 5–10/100,000 births (with a decreasing trend thanks to genetic counseling and prenatal diagnosis). We distinguish various disorders depending on the defect in the blood clotting system:

##### — Hemophilia A:

This is the commonest form, an X-linked recessive condition (gene locus Xq28). It involves a deficiency of factor VIII. Around 80% of hemophiliacs suffer from this form. Given its mode of inheritance, women are not affected by hemophilia A but can pass on the condition to their male offspring as carriers.

##### — Hemophilia B:

This condition involves a deficiency of factor IX, and its mode of inheritance is likewise X-linked recessive (gene locus Xq27.1–27.2). Around 15% of hemophiliacs suffer from this form, which is also known as »Christmas disease«.

#### — Von Willebrand disease:

This disorder involves defects in both factor VIII and platelets. Since the mode of inheritance is autosomal-dominant it can also affect females. As this form of hemophilia is relatively mild it causes few orthopaedic problems. Only 5% of hemophiliacs are affected by this form.

#### Clinical features

The clinical manifestations of hemophilia depend on the extent of the factor VIII or factor IX deficiency. If the plasma levels of these factors exceed 50%, the illness remains asymptomatic. The classification of the severity of hemophilia is presented in Table 4.27. As a rule, spontaneous hemorrhages occur only if the plasma level is less than 5%, while patients with a plasma level of under 1% are greatly at risk.

The bleeds occur mainly in the *joints*. The exposed major joints, i.e. the ankle, knee and elbow, are primarily affected. The hips and shoulders are rarely the site of bleeds. The severest changes are observed in the knee and elbow, while the lesions in the ankle, which is likewise frequently affected, are less likely to result in such a rapid onset of osteoarthritis [53]. In a severe case of hemophilia, a hemorrhage can occur in one of these joints even after a minor trauma. The bloody effusion remains in the joint and very rapidly leads to damage to the cartilage surface. As a result of the cartilaginous lesion, fluid can enter into the subchondral cancellous bone and form cysts. The blood deposits and the breakdown products of the joint cartilage cause further damage to the synovial membrane.

The hemorrhagic tendency is exacerbated by the inflammatory reaction of the synovial membrane. This produces a vicious circle of increasingly frequent bleeds, which can eventually occur on a daily basis. The joint undergoes further damage, resulting in subchondral irregularities, a narrowing of the joint space, osteophyte formation and, ultimately, in collapse of the joint. Severe osteoarthritis can develop as early as adolescence as a result of this process.

Hemorrhages can also occur in the *soft tissues*, particularly in the muscles, where large »pseudotumors« can form and also gradually erode the bone (Fig. 4.99). Large hematomas or pseudotumors can ultimately press on peripheral nerves and produce lesions. According to one study such nerve lesions prompted 81 out of 1,351 hospital admissions due to hemophilia [28].

Hemophilic joints have a higher risk of infection [15]. Another complication is the onset of avascular necrosis, particularly in the form of Legg-Calvé-Perthes disease. The patient is aware of the individual bleeding episodes and can usually state precisely when, and how frequently, they occur. Clinical examination of chronically altered hemophilic joints reveals both an effusion and a doughy swelling and thickening of the synovial membrane, which are distinguishable on palpation. Restriction of joint mo-

bility invariably forms part of the clinical picture of hemophilic arthropathy.

#### Radiographic findings

The development of hemophilic arthropathy is not quite the same as that of a degenerative arthritis. Subchondral sclerosis and osteophyte formation are less pronounced in hemophilic arthropathy, whereas cystic changes are more predominant. The radiological changes can be classified as shown in Table 4.28 (after Arnold and Hilgartner [3]; Fig. 4.100).



Fig. 4.99. CT scan of the pelvis of a 20-year old male patient with a huge hemophilic pseudotumor

Table 4.27. Classification of the severity of hemophilia

Plasma concentration of factor VIII/IX		Severity of hemophilia
25–50%	Factor VIII or IX	Mild hemophilia
5–25%	Factor VIII or IX	Moderate hemophilia
1–5%	Factor VIII or IX	Severe hemophilia
<1%	Factor VIII or IX	Very severe hemophilia

Table 4.28. Radiological changes in hemophilic arthropathy

Severity of hemophilia	Radiological changes in hemophilic arthropathy
I	Soft tissue swelling, no skeletal abnormalities
II	Osteoporosis of the epiphysis, joint integrity preserved
III	Slight narrowing of the joint space, subchondral cysts, widening of the intercondylar fossa in the knee
IV	Severe narrowing of the joint space with destruction of cartilage
V	Joint space obliterated, fibrous ankylosis and severe joint incongruity



Fig. 4.100a–d. The radiological stages of hemophilic arthropathy (in various patients). a Stage II, b Stage III, c Stage IV, d Stage V (see text)

### Differential diagnosis

The diagnosis of hemophilia is usually already known by the time the patient is seen by the orthopaedist, although the joint effusion is occasionally the first sign of the disorder. The most important condition to consider in the differential diagnosis is *juvenile rheumatoid arthritis*. Any doubt as to which of the two is present can be removed by means of a joint aspiration.

### Course, prognosis, associated disorders

Thanks to meticulous studies we are now very well-informed about the prognosis in hemophilia. One American study investigating 701 patients (between 1900 and 1990) found a 6-fold mortality rate for severe hemophilia, a 2-fold rate for moderate hemophilia and an unchanged mortality rate compared to the general population for mild cases of hemophilia. In the 1970's the life expectancy at the age of 1 year was 68 years. This had declined to 49 years in the 1980's as a result of the appearance of AIDS [22].

### Treatment

While initial *gene therapies* are available for hemophilia A [46], these are not yet used in everyday clinical practice. The *conservative treatment* in the event of an acute bleed primarily involves *replacement* of the deficient factor. This is the most important measure and prevents further bleeding due to the lack of coagulation. The affected joint can be temporarily *immobilized* during the acute phase, which also reduces further bleeding. Long-term immobilization must be avoided at all costs however. The application of *continuous passive movement* is more useful.

The value of *aspiration* is also disputed. Since the blood present in the joint damages the cartilage, aspiration would seem a sensible procedure. Even more effective is joint lavage.

Another conservative measure is *synoviorthesis* with osmic acid or radioactive substances [37], although the use of substances in children and adolescents with open epiphyseal plates is highly problematic.

### Surgical options for hemophilia

- Arthroscopic lavage
- Synovectomy (conventional, arthroscopic or using a laser device)
- Arthrodesis
- Arthroplasty

*Arthroscopic lavage* is useful for fresh bleeds, at a stage when chronic hemophilic synovitis has not yet developed. In such cases, joint lavage and coagulation of the bleeding vessel may be able to prevent the development of hemophilic arthropathy. *Synovectomy* is much more successful in patients with hemophilia than in the treatment of juvenile rheumatoid arthritis [14, 25]. The use of a laser device is particularly beneficial for synovectomy in hemophiliacs since the laser beam produces better hemostasis compared to cutting with a normal scalpel. In fact, the number of replacement units transfused during and after the operation can be reduced by up to a half thanks to the improved hemostasis. Last but not least, since the replacement units are very expensive, savings of up to US \$ 6,000 per patient can be achieved. Synovectomy is a worthwhile operation in patients with severe hemophilia and hemophilic arthropathy in stages II or III.

In contrast with the situation in juvenile rheumatoid arthritis, the synovectomy should not be delayed for too long in hemophilia. Once the arthropathy has progressed to the state where the joint is largely destroyed, the synovectomy will no longer be able to improve the condition.

In this case, the surgeon will have to decide whether to perform an arthroplasty or (as an alternative) an arthrodesis. Such operations are occasionally required in young patients [29, 34].

#### 4.6.6.12 Hypothyroidism

Hypothyroidism either occurs as a congenital condition or is acquired as a result of a goiter. In former times, hypothyroidism used to be common in certain regions due to a chronic iodine deficiency. Since iodine is now added to the water or salt in these areas, hypothyroidism has become very rare. The illness occurs three times more frequently in girls than in boys.

— In *clinical* respects, severe forms of hypothyroidism are characterized by dwarfism and severe mental retardation. The condition was formerly known as *cretinism*. In the infant, congenital hypothyroidism should be suspected if overweight, prolongation of physiological icterus, somnolence, flaccidity, loss of appetite and chronic constipation are present. Other signs are dry skin, an enlarged tongue, expressionless face and bulging abdomen. The children learn to sit and stand at a very late stage.

*Skeletal maturation* is greatly delayed. Specifically, the appearance of the epiphyseal ossification centers is delayed («epiphyseal dysgenesis»). The epiphyseal plate is often irregular and widened. The children have a relatively large head compared to their body length. The closure of the fontanelles and cranial sutures is delayed. Typical changes are observed in the spine: The second lumbar vertebral body (and possibly the adjacent ones as well) is usually abnormally wedge-shaped. Lumbar kyphosis with spondylolisthesis may also form.

The *acquired forms* of hypothyroidism are usually less obvious. The signs and symptoms vary according to the age at their initial onset. The most important sign of acquired hypothyroidism is delayed bone maturation. The epiphyseal plates are weakened, hence the increased incidence of epiphyseal separation [55]. Because of the delayed bone healing, this can occur at an age when growth is normally concluded. If an epiphyseal separation occurs in a girl older than 13 years or a boy older than 15 years, the possibility of hypothyroidism should always be considered.

— *The treatment* of hypothyroidism consists of thyroid hormone *replacement*. An adequate supply of iodine is important. If the hypothyroidism is diagnosed in good time, the typical complications can be avoided. The most important *orthopaedic complication* is *slipped capital femoral epiphysis* (■ Fig. 3.194, ► Chapter 3.2.6), which is managed in the usual way. Not infrequently, it is the orthopaedist who first suspects a secondary hypothyroidism when an epiphyseal separation occurs in association with severely delayed maturation.

#### 4.6.6.13 Sotos syndrome

This disease, which was described by Juan Sotos in 1964 [47], is characterized by substantially accelerated growth in the first years of life. The syndrome usually results from new mutations, the gene locus is 5q35. The uncoordinated accelerated growth is associated with cerebral gigantism, typical facies, a height at the 97th percentile, mental retardation and, secondarily, the occurrence of spinal (scolioses, kyphoses) and thoracic deformities (funnel chest). The orthopaedic treatment focuses on these deformities. The syndrome also appears to be associated with an increased frequency of malignant tumors [35].

The Sotos syndrome has similarities with the Weaver [54] and Beckwith-Wiedemann syndromes, which are likewise associated with accelerated growth [6, 57]. A specific problem of the latter disease is the macroglossia, which may require surgical treatment. Scolioses and thoracic deformities also occur in these two syndromes.

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## 4.7 Neuro-orthopaedics

### 4.7.1 Basic aspects of neuromuscular diseases

R. Brunner

#### Definition

Neurological diseases, which interfere with the control and functioning of the muscles, lead to functional disorders and structural deformities of the musculoskeletal system. Since the underlying neurological problems cannot usually be resolved at causal level, they act permanently on the musculoskeletal system. Since the growing skeleton is more plastic than the fully-grown counterpart, secondary skeletal deformities occur particularly during childhood. These can further aggravate the functioning of the locomotor system. Neuro-orthopaedics is concerned with the consequences of neuromuscular disorders on the musculoskeletal system.

#### Historical background

Even at the start of the last century appliance-based treatment for cases of paralysis, often after poliomyelitis, together with procedures for improving function, formed an important part of orthopaedics as a whole. Nowadays, patients with the sequelae of poliomyelitis are rare. Perinatal provision has improved significantly, hence the almost complete disappearance of kernicterus and the greater rarity of mild forms of cerebral palsy. The emphasis has now shifted toward the treatment of severely disabled patients, whose numbers have not declined. Such patients benefit from the latest techniques of anesthesia and surgery, enabling even those in a poor general condition to undergo the usually major and complex operations required. Adequate experience is a crucial basis for the often difficult and functionally relevant therapeutic decisions for the optimal treatment of the individual patient.

#### Etiology and pathogenesis

Widely differing clinical conditions lead to neuro-orthopaedic problems and these are addressed in the individual chapters (e.g.: *cerebral cause: cerebral palsies, craniocerebral trauma, infections, tumors; spinal causes: paraplegia, myelomeningoceles, post-polio syndrome; peripheral disorders: nerve lesions, plexus palsies etc.*). These neurological conditions cannot usually be treated at the causal level. Since the resulting functional *orthopaedic problems* are more uniform than their causes they will be grouped accordingly. The loss of control over part of the motor system affects everyday functions such as walking, standing, sitting or the use of the upper extremity. The underlying muscle activity may be spastic or flaccid.

A sensory disorder of varying severity is also usually present and can indirectly affect everyday functioning. Thus, impaired proprioception can lead to a loss of dy-

namic control over the joints and thus to an overloading of the ligamentous structures with progressive instability. Occasionally, whole extremities are not used because of sensory problems. The motor and sensory systems influence each other: The less an extremity is used, the less the sensory functions develop, and the less the extremity is used again. Motor training is important therefore for the development of sensory functions.

Physiologically the *muscles* adapt to the everyday needs in terms of power development and operating length. At the same time, stretching during everyday life ensures that the muscles are long enough to preserve adequate mobility of the joints. If high heels are worn constantly, for example, the triceps surae muscle is shortened, thereby restricting dorsiflexion. Spastic and flaccid pareses and the necessary compensatory mechanisms change the loading on the muscles, which then exert power in positions that differ from the physiological situation in healthy individuals.

The use of muscles with modified lengths and forces and the presence of spasticity interfere with their extension in everyday life. The muscles adapt to the new situation. Some muscles become too short (contracted), whereas others are too long, a situation that alters the extent of joint movement. Moreover, the optimal operating length for both muscles in relation to the optimal position within the range of motion is shifted, resulting in a weakness of both muscles. The still growing skeleton also adapts itself to the modified situation resulting from spastic and outwardly uncontrolled forces, with consequent secondary deformities. The aim of conservative and surgical measures is to prevent and correct these consequences. These secondary changes, in turn, often interfere with function and represent an additional handicap for the patient.

On the other hand, certain changes can prove functionally beneficial, for example an equinus foot in a case of a weak triceps surae muscle. The orthopaedist must be able to recognize and preserve such changes, and guard against therapeutic overzealousness and inappropriate treatment.

**! One important diagnostic step in neuro-orthopaedics is to distinguish between functionally beneficial and disruptive changes of the musculoskeletal system.**

Various principles can be drawn up for orthopaedic treatment that are based more on that signs and symptoms and functional consequences of the underlying disease rather than the actual basic neurological condition. Excessive spastic and weak, or absent, muscle activity are responsible for the two basic symptom complexes. Since the underlying neurological disease often cannot be influenced, or only influenced to a minimal extent, the same pathological mechanisms continue to act even after

treatment. This explains the high rate of recurrences after corrective procedures. Treatment invariably involves a variety of different measures involving, for example, the group of »therapies« (physical therapy, occupational therapy, speech therapy etc.), braces, operations and possible drugs as well. The numerous therapeutic strategies must be implemented in a planned and coordinated manner. As a rule, however, none of these measures is capable of solving the problem definitively.

**! Neuro-orthopaedics is concerned with the treatment of structural and functional changes of the musculoskeletal system that occur secondarily as a result of a neurological disorder. However, since the underlying disease is not treatable, or at least only treatable to a minimal extent, no definitive correction should be expected from the orthopaedic treatments in the long term.**

### **Clinical features and diagnosis**

The clinical evaluation of a patient with neuromuscular disease must always include a neurological assessment in addition to an orthopaedic examination.

### **Neurological evaluation**

Motor and sensory disorders are of particular interest from the neurological standpoint and must be included in the orthopaedic treatment plan. As regards *motor function*, a basic distinction must be made in connection with orthopaedic measures between neurological disorders with reduced, increased or altered muscle activity.

*Reduced muscle activity* and power are present in flaccid paralyses, e.g. after damage to a peripheral motor nerve, after poliomyelitis or in spinal muscle atrophy, but also after a muscle itself is damaged, for example as occurs in muscular dystrophies or other myopathies.

Testing the power of the individual muscle groups will reveal any muscle imbalances. During everyday activities, the body must be stabilized dynamically by the muscles so that individual parts of the body can move and be used in a targeted manner. Failures of the dynamic stabilizers and organs of propulsion explain the functional restrictions of the patients. Muscle tone – compared to healthy individuals – may be reduced overall or only in individual parts of the body. Such muscle hypotonia occurs, for example, during the period immediately following injuries to the CNS, or can often affect the trunk in otherwise spastic cerebral palsies. The postural function of the dynamic stabilizers in this patients is also often inadequate, although voluntary muscle activity is still possible. Thus, while a patient may be able to sit up voluntarily, he will otherwise sag down in his chair when seated.

*Increased muscle activity* occurs in the form of muscle hypertonia and spasticity. Muscle hypertonia manifests

itself as a stiffness that hinders joint movement in the relevant body sections in all directions. However, the range of motion is hardly restricted at all. Spasticity, on the other hand, involves an increase in muscle tone with exaggerated muscle reflexes. As a rule, only individual muscle groups are affected, and the antagonists are overstretched and therefore functionally inadequate. Spastic muscles are also weakened under the effect of their spastic power. Spasticity does not train the muscles!

A typical finding on clinical examination is the need for the application of a lot of force in order to break the spasm and continue the movement in the same direction. While the detailed pathogenesis of the spasticity is not clear, it is thought to be associated with an increase in gamma activity that makes the muscle spindles more sensitive, thus resulting in exaggerated muscle tone and reflexes.

In everyday clinical practice, the marked tendency toward muscle contractures, in particular, can cause problems. The excessive muscle activity can be triggered or avoided according to the positioning of the patients. Thus, an extension spasm in the leg can be elicited by stretching at the knee, while a flexed position will prevent the extension spasticity. Defective neuromuscular control in the upright position will lead to dynamic instability. Muscle tone increases by way of compensation, but this has a negative impact on tone stability.

*Any alteration of involuntary muscle activity* produces motor signs and symptoms that cannot be controlled directly by the patient. These include dystonia, athetosis and, in a wider sense, ataxia. In dystonia, individual muscles or muscle groups produce sustained tonic contractions. Athetosis is characterized by involuntary, irregular and slow movements that can cause extreme positions to be adopted at the joints, which keeps contractures at a low level. In ataxia, it is the coordination of muscle activity that is impaired, causing the patient to stand and walk unsteadily or to grab past objects.

A knowledge of the *sensory changes* is also important for orthopaedic concerns. Hypoesthesia or anesthesia predispose toward poorly healing pressure ulcers. But they also interfere with motor learning, since the necessary sensations are not perceived or incompletely perceived. By contrast, hyperesthesia is troublesome during the use of the affected extremity. For example, the patient may be unable to tolerate shoes on the feet or try to avoid any contact with the ground. Here, too, motor development is adversely affected.

### **Orthopaedic evaluation**

The main diagnostic aspects are described in the corresponding chapters on the individual clinical conditions. In any neurological disorder or functional and structural deficit, the orthopaedic problems must be assessed in respect of any functional impairment of the patient. Ac-

cordingly, this section will confine itself to a few general comments on the examination and functional evaluation of patients with neurological disorders.

**! In order to be able to stand with minimum muscle activity, we require slight hyperextension at the knees and hips. During normal walking as well, the joints are stressed almost to these extreme positions. Only the triceps surae muscle needs to perform postural work in order to produce a stable standing position. If this muscle is insufficient, a structural equinus foot can take over this task.**

Functional improvement often means enabling the patient to use more external forces without having to expend so much muscular effort.

### Muscle contractures

The search for *muscle contractures* forms an important part of the neuro-orthopaedic examination. Contractures lead to a restriction of the range of motion in the affected joints. The examination must be performed slowly and steadily in patients with spasticity, since sudden movements can trigger spasms which can then be mistaken for muscle contractures. Muscle tone and spasticity also depend on the position of the body, while primitive reflexes (asymmetrical tonic neck reflex etc.) influence the clinical picture [30]. Nevertheless, the mobility of the joints can be tested conclusively by patient examination. In our experience, examinations under anesthesia are *hardly ever* required.

In patients with muscular dystrophy, the possibility of pain must be taken into consideration in addition to the actual tensing of a muscle since, while patients will certainly feel the muscle stretching, the reflex muscle tensing observed in individuals with healthy muscles is not possible. Consequently, even slight stretching is painful.

At the *hip* level, shortening of the flexors is compensated for by hyperlordosis since, when the patient is lying down, gravity forces the legs downward, producing anterior tilting of the pelvis. If the other leg is flexed to its maximum extent, the pelvic tilt and compensatory hyperlordosis are cancelled. The thigh of this other leg then raises itself from the examination couch, thereby demonstrating the presence of a flexion contracture. Full extension can be examined with the patient in the supine position with the legs hanging freely over the end of the couch. The length of the knee flexors (hamstring muscles) can readily be evaluated by flexing the leg at the hip by 90° and then stretching the knee out of the flexed position. The angle at the knee serves as a measure of the contraction of the knee flexors. Alternatively, the extended leg can be lifted off the couch and the maximum flexion at the hip measured. A flexion contracture of the knee, by

contrast, leads to deficient knee extension even when the hip is extended.

The rectus femoris muscle may also be shortened. This shortening is tested with the patient in the prone position. The knee is flexed with the hip extended. Any shortening of the rectus femoris is manifested by a raising of the pelvis when the knee is flexed (Duncan-Ely test).

The *ankle* is examined in a similar manner: To enable the length of the triceps surae muscle to be determined, the foot must be adducted and supinated in an equinus position. This locks the lower ankle. The maximum dorsiflexion with the knee in a flexed position indicates the length of the soleus muscle. When the knee is extended, dorsiflexion is reduced because the gastrocnemius muscles tense. Finally, dorsiflexion at the ankle can also be tested with the lower ankle in a free position. This shows the functionally relevant dorsiflexion despite a possible shortened triceps surae muscle with an overextended lower ankle.

### Skeletal deformities

As well as measuring the range of motion in the joints and muscle contractures, the orthopaedist must also ascertain any *skeletal deformities*. The patient's position as he or she enters the office is particularly informative. The gait pattern is not usually consciously corrected and abnormal trunk postures are also still present.

The spine is examined with the patient in a sitting or standing position. The correctability of any deformity is checked while the patient is erect or lying down. At the hip, just shifting the range of motion in the direction of internal rotation will reveal any increased anteversion. A more accurate measure can be obtained clinically, with the patient's hip extended, by measuring the angle between the flexed lower leg and the vertical in the position in which the greater trochanter shows maximum lateral protrusion on palpation. A clinical parameter for the torsion in the lower leg is the angle between the knee axis and the malleolar axis with the knee in a flexed position.

### Functional examination

Any examination should, insofar as possible, include the testing of *functions* such as walking, standing or sitting in order to establish the functional consequences of the observed structural changes. The analysis of walking in everyday clinical practice is essentially based on the principles of gait analysis. While a basic clinical examination is usually sufficient for a general assessment of simple problems, a gait analysis is useful before radical therapeutic measures as this is the only way of determining the nature and extent of the functionally disruptive changes – together with details that are important for the treatment (► Chapter 2.1.3). In addition to the orthopaedic

examination, a short neurological assessment is also required in order to be able to incorporate the direct effects of the underlying disease on muscle tone and neuromotor development in the therapeutic strategy.

Hopping on one leg serves as a basic neurological test of *coordination* and *balance function*. Standing on one leg for a fairly long period involves a higher degree of difficulty. These two tests can provide a rough assessment of neuromotor control. Useful criteria for the orthopaedic evaluation of neuromuscular disorders are listed in ► Chapter 2.1.2. As well as checking leg length and balance of the *standing* patient, the examiner also assesses whether the hips and knees can be extended sufficiently and whether both legs are weight bearing. Patients with sitting problems of contractures in particular must be examined while *seated*. Ideally, the patient should sit on the examination table for this purpose. The orthopaedist can now test whether the patient is able to maintain this position independently or how much additional external help with stabilization is required.

Any necessary adaptations to seating aids can be clarified at the same time: First the pelvis is aligned horizontally without rotation, and the legs are spread apart as loosely as possible. The trunk is aligned as straight as possible over the pelvis in this position. This is a simple way of showing the extent to which movement restrictions render a symmetrical sitting position impossible and the extent to which these must be taken into account in connection with any seating aids. Examination of the patient in the *lying* position should not be forgotten, since these patients may lie for relatively protracted periods during the day, and posturally-related deformities are common.

Full extension at the hips and knees is required for lying stretched out in a supine or prone position. A flexion contracture at the knee subsequently leads to flexion at the hip. The leg is thus drawn up and falls inward or outward depending on the muscle tone in each case. The blanket or quilt also exerts a long-term »corrective« force through gravity. Flexion contractures of the hip produce the same effect, but in this case supplemented by a compensatory hyperlordosis of the lumbar spine. In the prone position the hip is always in approximately 10° flexion as a result of the conical shape of the thigh. Full extension can be achieved only by placing a support under the thigh close to the knee.

In the supine position, on the other hand, the hip is extended when the knees are stretched. Depending on any existing deformities, positional aids may be needed to prevent the development of compensatory deformities. Simple tests reflecting important everyday situation must be performed to evaluate the function of the *hand*. These examinations are very time-consuming and require experience. We therefore entrust this task to the occupational therapy department and discuss the resulting findings together with the patient.

Existing deformities must be assessed in respect of their *functional significance*: Thus, a contracture of the triceps surae muscle can compensate for a paralysis of the muscle, or a position of internal rotation of the legs in patients with poor postural function can be useful since such patients only sink towards the floor to the point where the knees press against each other, thus enabling them to stand in a stable position. If the legs were straightened they would drop to the floor.

A similar situation also applies to walking with internally rotated legs: If the knees give way, they do so in the direction of walking. But if they give way when pointing straight ahead or outwards (as in patients with legs that point straight ahead or outwards), the patients must balance their upper body over the poorly controllable legs in order to restore their equilibrium. This gives rise to a trunk-swinging or Duchenne limp that cannot be improved by treatment. On the other hand, a pronounced internal rotation during walking can be troublesome if the knees knock against each other or if the feet are almost at right angles to the direction of walking and catch on each other. Actual forward propulsion is no longer possible and walking is hampered. These examples demonstrate that the aim of treatment is not a »normal« configuration of the musculoskeletal system, but rather one that is best for the patient.

Functionally significant deformities must be differentiated from *cosmetic aspects*: An internal rotation of the legs can also simply be esthetically unappealing without any functional impairment. Hemiplegic patients may dislike their typical hand position with pronation, flexion and radial duction, in which case a surgical procedure may be indicated for cosmetic reasons. However, a functional deficit must be ruled out in connection with any surgical correction. By contrast, a severe foot deformity may be of no functional relevance if weight is not placed on the foot during everyday life. In these cases the deformity is merely of a cosmetic nature, and the patient's wish to wear standard shoes constitutes one of the few indications for treatment.

### **Therapeutic measures**

These patients often require treatment over a protracted period, and not infrequently for life. The costs are considerable, and the treatment time-consuming for all those involved. The treatment sessions (physical therapy, occupational therapy, speech therapy etc.), particularly in the case of children, take place once or twice a week. Braces are often expensive and must be worn and renewed regularly. Surgical treatments result in enforced bedrest. Functionally beneficial and troublesome deformities and compensatory mechanisms must be identified and differentiated if the patient is to receive the best treatment. For these reasons, a meticulous investigation of the patient's functions is essential and must include sensory aspects,



cooperation, coordination, intellectual abilities and concomitant neurological symptoms (for example epilepsy, athetosis).

Therapeutic mistakes have particularly serious consequences for the patient, since the potential for compensation is already used up as a result of the underlying neurological condition and the mistakes often end up with functional deficits that are no longer correctable.

The age of the patient was once considered to be a significant factor in the planning of therapeutic measures. But we now believe that this aspect should not be overrated. It can be beneficial to provide treatment for a functional deficit that hampers the patient's progress and accept the risk of a recurrence and the need for further surgery. Leaving a functional problem untreated can block the patient's progress and may result in the need for much more extensive correction at a later date. If a deformity is corrected at an early stage, the duration of the rehabilitation phase can be predicted. But if the surgeon delays surgery and the deformity worsens, the time required for rehabilitation increases disproportionately and can ultimately involve several years. During this time, the patients may become discouraged, with the consequence of poor results or complete relapses.

There are also limits on the correctability of deformities: If a muscle contracts, its antagonists become overlong. Correction of the contraction may result in a permanent functional insufficiency of the elongated muscles, particularly if the deformities are pronounced and have existed for a long time. The ligaments and capsular structures also adapt to the new situation. As well as a lengthening of the contracted sections, the surgical correction would have to include a shortening of the elongated sections, which is not usually performed and not always even possible in practice. Early corrections are therefore beneficial, and fixed age limits are not a good argument against such interventions at an early stage.

The *therapeutic objectives* that we wish to achieve for our patients can roughly be divided into two main groups:

- *Severely disabled patients* should primarily be able to lead pain-free and comfortable lives. Therapeutic measures may also be required to facilitate nursing care. One occasionally very ambitious objective is the achievement of transferability. This means that patients can stand, at least temporarily, on both legs and bear their own weight, even if they may need to be held in that position in order to compensate for any lack of balance. The patients' ability to bear their own weight simplifies nursing care considerably, and may enable them to live in a residential home – with a correspondingly enhanced quality of life (compared with a nursing home or a geriatric ward). Although major interventions are sometimes required to achieve such

small steps, experience shows that the effort is worthwhile.

- The objective for *slightly disabled patients* is to improve everyday functions (walking, sitting, standing and the functioning of the upper extremities). This will promote the patients' independence with the aim of enabling them to lead a largely independent life and possibly even to take up employment. Walking, as an important function of the lower extremities, can best be investigated by means of a gait analysis. This type of investigation often reveals important details that must be considered in the treatment planning.

### Prophylaxis

Although neurological changes cannot be prevented, measures to counteract the secondary deformities are useful. Impaired neuromuscular control regularly leads to a dysfunction in the locomotor system, thereby altering the loading of the skeleton and soft tissue which, in turn, produces secondary changes. These may be present only during a specific function (i.e. walking, standing, sitting etc.) or at rest without any loading (functional or structural deformities). Since these secondary changes can further restrict the patient's functions, prophylactic measures, whether surgical or conservative, are of major clinical significance.

*Physical therapy* plays the most important role in the prevention of deformities and is designed to preserve the correct length and strength ratios of the muscles and thus create the ideal preconditions for the proper functioning of the musculoskeletal system. In this way the mobility of the joints can be preserved in all planes. In parallel, the patient is given practical guidance in relation to the nervous system (tactile stimuli, position changes, balance training etc.), specific attention being paid to the individual deficits. Skeletal deformities, on the other hand, cannot be prevented by physical therapy.

*Orthoses* can also be used prophylactically. Thus, *lower leg braces* can prevent foot deformities or *trunk supports*, if used at the right time, can help prevent spinal deformities. Rotational problems however cannot be corrected by orthoses. Certainly a regularly acting rotational force, e.g. during the heel-to-toe roll while walking, appears to have a negative effect on the leg segments. *Twister cables* can be helpful in cases with excessive malrotations. These cables are fixed between a lower leg brace and a pelvic ring and exert a graduated rotational force on the brace that counteracts the torsion produced during the heel-to-toe roll. Although they cannot correct the deformity the braces can serve as a functional aid until the torsion is surgically corrected.

**! Existing skeletal deformities can only be corrected by surgery.**



## Treatment

### Orthopaedic measures for neuromuscular symptoms

All neuromuscular disorders lead to a lack of muscle control at the joints, resulting in dynamic instability. The aim of treatment, therefore, is to obtain maximum passive stability (by full extension or slight hyperextension at the knee and the hip) and to optimize the efficiency of the remaining muscle activity by correcting all lever arm deformities (feet and knees should point in the direction of gait). The ankle needs to be stabilized actively at 90°, or passively by muscle shortening at the same angle in the case of absent muscle activity. The deformities impeding function are best diagnosed by gait analysis. These should all be corrected at the same time, which often involves multilevel surgery, irrespective of the underlying neurological disease.

### Muscular insufficiency

Muscular insufficiency can result from a flaccid paresis, a muscle disease or muscle hypotonia. It should be borne in mind that a spasticity can also conceal a muscle weakness, which comes into play when the spasticity is efficiently treated. Innervated muscles can be built up by power training in children as well, although this requires regular controlled training and a motivated and cooperative patient.

Otherwise, from the therapeutic standpoint, only the deficit of muscle activity and power can be offset by *stabilizing measures*. Braces that firmly grasp and bridge the affected joints during functional use, i.e. walking and standing, are particularly suitable for this purpose. Alternative options are surgical stabilization procedures such as arthrodesis or tenodesis. Modern instrumental three-dimensional gait analysis is especially helpful in the diagnosis of functional muscle weakness.

### Excessive muscle activity

Excessively strong muscle activity such as spasticity or general muscle hypertonia interferes with the joint mobility of the affected extremities and can also lead to stiffness of the whole body. The patient is almost locked inside his own body as a result of the spasticity. Particularly spastic muscles have a strong tendency to produce contractures, in contrast with dystonic or atactic muscles. These problems disrupt the coordinated sequence of movements, prevent the interplay between the antagonists and agonists and therefore interfere with everyday functions such as sitting, standing, walking and the use of the upper extremities.

The disruptive excessive muscle activity can be curbed to a certain extent by appropriate *positioning* of the patient and his limbs. The spasticity usually affects one direction of movement at the joints, often extension in the legs and flexion in the arms. Positioning the legs in flexion while the patient is seated produces a looser position that also

facilitates the use of the proximal extremities. The spasms cannot be suppressed completely however. Thus, spastic reactions can be triggered repeatedly, for example even by emotions.

The muscle tone can be reduced, however, by stabilizing the patient in an upright position (with molded seats, standing aids or orthoses). This also improves the patient's voluntary motor control as the loosening of the spasticity restores the patients freedom to control his or her own muscles and use the limbs, particularly the arms and hands.

But such simple conservative orthopaedic measures are not always enough for preventing spasticity and increased muscle tone. Deciding on the actual treatment of increased muscle tone and spasticity, however, poses a difficult problem: The basic options are physical therapy, drugs and surgical treatment.

As regards *physical therapy*, stretching and rhythmic movements (e.g. as in hippotherapy or during cycling) produce positive effects. Other options include *manual* and *atlas therapy*. Although these methods result in a reduction in muscle tone [25], since they only produce a short-term effect, the treatments often have to be repeated. Unfortunately, there are only a few corresponding centers with trained and experienced personnel, particularly for children, and this considerably restricts the application of these methods.

Muscle tone can also be influenced by *drug treatment*, primarily with diazepam (Valium), baclofen (Lioresal) and tiznidine (Sirdalud). Certain antiepileptics (e.g. phenytoin (Epanutin)) can also produce positive effects. Diazepam (Valium), in low doses, generally loses its effect after a few weeks at the latest and is therefore particularly suitable for temporary tone control, e.g. postoperatively. In our experience, tiznidine (Sirdalud) is not very effective for spastic cerebral palsies. Baclofen (Lioresal), on the other hand, is also suitable for the long-term treatment of spasticity, although it is contraindicated in patients with uncontrolled epilepsy. It is generally administered by mouth initially (0.3–2 mg/kg body weight).

In most cases however, it subsequently has to be discontinued because of the major side effects, particularly fatigue, and to a lesser extent depression. In such cases, intrathecal administration via an implanted programmable pump represents one possible alternative. While the generalized side effects can be reduced considerably [8, 33], complications such as catheter dysfunction, leaks or infections are not infrequent, occurring in approx. 10% of cases. Finally, the pump must be changed after 5–7 years.

Before the pump is implanted definitively in the abdominal area, either subcutaneously or subfascially, a test treatment is administered. This can either involve an intrathecal bolus injection of baclofen, or else the post-

operative situation can be simulated with an external pump and an implanted catheter. This method appears to be particularly suitable for severely disabled patients, producing a positive effect on the arms. Unfortunately, the implanted pumps are still relatively large, thus restricting their use. Furthermore, since few parents and patients are willing to accept the implantation of a pump, our own experience with this method remains limited [1, 9, 27].

*Botulinum toxin (Botox, Dysport)* can switch off the motor endplates by irreversibly suppressing the release of acetylcholine and thus inactivating the relevant muscle. This method is used for individual muscles or muscle groups. The muscles that are affected by spasticity can best be identified by instrumented gait analysis. The therapeutic plan prepared on the basis of the gait analysis can then be checked after injection using the same diagnostic equipment. If the treatment proves successful, the botulinum toxin injection can either be repeated or the relevant muscle can be lengthened with the aim of making it weaker. This is one way of postponing a scheduled operation without the onset of additional deformities.

Another indication for botulinum toxin treatment is painful muscle hardening or insertion tendinosis. The drug can also be injected into muscles whose tendons are surgically treated. Since the effect roughly lasts for the period a tendon takes to heal, further protective measures are not required.

This botulinum toxin treatment has now become an indispensable part of the modern management of cerebral palsy. It has also been suggested that, if employed at a very early age (2 or 3 years), pathological compensatory mechanisms will not even develop in the first place, thus enabling better functional results to be achieved at a later stage through efficient early treatment of the spasticity.

The correct location of the needle in the desired muscle must be checked before the drug is injected, either by a functional movement test (accompanying movement of the needle), or by checking the position by EMG or ultrasound. No more than 50 U of Botox per injection site should be administered. The dose required for each muscle differs depending on the respective muscle, the age of the patient and the preparation. Appropriate tables should be consulted. The specified maximum dose is currently 30 U Botox/kg body weight. However, since general signs of weakness can even occur at this level, we avoid using such a high dose and do not exceed a dosage of 25 U Botox/kg body weight. The injection can be administered under local anesthesia, sedation or, if extensive, under a general anesthetic.

We administer a general anesthetic for the injection of very deep muscles (e.g. the iliopsoas) or if more than 8–9 needle pricks are required. The patient's sensation

is preserved and the effect can be graduated, with an onset after 12–72 hours and lasting for between 3 and 6 months. The injections may be repeated, and no negative consequences have been reported to date as a result [4, 13, 32, 38].

*Posterior rhizotomy* is another method for reducing muscle tone [31]. In contrast with the baclofen pump however, this is restricted to the lower limbs. In this technique, the surgeon performs an extensive laminectomy, selectively divides, as far as possible, afferent fibers of the extended reflex arc and then replaces the lamina. Although this procedure not only reduces muscle tone but may also affect muscle power, and can also lead to sensory problems in some cases, the reduction in spasticity does improve everyday functions such as sitting, standing and walking. On the other hand, various side effects of this treatment have been reported that are not inconsiderable, for example general weakness, heterotopic ossification [29] or progressive hip dislocation [17, 20, 26]. Although we ourselves have had very limited experience with this method, it should definitely be included in the repertoire of treatments for achieving efficient tone control, particularly if the problems are concentrated on the lower extremities [36].

Another way of influencing spasticity is to slacken the muscle and thus reduce its force. This can be achieved with a *tendon lengthening* or *aponeurotic lengthening* procedure. Tendon lengthening is a popular method in many places, particularly for the Achilles tendon. While this surgical procedure results in a permanent loss of power when performed on normal muscle, the power declines only temporarily after the aponeurotic technique and subsequently recovers completely [7].

Our experience in the hospital and gait laboratory has shown that while the spasticity is reduced by the tendon lengthening operations, the loss of power can lead to functional problems. On the other hand, the lengthening of a contracted muscle can restore the optimal muscle length at which the muscle produces its maximum force, so that it again acts within the correct range of motion of the joint. The muscle can even become more powerful, which explains the contradictory results reported after tendon lengthening procedures [11, 12, 19, 28, 39]. To achieve a reduction in power the tendon must be lengthened excessively. This procedure can be useful for individual muscles (e.g. for the posterior tibial muscle in neurogenic clubfeet). A prior test injection with botulinum toxin will help avoid any incorrect indications.

Spastic muscles can be temporarily disabled for differing periods (from a few hours to months) by means of *nerve blocks* involving the instillation of local anesthetics, phenol or alcohol (95%) [18, 24]. If only the motor sections of the nerves are infiltrated, this method is a good alternative or supplement to the functional treatment with botulinum toxin. Neurotomies, on the other hand,

are mutilating and irreparable procedures and their long-term effect is unpredictable.

*Electrostimulation* of the spinal cord is an efficient treatment for spasticity resulting from traumatic lesions of the cord, but does not appear to be very effective in cerebral palsies [15].

### Uncontrolled muscle activity

Patients with uncontrolled muscle activity lack a sufficiently stable basis for standing and walking. Orthopaedic *braces* that guide the joints of the lower extremity while allowing free movement in the sagittal plane can help such patients perform these functions.

### Orthopaedic measures for functional disorders and deformities of the musculoskeletal system

The therapeutic options include the various »therapies« (physical therapy, occupational therapy, speech therapy etc.), braces, cast treatments and operations.

### »Therapies« (physical therapy, occupational therapy, speech therapy etc.)

The »therapies« can be subdivided into two main groups depending on their purpose: one for diagnosis and the other for treatment. All patients must be investigated individually so that they receive the appropriate treatment. This includes extensive testing of daily activities, recording the neurological development, sensory function and coordination skills. As well as all the problems associated with the actual musculoskeletal system, any deficits in neuromotor control must be diagnosed and treated. The various therapies (occupational therapy, physical therapy, speech therapy etc.) can pinpoint these cognitive sensory and motor deficits.

The patients are then treated on the basis of these investigation results. Training is required both for global functions such as balance or coordination and cognitive deficits, and for the functions of individual muscles, sensory problems or joint contractures. This very broad spectrum of problems requires therapists with in-depth training, particularly in neurophysiology. Many associated types of treatment are available (Bobath, Vojta, Kabat, Doman-Delacato etc. [6, 14, 22, 40]).

Hippotherapy for the training of balance and body control is also included in this group. For the purposes of general practice, however, the specific neurophysiological basis is probably less important than an understanding of the problems of the patients in relation to their everyday life. This requires a flexible approach to their evaluation and type of treatment.

The ideal treatment can probably be formulated as follows: Out of all the available therapeutic methods, those steps that are required for the patient must be picked out, like raisins from a cake, and applied in a coordinated manner. Which therapist tackles which joint or problem

is less important than the fact that many of the patient's problems are covered by the whole range of therapeutic interventions. Only a joint and coordinated approach will be able to give the patient the ideal rehabilitation.

### Braces

The various braces available cover a wide range, from the simple shoe insert to the complex, individually adapted electric wheelchair. Most braces are discussed in connection with other therapeutic methods in relation to individual body regions. The basic principles are outlined in ► Chapter 4.7.2 below. The use of braces is primarily aimed at enhancing the patient's abilities, for example locomotion and the use of the arms and hands. wheelchairs help with the locomotion, while standing aids are used to hold the patient upright. Orthoses replace deficient muscle activity, stabilize joints and preserve the balance in relation to muscle lengths, which is more important than actual muscle power.

### Cast treatment

Corrective casts provide an efficient way of stretching contractures. Unfortunately, this form of muscle lengthening is invariably associated with atrophy, and relapses as soon as the muscle power is restored are not infrequent. Nevertheless, such cast treatments are very helpful and useful and can be repeated. As with orthoses, it is important that the plastered section of the skeleton should be corrected and held in the optimal position in order to achieve efficient stretching. Since structural changes can arise in the muscles after a cast treatment lasting 4–6 weeks, the cast should not be applied for more than four weeks. If the desired result has not been obtained by the end of this time, it is more useful to use an orthosis to maintain the acquired position and then repeat the cast treatment after several months. The cast treatment is more efficient if administered two weeks after an injection of botulinum toxin. The botulinum toxin also appears to have a longer-lasting effect as a result.

**! Skeletal deformities can merely be checked or their progression delayed by corrective casts. Genuine corrections are possible only in individual cases, and usually only on the foot.**

### Orthopaedic surgical measures

Before any operation, troublesome functional changes must be differentiated from useful ones. For patients who are capable of walking, a gait analysis, ideally in a gait laboratory, is essential. The basic aim of surgical treatment is to restore muscle lengths and, if possible, muscle strength, and lever arms. A muscle becomes contracted when it cannot be stretched properly because of spastic activity. More than the skeleton, the muscles require regular stretching in order for them to

be stimulated sufficiently to grow. In healthy children, this stretching is produced by everyday activities. In spastic muscles, however, stretching occurs to a much lesser extent or may even be completely absent. As a consequence, the muscles grow less than the skeleton, resulting in muscle contractures. These, in turn, hinder the patients and lead to further functional (the antagonists become overlong) and structural deformities (the joints are loaded in an abnormal position and the bone adapts to the new loading pattern). The short muscles must therefore be lengthened.

Surgical treatment is indicated particularly for deformities that respond poorly to conservative measures. The timing of the operation depends on the extent and progression of any existing deformity. The age of the patient is of secondary importance. In order to achieve the optimal functional benefit, all deformities at differing levels must be tackled at the same time. Such combined procedures have been shown to improve walking ability [34]. We can choose from a variety of *surgical methods*:

- *Aponeurotic lengthening procedures*: In this method the aponeurosis or an intramuscular tendon is divided transversely once or repeatedly. The muscle is then stretched by physical therapy and splints (stretching splints). This follow-up management has major advantages over the former cast treatments: The leg can be removed from the splint so that the skin can be checked. The rate of stretching can be adapted to the patient's pain. Nerve lesions have been described after cast treatments as a result of excessively rapid stretching [2].

In fact, we have observed this complication in one of our patients after lengthening of the knee flexors with cast treatment. Nerve damage can be prevented by using a stretch splint to stretch the conscious patient's muscle gradually. The aponeurotic lengthening does not lead to a loss of power [7] and strengthens the actual shortened section of the muscle. On the other hand, the treatment is much more painful than tendon lengthening.

- *Tendon lengthening procedures* can be performed as a Z-plasty alone or as a Z-plasty with the sliding technique (transcutaneously for the Achilles tendon). Both surgical techniques can be used for any tendon: In the conventional Z-plasty lengthening procedure, the tendon is split lengthwise and one part is shifted distally, the other proximally. The long ends are sutured.

In the sliding technique, the tendon is divided transversely across half its width at proximal and distal ends and then pulled lengthwise, causing the parallel tendon fibers to slide up and down. Since these operations lengthen the tendon but not the actual shortened muscle belly, this approach can be criti-

cized from the pathophysiological standpoint. If a recurrence occurs, the muscle belly will shorten even more.

On the other hand, these measures are effective and, in cases of severe shortening, often the only option. Follow-up management is also simple, being limited to a lower leg cast or splint, without the need for stretching by physical therapy. Accordingly, rehabilitation is less painful. Overcorrections occasionally occur, resulting in a loss of muscle strength. Recurrences are rare [10, 37].

- *Correction with external fixator (Ilizarov)*: This method is indicated particularly if severe long-standing contractures need to be stretched. In these patients an additional component has invariably developed in addition to the muscle contracture: The capsular ligament apparatus and all connective tissue structures in the fascia and subcutaneous tissue are also too short. The external fixator can correct the position either primarily or after muscle lengthening. The fixator is also a useful alternative to a repeat lengthening operation in the event of recurrences.

The external fixator is fitted so as to bridge the respective joint. The contracture is then stretched immediately. The rate of stretching can be adapted to the patient's symptoms. The procedure is burdensome for the patient, and there seems to be a higher recurrence rate than after corrective osteotomies. The method can lead to atrophy and fibrosis of the muscles, and fixator care is difficult [3, 21].

- In severe deformities a combination of tendon lengthening followed by a corrective extension osteotomy can be used alternatively to the external fixator. This approach is less irksome for the patient and does not endanger the joint stability.

- Another option is to *shorten the bone*: This method is used only in combination with other complex surgical procedures (e.g. hip reduction). However, this technique is not suitable on its own for the relative lengthening of muscle groups, as it results in a length discrepancy in the skeleton.

After each of these therapeutic methods the relevant muscle gradually adapts itself to everyday use. Particularly if length growth is not concluded, the still spastically active muscle may again become too short relative to the bone with the consequent risk of recurrence. Each of these methods can be repeated as required, however. The recovery of the antagonists is important. If these fail to recover despite training, a shortening operation is carried out.

**! After all muscle-lengthening procedures the risk of recurrence is high, particularly during growth, although all measures may be repeated.**

— One further surgical measure that may be required involves the redirection of muscle power, i.e. *a muscle transfer*. Clinical experience indicates that this regularly results in a loss of muscle strength because, on the one hand, the muscle is not ideally structured for its new use (in respect of its geometrical structure and fiber length) and, on the other, the fixation of the muscle always involves a reduction in the basic tension. Since, in physiological respects, the tension of the muscle depends on its length, the reduction in tension is equivalent to lengthening. Just as with tendon lengthening procedures, the muscle will adapt itself, resulting in a reduction of strength.

In spastic syndromes the muscles cannot be controlled in a refined manner. A tendon transfer procedure in such patients is therefore equivalent to a tenodesis.

— *Reconstruction of lever arms*: This can be achieved with orthoses, provided these are worn on an almost permanent basis. An alternative option is *tenodesis* or *arthrodesis* of the bridged joints. These procedures are particularly suitable for the spine and rearfoot. Our clinical experience with these operations is good. The patients remain orthosis-free postoperatively and can wear simple shoes, possibly with slight corrective tensioning, or may require a few seating aids, but they retain the same functional status as before the operation. *Arthrodeses are contraindicated for major joints such as the knee, hip, shoulder or elbow*. These joints may not be fused otherwise a long lever arm substantially increases the risk of pathological fractures in patients with fragile osteoporotic bone. Even in cases of severe dynamic instability due to muscle weakness, major joints must be managed with orthoses. Only in the hip may an instability due to a bone deformity be managed by surgical correction.

A second step is the *correction of torsions*: the alignment of the knees and the feet needs to be set correctly in the direction of gait. In some cases the long bones of all four segments of the legs need to be corrected at the same time for this purpose.

**! During growth, the foot and spine are stabilized with the aid of orthoses. Alternatively, arthrodeses may be implemented towards the end of growth.**

### Prognosis

Neuro-orthopaedic problems arise as a result of a neurological disorder which, as a rule, can neither be influenced nor cured. Orthopaedic measures can correct deformities and improve the functional status, but recurrences are almost unavoidable since the underlying neurological condition continues to exert its influence unchanged.

During this period the skeleton is plastic and adapts itself relatively quickly to modified conditions and forces. If the muscles are affected, their length and force must not only be preserved, but additional length must be gained in order to cope with the growth in bone length. Despite all the above, the risk of recurrence has proved to be low, as is also the case with our own patients [35]. For this reason – not to mention the need for increasing corrections as contractures persist – postponing any necessary surgical corrections is not a good idea.

### Rehabilitation

The treatment of neuromuscular disorders is rarely a purely orthopaedic problem. In contrast with other paediatric orthopaedic conditions, the correction and cure of the orthopaedic disease does not mean that the patient's problems are solved. Since neuro-orthopaedic patients are usually disabled by their neurological condition, the integration of the patient in everyday life is more important than the treatment of the deformities. The orthopaedist is only one specialist in a team of many who are responsible for the rehabilitation of the patient.

Besides the orthopaedic problems, sensory and cognitive functional deficits and other malfunctions of the nervous system must be diagnosed and treated. A whole range of braces is often required in addition to the therapeutic measures. Optimal rehabilitation of the patient can be ensured only through the efforts of a team of specialists from the fields of occupational, physical and speech therapy, orthopaedics, pediatrics and rehabilitation therapy.

Only with the aid of modern instrumental gait analysis is it now possible to determine whether an increase or a decrease in muscle power has resulted in the prevailing functional disorder. While the numerous methods outlined above can be applied to correct the abnormal muscle power or activity, the treatment of the muscle weakness primarily consists of a medical training program. If adequate power cannot be developed despite the patient's cooperation and sufficient time (at least six months), surgical shortening of the muscle-tendon apparatus to restore the correct muscle tension must be considered. Although such shortening procedures are difficult and have a poor reputation in terms of their results, they are certainly helpful for the knee extensors in particular [5]. We have started shortening the anterior tibial muscle together with lengthening of the Achilles tendon in severe equinus deformities to overcome the dropfoot problem generally, and the results seem to be favorable.

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## 4.7.2 Braces

R. Brunner

### ➤ Definition

Braces can be divided into orthoses and mobilization aids.

### 4.7.2.1 Orthoses

#### ➤ Definition

Orthoses are supports applied externally to correct a deformity or to control the forces acting on the musculoskeletal system. In neuro-orthopaedics they always fulfill two functions: They provide *stability* while at the same time *correcting abnormal function and/or shape*.

Orthoses can only act from the segment that is stable in space on the more unstable segment. For example, the stable ground can be utilized to shape the overlying foot (more unstable or more mobile than the ground). The appliance in this case is known as a shoe insert and acts only when corresponding pressure is applied by the foot against the ground, i.e. during the stance phase of walking or while standing. During the swing phase, by contrast, the foot must be held by the lower leg (ankle-foot orthosis) or at least in a comprehensive support (Nancy-Hylton orthosis) in order to correct the shape. A distinction must therefore be made between stance-phase and swing-phase deformities.

Our skeleton consists partly of long bones, while other sections of the body are more like joint chains, for example the spine or the foot. While these joint chains remain in the anatomically correct position they are loaded in the optimal manner, for example as in a normally shaped foot. Only in this way can the foot remain a stable lever arm for the triceps surae muscle, which is a key muscle for posture control. The precondition for a normal shape of the foot is adequate muscle activity and dynamic control during weight bearing. Only then can disruptive external forces be intercepted. Since this dynamic control is usually lost in neuromuscular disorders, progressive deformities develop, particularly in joint chains such as the foot or spine. Consequently, inappropriate muscle stabilization occasionally leads to severe deformities such as flat feet or club feet, resulting in a loss of lever arm function and dynamic stability.

A similar situation applies to the spine in respect of scoliotic or kyphoscoliotic deformities. Ideally, the axially acting forces are dissipated via the vertebral bodies. Once the vertebral bodies break out sideways from the chain, the scoliosis can progress rapidly.

In neuro-orthopaedics, deformities occur particularly when patients with deficient body control are

straightened up. The joint chain then gives way on the one or other side, depending on the difference in muscle tone and the externally acting forces. Gravity acts as a permanently deforming force in this position. Since gravity does not apply while the patient is lying down, and muscle tone is also lower as a rule, problems arise for the neuro-orthopaedic patient especially in the upright position. For this reason, braces are particularly efficient if they are worn throughout the day as functional orthoses.

This important active element is missing in relation to nocturnal splints. Although they retain the secured body sections in an ideal position at night, functional control is not ensured during the day. Moreover, the patients experience stretching as an unpleasant, and possibly even painful, sensation. Although the patients may tolerate such unpleasant sensations during the day while distractions are available, their sleep at night is disturbed. Consequently, patients will often minimize any stretching or even dispense with the use of the orthosis altogether, making the nocturnal splint completely ineffective. For these reasons we prescribe nocturnal splints only in rare individual cases.

❗ **Since uncomfortable corrective forces are poorly tolerated at night, while abnormal postures and deformities interfere with function, functional orthoses are more useful than splints.**

Orthoses should correct the shape of, and stabilize, the bridged body sections. However, if the skeleton can no longer be aligned in the correct anatomical position, force is transferred to the orthosis at the deviating points of the joint chains, resulting in the regular development of pressure points at these sites. If the orthosis is cut away at these pressure points, this allows further deformation to develop. This postpones the problem of the pressure points, while the deformity becomes increasingly accentuated to the point where the orthosis requires further adaptation. The correct procedure in the event of pressure points is to check the position of the body section in the orthosis and, if necessary, order a new orthosis in the correct position rather than make further adaptations.

❗ **Pressure marks are hardly ever a local problem but are usually due to incorrect or incomplete correction of the embedded body segments.**

It is important that patients are hindered as little as possible by their braces in their everyday life. Moreover, braces should not produce any pressure points. This aspect deserves particular attention in patients who are uncooperative. Once they have learned that the braces cause pain, they will be unlikely to tolerate them any more. In other words: no appliance is better than a troublesome appliance. Functional orthoses should, if possible, be of

## 4.7.2 · Braces

lightweight construction in order to minimize the oxygen and energy consumption. Modern technology and the use of plastics are therefore preferable to the old designs made from metal and leather [1, 3]. In addition, leather is not washable and therefore hygienically suspect, particularly for those parts of the body where profuse sweating occurs (feet, hands and trunk).

### Shoe insert and shoe modifications

#### Definition

Inserts can be incorporated in the shoe loosely or as fixed components. They support the calcaneus by the application of pressure from below and straighten the foot deformity. The footwear may need to be strengthened in order to keep the foot positioned over the insert.

A *shoe insert* can be considered as the simplest form of orthosis. For foot deformities such as pes planovalgus, it serves as a tried-and-tested resource as long as powerful spastic muscle forces are not additionally present. The aim of the insert is to correct the shape of the foot by applying counterpressure to the foot. This will prove successful if only the calcaneus is supported (at the rear on the medial side) and thus embedded in a varus position (■ Fig. 4.101). Severe deformities or strong forces may require this embedding to be shifted further forward. The shape of the foot is corrected by the straightening of the calcaneus. However, the preconditions for a successful outcome with this treatment are that

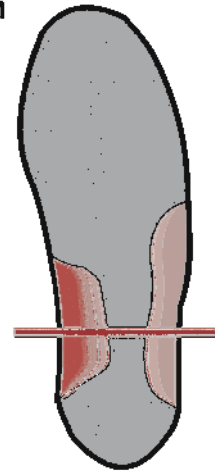
- the foot – particularly the heel – presses against the insert, i.e. that the patient walks with a heel-to-toe roll and not with an equinus gait (■ Fig. 4.102), and
- the shoe and insert grasp the foot as a whole. Often the footwear is too weak and the insert is pushed aside, or else the foot deviates away from the insert. An adequate correction shows correct foot alignment when the orthosis is worn compared to the situation without (foot in the direction of gait).

The inserts can be integrated in the shoe to prevent them from slipping out of place, although each pair of shoes will need to be adapted in this case. This type of shoe insert is required for feet that can only be grasped with difficulty. A loose insert is cheaper and allows the patient to change his or her shoes. Loose inserts have proved to be highly effective and are sufficient in most cases. Small rubber nodules under the heel can also prevent slippage of the insert in relation to the shoe. If stronger forces are involved, e.g. in heavy patients or in the case of pronounced deformities, it may be necessary to improve the securing of the foot.

This can be achieved with stabilizing medial and lateral reinforcements in the shoe, so-called *upper reinforcements*. However, these upper reinforcements must extend well to the fore to ensure that the foot really is adequately

#### View from top

medial side lateral side



#### Cross-section



■ Fig. 4.101. Schematic view of an *insert* that supports the calcaneus. The heel is balanced, and the whole foot thus indirectly straightened, via the medial and lateral support, which must be located under the calcaneus



■ Fig. 4.102. This correctly prepared *insert* does not achieve its objective. As shown by the marks of everyday use, weight has *only* been placed on the forefoot section (the patient walked with an equinus gait)



■ Fig. 4.103. This *standard shoe*, with stabilization of the rearfoot on both sides, shows deformation after just two weeks' use. The leather is too soft and unable to withstand the force of the abduction flat valgus foot

stabilized (■ Fig. 4.103). Today's modern shoes are soft and can rarely withstand strong forces, even if the upper reinforcements extend up around the ankles. The leather gives way and the whole shoe soon becomes warped. If the acting forces cannot be intercepted even with these reinforcements, then lower leg splints will be required. Alternatively, such modified shoes are worn during periods when the orthoses are not worn.

As a rule, however, inserts should not be prescribed lightly: If they really are indicated, then walking barefoot is not particularly useful, or even healthy. The inserts must then be worn in slippers too, which must already be considered a radical measure in today's households where children usually run around in their socks.

### Ankle-foot orthoses

#### ➤ Definition

Ankle-foot orthoses are splints that hold the foot in a corrected position and guide and stabilize it in relation to the lower leg. They therefore control the foot and its position in relation to the ankles. They are used as functional orthoses (during walking/standing) or as postural splints (at rest, usually with a relieving effect).

— Abbreviation: AFO

#### Objectives of ankle-foot orthoses

- Reestablish lever arm
- Stabilize during function
- Correct abnormal skeletal shape
- Stretch the muscles (particularly the triceps surae)
- Substitute for paretic muscles
- Reduce spasticity and tone

The efficacy of ankle-foot orthoses in improving walking ability has been demonstrated in gait investigations [4, 7, 9–11, 15]. Depending on the type of functional problems, they can support and stabilize, reestablish the foot as a lever arm for the triceps surae muscle, or else guide the ankle by grasping it with a freely movable orthosis joint. If the foot alone needs to be controlled, orthoses that grasp the foot (Nancy-Hylton orthoses) are sufficient. In this case the orthoses correspond to particularly tall support shoes.

Movement in the upper ankle can be restricted by stops on the orthosis joints or by designing the splint as a spring (leaf spring orthosis), thereby reducing the load on the muscles while at the same time stabilizing the joint. The foot is also held in a better position. In order to achieve control via the upper ankle, and thus via an equinus foot position, a lever arm on the proximal lower leg is always required [14].

Finally, the splints can also be designed as completely rigid components. Although these afford the greatest stability, walking, particularly the heel-to-toe roll and push-off, are made more difficult [11]. Nevertheless, such stiffness is required if they have to serve as a functional replacement for a paretic triceps surae muscle [9, 10, 13]. An extensive range of orthoses is available, and the task of the orthopaedist is to identify the most appropriate one for the individual patient [2] (■ Fig. 4.104 and 4.105).

Patients who do not possess adequate intrinsic dynamic control of their limbs show increased muscle tone. The uncontrolled positioning of the lower limbs on foot-strike and during the stance phase can trigger spastic reactions that patients are unable to intercept. The aim of orthoses is to stabilize the joints and thus avoid abnormal postures during standing and in the stance phase of walking and thereby reduce tone and spasticity [2, 7, 13]. While a ramp under the toes (toe extension) can positively influence the spasticity in patients who have suffered damage to the already fairly mature brain, in our experience such toe ramps in the shoe are not very effective in patients with cerebral palsy and can prove irritating.

At any rate, the orthosis will fulfill its purpose only if it sits optimally and holds the foot correctly. The *foot skeleton* must be aligned in the anatomically correct position



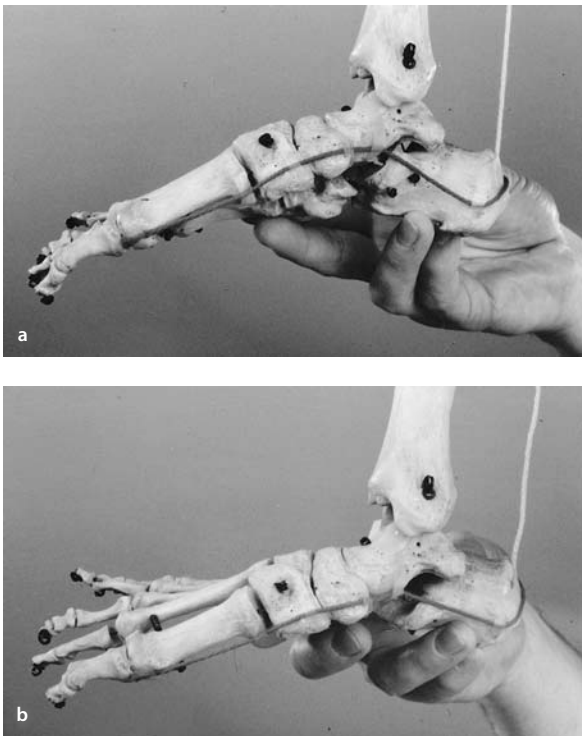


■ Fig. 4.104a–d. Ankle-foot orthoses. a Rigid functional ankle-foot orthosis, b Elastic functional ankle-foot orthosis that permits approx. 25° of dorsiflexion, c Functional ankle-foot orthosis with free joint, d Lower leg brace



■ Fig. 4.105a, b. Two ankle-foot orthoses of the same age (a). That for the right foot shows no signs of adjustment, that for the left repeated signs of adjustment. b The symmetrical abduction flat valgus feet in the orthoses. The right orthosis corrects the foot, but the left permits the deformity to persist, resulting in typical pressure points on the medial side beneath the navicular bone, where the orthosis has been adjusted repeatedly without effect





■ Fig. 4.106a, b. Normal *foot skeleton* in the anatomical position (a). When the foot is at right-angles to the ankles, the triceps surae muscle, represented by the woolen thread attached to the calcaneus, is taut and straight. **b** When the *foot is bent*, causing the calcaneus to subluxate in the lower ankle, the woolen thread slackens conspicuously without any change in the position of the foot in relation to the lower leg

so that the development or progression of deformities can be counteracted. Thus, an abduction pes planovalgus must be adducted and the heels, in particular, placed in a varus position, while the opposite maneuvers are required for clubfoot. If the position is not corrected, pressure points will arise at the typical sites: medially beneath the navicular bone in abduction pes planovalgus (■ Fig. 4.106a, b) and on the lateral edge of the foot in clubfoot.

However, the triceps surae muscle is often contracted in patients with such foot deformities. The exaggerated valgus or varus position of the calcaneus reduces the distance between the origin and insertion of this muscle [5], thereby concealing the contracture and making any extension largely impossible. In fact, the muscle becomes even more contracted and the subluxation in the lower ankle more exaggerated. The contracture of the triceps surae muscle prevents full dorsal extension, usually leaving an equinus foot of varying severity when the foot skeleton is anatomically aligned. The doctor and orthopaedic technician are left with the alternative of either ignoring the foot deformity and positioning the foot in a plantigrade position (i.e. at right angles to the ankle) or of keeping the

foot skeleton in an anatomical position and integrating the equinus foot in the splint.

Since we have never encountered the situation where an equinus deformity makes walking impossible (even though orthoses may be required for walking), and since an orthosis can fulfill all the therapeutic goals only in this position, we always place the foot in the orthosis in an equinus position as much as required. A really troublesome contracture of the triceps surae usually improves during orthosis treatment in this position, albeit only over a protracted period. Articulated orthoses are more efficient, but should only allow dorsal extension while blocking plantar flexion. If quicker correction of a contracture is required then other methods (cast correction, surgery) must be employed.

An attempt should be made to align the *foot axis in the direction of walking*, particularly in those patients that are able to walk. Many patients, however, adopt an intoeing gait as a result of internal rotation at the hip and then compensate for this by bending the foot. If the foot is held anatomically in the orthosis, we try and align the 2nd ray, as a reference for the foot axis, with the femoral axis. As a result, and because of the internal rotation at the hip, the foot must stand at an inwardly rotated angle in relation to the direction of walking, which is not the case without the orthosis. Unfortunately, few compromises are possible for solving this problem when positioning the foot in the orthosis, since any correction of the foot position in relation to the direction of walking worsens the skeletal position in the orthosis. This phenomenon must be explained to the patients and their parents and carers, as they will often feel that »the patient walks better without an orthosis«.

The aim of the orthosis is to produce a *heel-toe gait*. This requires adequate extension of the knee at the end of the swing phase. An equinus gait pattern will result, however, in patients with knee flexion contractures or spasticity of the knee flexors despite the fitting of a correctly designed orthosis and even with the foot adjusted in a plantigrade position. In these cases the orthosis supports the weight-bearing equinus foot and prevents its deformation. It is important that parents and carers should be told of the reasons for the failure to achieve a heel-ball gait so that the orthosis is nevertheless accepted.

### Knee-ankle-foot orthoses

#### ► Definition

Knee orthoses used in neuro-orthopaedics always include the foot. In many cases, an ankle-foot orthosis on its own will be sufficient for stabilizing the knee, provided this incorporates a backward lean. In rare cases, the lower leg orthoses will need to be extended up to the thigh, either via a hinged joint or rigidly, and bridge the knee.

— Abbreviation: KAFO



■ Fig. 4.107. Ankle-foot orthosis with a slight forward lean and corresponding shoe



■ Fig. 4.108. Knee extension brace with a free knee joint and extension bar that allows rapid tightening and loosening. The brace can be fitted in flexion, which is easier compared to the extended position, and then quickly extended

Functional braces can guide the knee indirectly in the sagittal plane (flexion/extension) and do not need to encroach upon the knee for this purpose [13] (► Chapter 3.3.7, ■ Fig. 3.309). Increasing the backward lean of an ankle-foot orthosis (i.e. increased equinus foot position) causes the knee to extend more, while a greater forward lean will increase flexion. An important factor is the angle between the orthosis shaft and the sole of the shoe, whereas the position of the foot between the two is not relevant. It is advantageous if the orthosis can help the patient produce an indirect extension moment at the knee during walking, as occurs physiologically. During walking, the ground reaction force very quickly moves in front of the knee, causing the knee to extend indirectly (passively). If this effect is to be reproduced with an orthosis, it will need to incorporate a slight backward lean in a system consisting of the orthosis and a corresponding shoe (a forward lean of the whole system is never desirable; ■ Fig. 4.107).

The knee can be guided indirectly by means of this backward lean. An articulated knee-ankle-foot orthosis can basically intercept only sideways-directed forces when instabilities in the frontal plane are present. If the orthosis must also be effective in the sagittal plane, the knee must be placed in a fixed position, which greatly interferes with walking. The only exception is knee hyperextension in stance, where the KAFO limits extension while leaving flexion free. *Knee extension braces* are useful primarily in the postoperative treatment after muscle lengthening procedures (■ Fig. 4.108). The extension braces serve as a substitute for casts and afford much better control of extension. An overextension of 10–15° in the orthoses is required, however, in order to achieve full extension of the knee, since this overextension is lost as a result of compression of soft tissues and the deformation of the orthosis. These braces may only be worn for a limited period, e.g. during rehabilitation after a knee flexor lengthening procedure.

The compressibility of the soft tissues of the thigh also jeopardizes the efficient mechanical control of varizing or valgizing forces. Orthoses can probably prevent extreme positions only.

#### **Hip-knee-ankle-foot orthoses**

A distinction is made between stabilizing orthoses that replace missing muscle function and orthoses that guide the hip.

#### **Hip guidance orthosis for spina bifida**

##### ► **Definition**

Hip guidance orthoses include the feet, ankles, knees and hips, and can even extend to the trunk. They are used to stabilize the bridged joints and replace missing muscle function. They are used as functional orthoses for standing and walking.

— **Abbreviation:** HKAFO



■ Fig. 4.109. Swivel walker (left) and Parawalker (right) from ORLAU, Oswestry, GB (see text for details)

Hip-knee-ankle-foot orthoses (MMC-type orthosis) are used when the lower extremities, including the hips, cannot be actively controlled, or are only inadequately controlled. This is typically the case in patients with flaccid pareses, i.e. patients with spina bifida. The stabilization of the bridged joints allows the patients to stand and walk. Since the active motor function at hip level is insufficient, a positive effect on walking can be achieved if an automated step mechanism can be incorporated in some form (reciprocator, special joints). Shifting the weight onto one leg causes the other leg to move forward without muscle power. Thus, steps are produced simply by weight transfer. Since the opposing leg must be raised off the ground to enable it to swing forward independently, the orthosis itself must be a very stable and rigid construction. The actual weight is less important (example: »Parawalker« (from ORLAU, Oswestry, GB); ■ Fig. 4.109).

If the orthosis is sprung, the upper body only swings to the side over both legs, without releasing one leg. The orthosis can also be combined with a trunk orthosis (corset) if the trunk is also unstable. It should be borne in mind, however, that walking is more difficult and consumes more energy, the higher up the orthosis extends.

Apart from paralyzed patients, such orthoses can also be used in patients with myopathy. Factors that limit the use of such high orthoses are obesity, old age, spasticity and spinal deformities [8].

If orthoses extending to the knee or higher are required for walking, locomotion in a wheelchair is more efficient, which is a particularly important consideration for longer distances [6]. High orthoses should therefore be considered as training devices rather than actual functional braces. An improvement in, or even the imparting of, the ability to walk in everyday life is achieved only in isolated cases. Walking in these devices can more appropriately be viewed as a sporting activity.

In order to train the patient in the *balancing reactions* required for the weight transfer; A »Swivel Walker« (from ORLAU, Oswestry, GB) can be used with small children (2- to 3-year olds) (■ Fig. 4.109). This walking apparatus essentially consists of a rigid frame mounted on a baseplate. This baseplate rotates over two large »feet«. The system is more stable than an Hip guidance orthosis and allows the child to learn how to walk by weight transfer.

### Abduction splints

#### ► Definition

Pure abduction orthoses prevent adduction at the hip. They can be used functionally or as postural aids.

*Functional abduction orthoses* (Machini brace type) consist of two movable opposing shells that accommodate the upper legs. The orthoses are designed to prevent the legs from crossing over (scissor gait) and are set in a position of slight abduction.

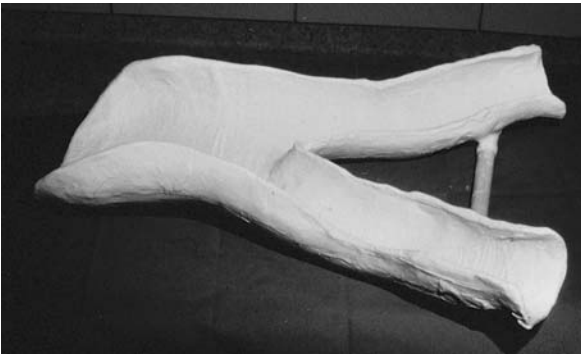
The SWASH orthosis (SWASH = Sitting, Walking, Abduction, Standing, Hip) has a similar design. These orthoses are worn during walking and are designed to facilitate walking. *Postural abduction splints*, which hold the legs in abduction, and possibly also in a desired position of rotation and flexion, are used to prevent hip dislocations and adduction contractures. These rigid orthoses can be made from plaster of Paris or plastic (■ Fig. 4.110). For heavy children, an abduction wedge placed between the legs at night and fixed in relation to the mattress can serve the same purpose. Movable orthoses allowing varying degrees of abduction (e.g. those described above) can be used alternatively. Rotation at the hip, however, can only be controlled if the lower leg is included in the orthosis.

### Trunk orthoses

#### ► Definition

Trunk orthoses stabilize the trunk in the sagittal and frontal directions. They counteract the onset or progression of spinal deformities.

— *Synonym: Corset*



■ Fig. 4.110. Plaster splint for positioning the legs as prophylaxis against hip dislocation



■ Fig. 4.111. Without a corset the patient collapses into extreme kyphosis and is unable to look straight ahead

Patients with *flaccid paresis* with a substantial neurological component lack not only control over their extremities, but also trunk stability. But even patients with pronounced spastic tetraparesis and spasticity and hypertonicity of the extremities will often show muscle hypotonia in the trunk, particularly the lumbar spine. The objective of the trunk orthosis (corset) is to compensate for this instability and stabilize the patients in an upright position. If the patients are straightened without an external stabilizer, the spine will collapse into a scoliotic and/or kyphotic position (■ Fig. 4.111). These deformities will subsequently become fixed at bone level.

*Spinal deformities* constitute another indication. Scolioses or kyphoses or combinations thereof are not uncommon in patients with poor neuromuscular control of the trunk. The prevailing muscle tone indicates the direction in which the spine collapses. In such cases, gravity constitutes an important pathological mechanical factor that cannot be compensated for by the weak muscles. Any deformities that are present are usually progressive.

Corsets are also indicated for frequent bad posture, even if the patient is able to straighten up actively. His or her ability to do this is of no benefit if it is not put into practice. Otherwise, the common scenario of bad posture on one side will, in the long term, end up in a spinal deformity that will not be amenable to correction by conservative means.

Adaptation of the corset is also made more difficult, and the corset must be worn tighter in order to counteract progression. For a corset fit without pressure points, the axially acting force must be dissipated via the spine. The corset must therefore support the whole trunk. The best fit that produces the fewest pressure points is achieved if the plaster mould is taken in an overcorrected position. In the corset the trunk springs back slightly into the deformed position, but less so than if the impression is taken with a straight trunk. Using this method, corsets can be adapted even in difficult situations, for example for children with spinal muscular atrophy for whom surgical correction is out of the question.

Furthermore, the corset should not rest on the iliac crest, otherwise it will tend to ride up, particularly in seated patients. It must rather be wedged between the iliac crest and the rib cage, firmly occupying the whole waist while, at the same time, stabilizing the movable and dynamically uncontrolled lumbar spine. If they are only counteracting scoliotic forces, such corsets permit flexion and extension movements, at least while no serious spinal deformity exists. It has also been shown that this mobility can be beneficial, even in severe deformities, since fewer pressure points form and the corsets are more likely to be worn than the theoretically more correct completely rigid versions. If there is only dynamic instability of the lumbar spine, short braces extending from the pelvis up to the bottom of the rib cage may suffice. These may be worn only for situations where trunk stability is required (such as in some types of occupational therapy or school).

The efficacy of any corset used for neuromuscular spinal deformities must be checked radiologically. To this end, and ideally in the same session, general x-rays of the spine under load should be recorded, with and without the corset, with the patient seated or standing. The corset should correct at least 25% of the curve.

Generally speaking, corsets may also be indicated in patients with *muscular dystrophy*, although surgical correction should be performed as soon as possible in these patients. Progression is certain, and the patient's general condition will only deteriorate. The *duration of corset use* will depend on the therapeutic objective. Since the primary effect is to compensate for the action of gravity, it should only be used in the upright position. The constant fear of muscle weakening should not be a primary consideration in the use of trunk orthoses. The muscles are permanently overstretched in any case as a result of the bad functional posture and their insufficiency is demon-



strated by their inability to hold the trunk in the correct position. The corset is not worn all day, thereby allowing sufficient muscle activity for maintaining strength. Finally, the muscle strength has been impaired merely by virtue of the dynamic instability, and the patients have to make the extra effort to withstand the effects of gravity.

### Braces for head control

In many patients the head control is impaired as well as trunk control. However, since such patients can usually manage to balance the head above a stabilized trunk, the braces are designed to minimize the force required to hold the head in place. It can be difficult, if not impossible, to satisfy all the demands of the patient, parents, therapists, carers and the outside world in a single appliance. A simple way of improving head control is to ensure that the patient remains in a *half-sitting position* and place the head on a *head-neck support*. However, this method means that the patient can only look directly straight ahead and thus restricts contact with the outside world.

While an additional tabletop allows the arms to be used for added support and thus improves trunk stability, the arms in this case are being used as support organs

rather than for their proper purpose as functional tools. This hinders their use and serves as an obstacle to improvements in arm and hand function. A better solution is a trunk orthosis that reduces the patient's postural effort and facilitates, or even allows in the first place, balancing of the head. A trunk orthosis will also stop patients with thoracic, and in some cases lumbar, hyperkyphosis from having to sit down and hold their necks in extreme lordosis by way of compensation in order to look straight ahead. Other options include *lengthening of the corset* in the manner of a Milwaukee brace, a *cervical collar* or *Glisson sling*. All of these braces are poorly accepted, however, even though a Glisson sling, for example, is ideal since it holds the patient's head while allowing movement in all directions (■ Fig. 4.112).

### 4.7.2.2 Mobilization aids

#### Wheelchairs

##### Definition

A wheelchair is a seat mounted on four wheels that provides locomotion for patients who are unable to walk. A distinction is made between an active wheelchair, in which patients propel themselves forward by their own muscle power, and a pushed wheelchair. The modular wheelchair should ideally be adapted to the patient. The electric wheelchair provides locomotion if the patient's own muscle power is not enough.

Wheelchairs are technically designed to be smooth-running and can be maneuvered by patients with the minimum of effort. To this end, large wheels with a low rolling resistance are fitted at the back, where most of the weight is located. Small wheels at the front provide stability. If the chair is used outdoors then these wheels should not be too small, otherwise they will catch on small obstacles, e.g. stones. On the other hand, patients with a good sense of balance can maneuver their wheelchair themselves in almost any situation. In such cases small front wheels are better in functional respects. The weight can be shifted from the front wheels to the back wheels by weight transfer. The wheelchair travels more easily, but is less stable to backward falls. Such designs are suitable for patients with good balance. Otherwise, the weight should not be transferred too far back over the back wheels, or else brackets should be fitted at the back to prevent a fall.

Depending on the use and the needs in each case, wheels with drum brakes or obliquely angled wheels can be fitted to improve stability and protect the fingers from getting trapped. A wheel guard to counter splashes from the tires is also recommended.

Many patients with neuro-orthopaedic problems are partially or permanently reliant on wheelchairs, and optimal wheelchair adaptation can help the patient considerably in terms of integration in everyday life.



■ Fig. 4.112. Glisson sling



Ideally, patients should be sitting upright loosely in the wheelchair so that they are neither hampered by spasms, which can affect the whole body, nor obstructed in their contacts with the outside world as a result of an inappropriate sitting position. Wheelchair patients must be *instructed* in the use of their chair. This includes balance training, weight transfer, overcoming obstacles such as steps and standing the chair upright and getting back onto the chair after a fall. Where possible, a corresponding training program should be arranged under the direction of suitably trained therapists or in a rehabilitation center. Not all patients are strong enough to control an active chair.

But even severely disabled patients can manage very well with an electric wheelchair. This gives them freedom of movement and independence. Ideally, a variety of models should be tested before purchase so that the best wheelchair can be determined for the individual patient. Various electronic control systems are available on the market. Whereas some are small, handy models that can be positioned on a tabletop, others are equipped with an electronic filter against shocks or sudden movements. Only a test run will reveal which of the (usually expensive) models provide the best features for the patient's situation.

Some electric wheelchairs are also equipped with a standing device. This function can be used not only for training in standing, but also allows the patient to reach objects at a much higher level. Ideally, the patient should be able to stand completely upright, and this option is offered in certain wheelchair models. This enables the patients to use their full arm length, which can be very useful in kitchens or bathrooms where wall units are usually shifted to the back. The functional benefit offered by a correct standing function should not be underestimated.

### Seating adaptations

#### ➤ Definition

Seating adaptations allow the patient with a dynamic instability or with deformities of the locomotor system to sit upright.

If the patient has good body control, major *seating adaptations* are not required. But the worse the control, the more the body must be supported. An optimal adaptation reduces the effort required by the patient to remain in an upright position, and thus gives him or her to use the upper limbs in particular more freely [12] (■ Fig. 4.113).

The first step is to stabilize the pelvis by an anatomically shaped cushion. This should maintain the pelvis in an unrotated and horizontal position as a stable basis for the spine and trunk. After the pelvis has been adjusted, the legs should be positioned in an unstressed position. A symmetrical position of the legs is not important, nor

is it possible if asymmetrical deformities are present. If a symmetrical position is attempted in a patient with asymmetrical deformities, the pelvis will always be distorted in relation to the long lever arm of the legs. This exerts torsion on the spine, potentially resulting in scoliosis. Since it is not possible to actually fix the pelvis in the seat, the positioning of the legs must be adapted to the position of the pelvis.

The simplest solution is to sit the patient on a stable base and allow the legs to fall loosely. Slight distraction or a posture with spread legs is desirable as this enlarges the sitting area and improves stability. The distraction likewise does not need to be symmetrical. For patients with flaccid paralyses, a leg position with slightly extended knees is advantageous as this facilitates balancing of the wheelchair. In sports wheelchairs, however, greater knee flexion affords greater mobility. In spastic patients, by contrast, a right-angled position at the hips and knees has proved effective as this minimizes muscle tone and improves their ability to use the upper extremities in particular.

An increase in muscle tone is sometimes desirable since it can allow the patient to sit in a more »active« manner, i.e. remain upright without external control. However,



■ Fig. 4.113. Poor wheelchair adaptation. The patient is not held comfortably, even with numerous straps

such an increase in tone always affects the whole body and thus blocks free arm and hand movements. Wheelchair maneuvering itself is also hampered. In addition, all muscles are under increased tension, which accelerates the onset of fatigue. A looser seating position with minimum tone must therefore be attempted, even if any resulting muscle weakness of the patient must be compensated with braces. In accordance with the increased anteversion that is invariably present in these children, the hips are always centered best if the legs are held in a slightly internally rotated position.

If trunk stability is inadequate, lateral stability can be provided with an additional *back cushion* or *back section*. The side supports that hold the trunk must be arranged asymmetrically if necessary, although the pelvis must be grasped in a stable position in such cases. A corrective force cannot be exerted either with this back section or with the cushion or molded seat. Aids such as cushions or molded seats are not remotely fitted precisely enough to the body to achieve a corrective effect. A corset will be needed if functional, and especially structural, deformities are present, or if the effect of the back section is inadequate. Back sections on their own are not very promising, particularly if an abnormal kyphosis is present, and an anatomically shaped back section can be dispensed with once the corset has been adapted. Side supports, on the other hand, may still be needed in order to keep patients with poor balance upright. In small children, a vest that secures the upper body to the back section can replace the corset for a certain period (■ Fig. 4.114).

### Standing aids

#### ► Definition

Standing aids are braces that enable patients to stand upright, including those who are incapable of standing.

Training in standing is important for all patients who are unable to stand upright actively. It helps prevent osteoporosis, extends the hips and knees, ventilates the lungs and serves as cardiovascular training. The upright position also has a positive psychological effect. Even if patients no longer have any postural function at all, they can still be positioned in tiltable standing frames, with the hip slightly overextended and the knee fully stretched. At foot level, the load should be placed over the midfoot area and not the heel. This allows the whole skeleton to be loaded. Achieving this position is particularly difficult if the abdomen is faced toward the frame. When the patient lies face down on the tiltable standing frame, the hips are generally flexed by approx. 5–10°.

In the standing position it always looks as if the patients wants to sit down again. A better posture can be achieved if a padded wedge is placed under the thighs in the prone position (■ Fig. 4.115). This ensures that the hips are fully extended. For patients with better body



■ Fig. 4.114. Severely disabled patient who is unable to sit independently. Thanks to correct positioning in the *molded seat* she is able to sit in a relaxed manner with just one strap

control, standing frames that allow active standing with balancing reactions are more suitable. Various braces are available on the market for this purpose. These are fitted with just two vertical supports that are securely linked to a baseplate. Straps, or more rigid fixation elements, are attached to these supports to keep the patient upright. In this way, the various joints of the spine and lower extremities be released step-by-step in order to train the patient's body control. For added mobility, some standing aids are fitted with wheelchair wheels, although the adaptability of these mobile devices is limited.

### Walking aids

#### ► Definition

Walking aids are braces designed to facilitate assisted walking.

Various types of *walkers* are available. Some are fitted with hoist attachments and are particularly suitable for



■ Fig. 4.115. Good posture in the *standing frame* with extended hips thanks to the wedge placed under the thighs before the frame is righted

older and heavier patients. Walkers help train the patient in walking without the need for additional postural support from nursing personnel. Half-way solutions between standing frames and walkers include frames on wheels that allow the patient to take controlled steps. These devices compensate for the patient's poor balance by holding the body in a secure grip, although the arms remain free. Patients can therefore choose their own direction of walking, while the hands are free for grasping. This enables them to move independently on their own initiative within their immediate environment, grasp and transport objects and experience how walking can represent targeted locomotion.

A *walking frame* on wheels is suitable for more mobile patients. A better option than the anterior walker, over which the patient bends forward and thus flexes the hips and knees, are posterior walkers, which promote extension at the joints, particularly the hips. The simplest and most familiar walking aids are *crutches*, which afford bal-

ance and stability. Crutches with three legs are available for children obliged to learn to walk on crutches. The stability is greater and the crutches remain upright even when released.

### Bicycles

#### ➤ Definition

Bicycles for disabled patients possess maximum inherent stability either thanks to two large back wheels or stabilizers.

Bicycles enlarge the radius of activity, which is important for the psychological development of patients. The uniform movements reduce tone and spasticity. However, since pedaling does not require full extension at the knees and hips, in contrast with walking, bicycles are not suitable for use as training devices for building up strength in order to improve walking.

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### 4.7.3 Cerebral lesions

R. Brunner

#### 4.7.3.1 Cerebral palsy

##### ➤ Definition

Cerebral palsy is a permanent motor disorder due to a non-progressive defect or lesion of the brain in the early stages of development (definition of the 4th international congress of the study group on child neurology and cerebral palsy, Oxford 1964) [6]. It is therefore a symptom complex rather than a specific diagnosis.

— **Synonyms:** Infantile cerebral palsy, cerebral motor disorder (CP, ICP)

##### Historical background

In 1862 Little described cerebral palsy as a congenital spastic stiffness of the limbs with exaggerated tendon reflexes and increased muscle activity of symmetrical muscle groups, particularly the adductors, thigh flexors and calf muscles. Freud classified this symptom complex in 1892 on the basis of brain damage. Even today the term »cerebral palsy« covers a heterogeneous group of disorders.

##### Etiology and pathogenesis

The term »cerebral palsy« refers to a typical combination of signs and symptoms rather than a defined illness. Since the arrival of MRI scans, so-called idiopathic cerebral palsy has become rare. The causes can be subdivided into pre-, peri- and postnatal. During the prenatal stages, cerebral malformations (alone or as part of a pediatric syndrome) and infections (measles, cytomegaly, rubella etc.) and metabolic disorders lead to brain damage. A particularly high risk applies in cases of premature birth or small-for-gestational-age babies. The risk of cerebral palsy when the birth weight is less than 1.5 kg (3.3 lb) is 70 times greater than that for a term baby of normal weight [1]. The risk increases accordingly with multiple pregnancies [8].

Hypoxemia and infections in the perinatal phase, and particularly infections, craniocerebral injuries and tumors and, more rarely, hypoxemia in the postpartal phase, can lead to permanent brain damage. By definition, the damage affects a still immature nervous system and also influences its development. This explains why the full gamut of signs and symptoms is often not apparent from the outset. The children are initially often hypotonic, and the spasticity only manifests itself during the subsequent development of the nervous system and a change in basic muscle tone. The tone also often changes during puberty, and unfortunately a further subsequent increase usually occurs as well. Diagnostic classifications based on the affected regions and tone abnormalities often have to be corrected at a later stage. Spastic forms of cerebral palsy predomi-

nate nowadays, and can be combined with atactic, athetotic or dystonic components. Once common forms such as athetosis after kernicterus are largely preventable thanks to prophylactic and therapeutic measures of modern medicine and have now almost disappeared completely.

##### Occurrence

The prevalence of cerebral palsy in developed countries is just over 0.2% [1, 9, 10, 11, 13, 15–18]. Accordingly, it constitutes the commonest cause of disability during childhood. The improved management of premature births and small-for-gestational age babies has not resulted in a reduction in the number of cerebral palsies [3, 7]. Although children with a comparable risk are less likely to suffer damage to the brain, the minimum weight required for survival is always decreasing, thus producing a further rise in the risk of brain damage.

##### Clinical features and diagnosis

Cerebral palsy is most commonly diagnosed on the basis of missing or asymmetrical movements in the newborn infant. Delayed development, with the persistence of primitive reflexes and the late appearance, or complete absence, of motor functions form part of the clinical picture. Other typical signs, including increased basic muscle tone and spasticity, often appear only at a later stage. Some children, however, can also show a lifelong, permanently low tone in the muscles of the whole body or individual regions, often in the trunk.

Although the treatment is based on the clinical signs and symptoms, clarification of the causes is useful. Inherited disorders are occasionally concealed behind a picture of cerebral palsy, in which case the parents will need to be counseled accordingly. An *MRI* or *CT scan* is usually required to investigate the causes, and *metabolic investigations* can also be helpful.

Although, according to the definition, cerebral palsy is attributable to non-progressive brain damage, its manifestation does change during development and growth. In the initial stages, the maturation of the nervous system can produce a change in the neurological appearance. Secondary deformities of the musculoskeletal system that may be of functional relevance can arise during later growth. Thus, a patient who is initially able to walk can, for example, lose the ability to walk and appear to become increasingly weak, even though the underlying neurological condition is not progressive. Nevertheless, if the patient's clinical picture does change for no apparent reason, further neurological investigation must be considered.

! **Cerebral palsy is a mixed bag of etiologically very differing clinical conditions that exhibit similar signs and symptoms.**



Most striking of all are the *motor disorders*, which manifest themselves in the form of spasticity and muscle hypertonia, and are usually more pronounced distally. Muscle weakness of the antagonists or proximal muscle groups is often present at the same time. The psychomotor development of the patients is retarded as a result of these motor dysfunctions. The patients find it difficult to develop the necessary body control and learn balance reactions. As a result, the onset of walking is delayed and, if the child is severely disabled, head and trunk control is delayed or may not even develop at all.

The motor disorders are frequently accompanied by changes in *sensory perception*. These can exist in the form of hypesthesia, paresthesia or hyperesthesia. In patients with hemiparesis, the affected side can readily be compared with the healthy side, which explains why sensory disorders are so well known in connection with this distribution of neurological symptoms. But these problems also apply to cerebral palsy patients with differing topical distribution patterns. Thus, patients with diplegia, not infrequently show sensory problems on closer examination, while patients with tetraparesis sometimes refuse to wear shoes, or even socks, or else they avoid placing their feet on the floor, which also suggests the presence of a sensory problem. Such sensory disorders may also be responsible for the weight-bearing problems that occasionally arise after corrective cast treatments.

The defective speech and lack of cooperation exhibited by severely disabled patients often renders detailed examination of the sensory function impossible. The brain functions of severely disabled patients are often affected generally, resulting in *additional disorders of the nervous system*, e.g. cognitive damage, coordination problems, diminished intelligence, problems with vision and hearing, and occasionally with breathing, and abnormal circulation and temperature control [2]. These additional problems interfere considerably with the patients functioning and thus the rehabilitation.

If proprioception is impaired, as is the case with many cerebral palsy patients, intact vision is all the more important by way of compensation. Unfortunately visual function, in particular, is impaired in up to 67% of patients [14]. Since the frequency of refraction anomalies is no more common than in neurologically health individuals [12], the existence of a central visual disorder must be assumed. This also explains why patients can develop adequate balance control only with some difficulty.

An upright posture overtaxes the patients' balance control. They feel insecure in this position, becoming stressed as a result of having to guard against falls. But every stress leads to an increase in muscle tone, which further restricts the patient's reactions. Balance training must therefore form an integral part of the therapeutic

program. A position that is experienced as secure helps the patient loosen up and react more freely.

### Classification

#### Tetraparesis (or whole body involvement cerebral palsy)

These patients typically show distinct spasticity of all extremities with concurrent hypotonia of the trunk and neck muscles. The mimic and swallowing muscles are also affected, resulting in poorly articulated speech and salivation.

The *severity* of the neurological condition can vary considerably. Some patients remain independent and can even take up employment, whereas others are completely helpless, reliant on outside help and care, and are unable to communicate (■ Fig. 4.116–4.118). In our experience, the slightly disabled cases are, unfortunately, the exception rather than the rule. Since the brains of these patients is globally damaged, *functional disorders* in various areas are often present. Intelligence is more diminished the more severely disabled the patient. The actual damage to the brain is just one factor. As a result of the motor and sensory problems, the children are also handicapped in their mental development.



■ Fig. 4.116. Patient with *spastic tetraparesis* who is capable of walking when held





■ Fig. 4.117. Patient with *spastic tetraparesis* who is able to sit with the aid of a molded seat



■ Fig. 4.118. Severely disabled patient with *spastic tetraparesis*. She has no abilities whatsoever and must be fed via a *gastrostomy*

Severely disabled patients, in particular, also lag behind in their *growth*. The lack of cooperation and the impaired chewing and swallowing functions can lead to malnutrition. In addition, the spastic muscle activity increases the calorie requirement. Apart from the nutritionally related factors, a reduced level of growth hormone has also been described as a cause of the frequently observed cases of stunted growth [5]. More far-reaching disorders of basic

function such as temperature regulation or breathing can also occur in severely disabled children.

### Diplegia

In diplegic patients the lower extremities, in particular, are affected (■ Fig. 4.119). The trunk and neck muscles are normotonic and the patients develop good trunk control. Frequently, however, the two upper extremities are also affected to a mild extent. The onset of walking is often delayed, although the patients are usually capable of walking.

### Hemiparesis

In patients with hemiparesis, the arm and the leg on the same side are mainly affected (■ Fig. 4.120). As with diplegia, innervation of the trunk muscles remains almost normal and these patients also develop good trunk control. On closer examination, the extremities on the other side also usually show slight functional problems. Since the patients are able to compensate, with their normal side, for the restrictions of the affected side, psychomotor development is either not delayed at all or only slight delayed.

### Other forms

Some authors make a further distinction, referring to tri paresis (usually both legs and one arm affected) or monoparesis. Most patients however can be classified in one of the first three categories.

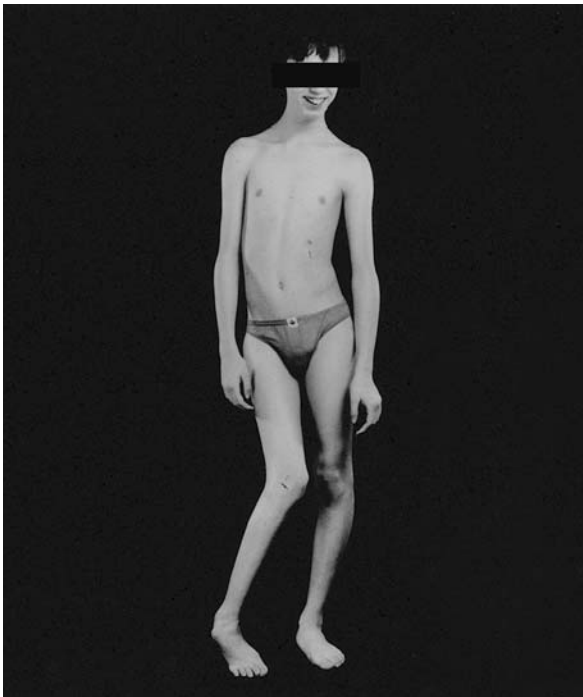
In addition to the topical classification, another *sub-division*, based on the *prevailing neurological signs and symptoms*, is useful: Depending on the muscle tone and muscle activity, we can distinguish between spastic, dystonic, atactic and athetotic clinical pictures.

### Treatment

Increased tone and spasticity block the patient's motor functions, making controlled voluntary movements impossible. This hinders not only the motor development, but ultimately the overall development of the patient. Reducing the muscle tone in spastic patients with muscle hypertonia is therefore a crucial *therapeutic* objective. The various »therapies« (physical therapy, occupational therapy, speech therapy etc.) promote psychomotor development and help prevent secondary deformities. Braces that stabilize the skeleton in an anatomical position during weight-bearing work towards the same objective. Since limited time is available for treatment sessions, braces should be used wherever possible so that the valuable treatment time is left free for measures that cannot be conveyed in any other way. Other conservative and surgical measures are designed to correct any functionally troublesome deformities, balance muscle forces and provide stability. The longer deformities persist, however, the greater the additional adaptive changes, increasing the time and effort required for corrections and rehabilitation.



■ Fig. 4.119. Patient with *spastic diplegia*. Typical *crouch position* of the legs with slight internal rotation, good trunk control



■ Fig. 4.120. Patient with right-sided *spastic hemiparesis*. The leg on the affected side is internally rotated and the knee and hip are flexed. The patient stands in an equinus position

Spasticity often conceals muscle weakness, which changes the therapeutic plan considerably: Power training is provided instead of stretching exercises and muscle weakening measures. For this reason the treatment of patients that can walk should, wherever possible, be based on gait analysis data.

### Prognosis

Since, from the neurological standpoint, cerebral palsy involves non-progressive damage to the brain, deterioration in the neurological picture can be ruled out. On the other hand, the cerebral damage will not fully recover. The basic prognosis for the clinical condition therefore depends on the extent of the primary neurological damage, which also restricts the life expectancy. Twenty-year survival rates of 99% and 50% have been observed for slightly disabled and severely disabled patients respectively [6].

Regardless of the life expectancy, the question of the orthopaedic prognosis is frequently raised, particularly in respect of the ability to walk. This is where the clinically topical classification is helpful. Patients with good trunk control, i.e. patients with hemiparesis or diplegia, usually learn how to walk. The onset of walking is generally delayed to a greater or lesser extent, however, depending on the severity of the impairment in each case. Walking freely is often difficult for patients with tetraparesis, and only those with mild symptoms will be able to acquire this skill. In many cases, balance is impaired by deficient trunk and head control to such an extent that the patients required added support (crutches, canes or a walker). In any case, many patients are at least able to achieve a transfer function in this way. Only the most severely disabled patients are hardly able to walk or stand at all, although here too any prognosis should be made very carefully. It is often asserted that walking can no longer be learned after the age of seven.

Clinical experience has taught us, however, that it is perfectly possible to achieve a certain ability to walk beyond this age. One needs to define »walking« in this context: It is, of course, highly unlikely that patients of this age will be able to acquire the walking ability of neurologically normal individuals. The important point is that they can learn how to move their own weight over a few meters, perhaps only with support. The chances of acquiring such a limited walking function at this stage are particularly good if any existing orthopaedic deformities such as hip dislocations or severe contractures can be corrected. Since a motor-related prognosis is unreliable in this context, we are particularly circumspect when making any statements, particularly negative ones. Bluntly informing parents that »your child will never learn to walk« is demotivating and jeopardizes many therapeutic steps. If functional progress is subsequently made, contrary to the prognosis, the parents' confidence in medicine will be shaken.

### 4.7.3.2 Subsequent brain damage

The brain can also be damaged after it has fully matured. The commonest causes are accidents with craniocerebral injuries, infections, hemorrhages and resuscitation. In contrast with the situation for cerebral palsy, a fairly pronounced and troublesome spasticity develops. On the other hand, patients are able to draw on their experience of life before the brain damage occurred. As a result, some astonishing results are occasionally achieved despite the presence of severe damage. Treatment is based on the principles described in ► chapter 4.7.4.1.

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## 4.7.4 Spinal cord lesions

R. Brunner

### 4.7.4.1 Myelomeningocele

#### Definition

Myelomeningocele involves a cleft malformation in which the vertebral arches are not closed, the dura either protrudes in a sack-like manner or is simply exposed, and the spinal cord has not closed to form the neural tube. Neurological function is impaired at the level of the myelomeningocele and distally. Motor function (usually as flaccid paralysis), sensitivity and bladder and bowel function are affected.

— **Synonyms:** Spina bifida

— **Common abbreviation:** MMC

#### Etiology and pathogenesis

Myelomeningocele is the most common disorder of the spinal cord during childhood. Although the cause of the cleft malformation is not known, a multifactorial process is probably involved: Myelomeningocele, together with anencephaly, tends to run in families. Social and environmental factors also play a role. Thus, an increased incidence is observed in the lower social classes [42]. Diet is also important, and folic acid in particular is known to be capable of preventing the condition [36, 53]. The frequency of neural tube defects appears to be on the decline [34]. A myelomeningocele can develop either as a result of failed closure of the neural tube or a rupture of a closed neural tube [35].

#### Historical background, occurrence

Morgagni described cases of myelomeningocele as early as 1769. In 1886 Von Recklinghausen observed that it occurred as a result of failed closure of the neural tube [61]. The *incidence* of myelomeningocele is dependent on region and race. Whereas the incidence in Sweden is 72/100,000 inhabitants [24], the figures cited for England are around 300/100,000 [30, 33]. An incidence of up to 100/100,000 has been calculated for the United States [54, 63]. The white population is 3.6 times more often affected than blacks [22]. Both sexes are affected with equal frequency [14, 22].

#### Clinical features and diagnosis

Myelomeningocele must be differentiated from cleft malformations without any involvement of the spinal cord (meningoceles, exclusively bony defects). No neurological changes are expected in these patients [59]. In a patient with myelomeningocele the spinal canal, meninges and neural tube are open. The cele may be completely exposed



■ Fig. 4.121. Skin-covered *myelomeningocele*

or covered by a thin membrane that becomes covered with skin over time (■ Fig. 4.121).

The *spine* is always deformed. The deformity may involve just the spinal canal, although segmentation defects with bar formations and hemivertebrae are also possible. In addition to neurogenic scoliosis resulting from persisting paresis, congenital scoliosis or combinations of the two can occur. If the spinal canal is wide open, the muscles that are normally located behind the vertebrae move in front of the load-bearing axis of the vertebral bodies, resulting in the absence of functional posterior tensioning and the development of extreme hyperkyphosis. Even after closure of the cele, patients with myelomeningocele can show *neurological symptoms* at the level of the cele and below. Most patients show flaccid paresis of the muscles of the locomotor system, combined with hypesthesia or anesthesia. Spasticity is not typical. The tendon reflexes are usually diminished or absent. Urinary and fecal incontinence are present. The neurological symptoms usually remain unchanged or show only slight improvement during the first few months of life. A scarred adhesion of the spinal cord or individual roots in connection with a tethered cord syndrome (▶ Chapter 4.7.4) is part of

the pathology, but usually remains asymptomatic [39]. If spasticity or neurological deterioration occurs, investigation for a possible tethered cord or additional medullary malformations (e.g. syrinx, diastematomyelia etc.) are indicated.

After surgical correction of the pathology underlying the spinal cord adhesions, the neurological deterioration may either regress or remain unchanged [4, 18]. Since it is important therefore to identify any neurological changes as soon as possible, we recommend an annual muscle status assessment. Only in this way will it be possible to identify the onset of any weaknesses before the compensatory mechanisms finally collapse and an obvious functional deficit appears.

A positive effect even on existing spinal malformations has been described after »detethering« [9, 47]. *Concomitant malformations* include the *Arnold-Chiari malformation* (caudal displacement of the medulla oblongata with herniated cerebellar tonsils) and *Walker cysts* (dilations of the fourth ventricle), which is the most common additional abnormality of the CNS. Magnetic resonance imaging should be arranged before any spinal procedure and for all patients within the first 8–10 years of life since accompanying pathologies, including within the spinal cord, are common. The frequency of *epilepsy* in patients without hydrocephalus is approx. 2%, but approx. 22% in those with hydrocephalus [10]. In one investigation a dysfunction of the upper extremities was found in 46% of patients, regardless of the presence and treatment of hydrocephalus. Only 3% showed normal neuromotor development [28]. In a study involving 527 children, 32% were able to walk without assistance, 60% had urinary incontinence, and 4% were completely incontinent. 70% of the children were severely disabled [25]. Intelligence is often diminished, although a normal IQ was measured in 76% of the patients in the same study [25]. Other authors have found significantly reduced IQ levels in patients without hydrocephalus, and the functioning of the upper extremities was also worse than that of patients without myelomeningocele [37].

In addition to all these neurogenic problems, the possibility of latex allergy should also be mentioned, as this is common in patients with spina bifida and results from increased antibody formation [11].

### Treatment

Left untreated, 86% of patients with myelomeningocele will die during the first year of life [23]. The celes should therefore be closed immediately after birth or, at the latest, within the first few days of life. If hydrocephalus is also present, a drain must be inserted at an early stage in order to prevent any pressure-related brain damage. The predominant factor in patients with myelomeningocele is usually the flaccid paresis (50%). The other patients show a mixed pattern of spasticity and flaccid paresis of



the upper and lower extremities or spasticity on its own. The impaired neuromuscular function alters the forces that act on the growing musculoskeletal system, causing secondary deformities to develop. The pareses, combined with the loss of sensory functions in the area of the affected segments, lead to a loss of dynamic stabilization of the individual joints, e.g. the hip, which can then work loose and dislocate.

The paresis also always affects the muscles at and below the level of paralysis. The resulting dynamic muscle insufficiency explains the high risk of spinal deformities, particularly scolioses, even if malformation is not present. Particular caution is indicated in this context since the mobility of the lumbar spine, which has to be sacrificed during surgical corrections, is needed for numerous everyday activities (even for putting shoes on for example). The patient's functions must, wherever possible, be restored via the correction of deformities.

All patients with paralyzes tend to develop skeletal deformities of varying degree that also restrict function. Braces must be used and operations performed in order to create the best possible conditions for the rehabilitation of the patients. The treatment of the individual orthopaedic problems is addressed in the respective chapters for the specific areas of the body.

*Rehabilitation* is designed to promote the age-matched development of motor, psychological and social skills. Ideally, patients should be able to perform the same activities as healthy children of the same age. For this reasons, braces are indicated from an early age (from 1 1/2 years) in order to enable the child to stand. Deformities can occur at any time and require correction, particularly if they interfere with function. Parents and patients are often anxious about major operations on the hips or the spine. However, given the therapeutic objective of normal psychomotor development it does not seem a good idea on the basis of this anxiety to forbid children from undertaking activities in order to prevent the development of deformities (in order to prevent a hip dislocation for example), and thus at the same time delay their development.

While children must develop at their own pace and this process can only be assisted, but not replaced, by treatment, nowadays skeletal deformities can be corrected, albeit with considerable time and effort. The restriction of motor skills that is present in any case as a result of the myelomeningocele always leads to a focal loss, of varying degree, of other abilities (e.g. in perception or cognition), which requires a corresponding program of physical therapy, occupational therapy and education. This extensive range of therapeutic services often means that the patient has to attend special schools. Regular medical check-ups are required, particularly during the years of growth, in order to monitor, inter alia, the orthopaedic situation, the urinary tract and the neurologi-

cal system. The patients must be encouraged gradually to take responsibility for themselves, learning how to manage their orthoses on their own or catheterize themselves. Only then will they be able subsequently to lead a normal life with maximum independence. The intensive rehabilitation often interferes with the quality of life of the individual patients and places a great strain on the family and healthcare system. Tests are now available for diagnosing a myelomeningocele at an early stage, within the uterus, on the basis of the investigation of the amniotic fluid [45, 62].

From the orthopaedic standpoint the most relevant issue in children with myelomeningocele is their *ability to walk*. The most important factor is the level of the neurological lesion. Whereas patients with thoracic or high lumbar myelomeningoceles are barely able to walk, approx. half of patients with lesions at L 3, two thirds of patients with lesions at L 4, and four fifths of patients with lower level lesions are able to walk [48]. Authors of various studies have attempted to establish parameters for effectively predicting the subsequent ability to walk. The level of the neurological lesion is just one parameter [2, 8, 19, 58], whereas other parameters allow a better prognosis to be made at a more functional level (e.g. the ability to walk freely outside the house compared to the use of a wheelchair at the age of 7 years [17], or the development of good sitting balance [57, 58].

An important requirement for the ability to walk is the presence of sufficient power in at least one muscle group that controls the upright position. Since both plantar flexors and hip extensors are innervated by nerves located at a low level, the knee extensors constitute the last functionally relevant muscle group with a higher innervation level that are able to compensate for the failure of the other postural muscles are. Powerful knee extensors are therefore a precondition for free walking [50], and it is particularly important to avoid the scenario in which a growing knee flexion deformity overtaxes these muscles.

We know that children with myelomeningocele learn how to walk at a later age than normal [57]. One important reason for this is the inability of patients with paralyzes to perceive at all the ground on which they are supposed to walk or some part of the legs, which they are supposed to keep upright. They are able to control their lower extremities only indirectly, which places much greater requirements on the balance function. The development of the balance reactions is delayed. In fact, the balance reactions are often worse than those in patients suffering from traumatically induced paraplegia, as shown by the fact that the latter patients are often able to walk, at least for short distances, even when the lesion is at a fairly high, i.e. thoracic, level. This is rarely possible for patients with a myelomeningocele at the same level.



#### 4.7.4 · Spinal cord lesions

Walking ability should be the goal for every patient wherever possible. *Braces* of various kinds and/or operations are usually required to enable patients to stand and walk. They replace the missing muscle power, prevent or correct deformities of the musculoskeletal system and provide stability. It is useful therefore to provide even small children, i.e. at the age of 1–2 years, with orthoses in order to induce standing and walking. This is important even if transferability is the only future objective, since balance, body control and muscle power must be developed for this function as well. Patients who are capable of walking suffer fewer fractures and fewer pressure points than those confined to a wheelchair. On the other hand, more energy is required for locomotion by walking compared to locomotion in a wheelchair [1, 15]. Locomotion with a swing-through gait is only slightly less favorable than reciprocal walking in terms of energy use [38]. In any case, the increased energy consumption of walking obviously causes the patients to become more fatigued [18].

The shoulders are also unable to cope with the strain over the years and patients develop painful arthritis of the shoulder. An appropriate balance must therefore be established between walking ability and locomotion in the wheelchair. We know that patients lose their walking ability in the long term [8], partly as a result of skeletal deformities and partly no doubt based on the extent of the braces and the actual purpose of walking. In our experience, patients who walk for sporting or therapeutic purposes tend to lose their ability to walk when they take up employment.

On the other hand, myelomeningocele patients who use their walking ability day-in, day-out for beneficial routine activities tend to remain on their legs. A daily routine must therefore be developed during rehabilitation that requires beneficial walking by the patient. But adapting high-fitting orthoses in particular to the needs of everyday life can be very difficult, if not impossible (■ Fig. 4.122). Much better preconditions can therefore be achieved with orthoses that do not extend above the knee than high-fitting braces, and the long-term prognosis in respect of walking is better.

Small children with high-level lesions should initially be fitted with rigid *Hip guidance orthoses* (walking braces that secure the pelvis and lumbar spine and extending down to both feet) and perform balancing exercises while standing. Walking is subsequently introduced, first by means of braces such as the Swivel Walker and later with Hip guidance orthoses with a rigid frame (e.g. Parawalker) in combination with a rollator and, later, crutches or canes (► Chapter 4.7.2). But walking is difficult to maintain in the long term with high-fitting orthoses.

As regards the design of high-fitting orthoses that include the pelvis (Hip guidance orthoses), micturition control represents a major problem in patients with high myelomeningoceles. The rigidity required for walking is



■ Fig. 4.122. Patient with *myelomeningocele* who is able to walk with a rollator in conjunction with an *orthosis* that secures the distal trunk and both legs

technically incompatible with adequate abduction of the hips or good practicality of the appliance. If patients have to catheterize themselves or empty their bladder several times a day, the Hip guidance orthosis will usually have to be removed for this purpose. Whereas patients without an Hip guidance orthosis can empty their bladder on their own, those with such an orthosis are reliant on a helper for removal and re-fitting. In such situations emptying the bladder is just too time-consuming, and these high-fitting Hip guidance orthoses are eventually discarded and the patients take to their wheelchair.

Optimal adaptation of braces must be attempted in each individual patient. It may be appropriate to accept a »less attractive« gait pattern as a compromise and provide the patient just with ankle-foot orthoses. These should be adapted even in early childhood, since the muscles and body control must be trained accordingly. Patients with paresis of the plantar flexors and knee extensors are still able to walk with ankle-foot orthoses. Rigid ankle-foot

orthoses replace the loss of the plantar flexors and extend the knee indirectly, thereby ensuring walking function in the long term [5, 27].

Alternatively, a rearfoot arthrodesis after completion of growth can provide the same stability. Ankle-foot orthoses can even be attempted in patients with deficient hip stability, although high-fitting Hip guidance orthoses will need to be fitted if no hip stability is present. These orthoses incorporate a reciprocal gait mechanism, i.e. when the weight is taken off one leg, this leg is swung forward by the orthosis itself. In this way, the orthosis produces a step almost by itself. The patient must learn how to shift the weight onto one leg in order to take the weight off the other and thus allow a forward swing. Of course, such orthoses work only if the frame is completely rigid and does not give way when the weight is transferred, which makes them difficult for adult patients to use.

Another aspect that can significantly interfere with walking is the *development of deformities*. External or internal rotational deformities of the lower leg are common. As a result of such deformities, the feet do no longer face the direction of walking and the necessary postural function is not ensured. These rotational deformities can progress rapidly once they reach a certain level. Other adverse deformities are those of the feet, which lead to pressure points, and deformities of the spine, which patients with poor balance are unable to compensate for properly [13, 20, 57]. Functionally relevant deformities must therefore be treated and corrected, particularly in patients who possess some degree of walking ability.

On the other hand, studies have shown that hip dislocations do not interfere with walking ability [52, 60]. Nor do the kinematic gait data depend on the centering of the hips [60]. The therapeutic options are described in the individual chapters for the specific areas of the body. Patients who remain on their legs are less susceptible to *fractures*. Apart from actual fractures, epiphyseal separations can occur, particularly in the lower leg, and can lead to extensive periosteal reactions. However, such skeletal injuries heal rapidly and only require short-term immobilization, which can also soon be ensured again with an orthosis. Long periods of immobilization must be avoided because it can lead to a loss of walking ability [17].

The myelomeningocele is surgically closed at an early stage. Urinary and fecal incontinence is common and the tendency for musculoskeletal deformities to develop leads to frequent operations. For this reason, patients often come into contact with rubber, particularly latex. *Latex allergies* are therefore a frequent problem [12, 40, 41]. Frequencies of between 18% and 50% have been cited for latex antibodies in these patients. These latex allergies can be life-threatening in some cases, since anaphylactic and severe allergic reactions can occur. At any rate, caution is always indicated when using rubber products and latex.

#### 4.7.4.2 Spinal cord injuries

##### ➤ Definition

Permanent, complete or incomplete damage to the spinal cord as a result of an accident or as a complication of surgical procedures.

Fortunately, *accidents* that produce spinal cord injuries are rare. The spinal cord can also suffer damage without any detectable signs of actual spinal injury (SCIWORA syndrome: »spinal cord injury without radiographic abnormality«). In the event of fractures, the reduction and any stabilization are implemented immediately in order to create the best conditions for neurological recovery (► Chapter 3.1.11), although residual neurological deficits can still occur.

Spinal cord damage can also occur as *complications of surgical procedures*. While the spinal cord can be injured directly during *spinal operations*, more often the cause is an adhesion of the spinal cord in the spinal canal, which can then damage the spinal cord via traction if distraction is used in the correction of deformities. Investigation of the spinal canal is therefore indicated before procedures for correcting deformities. Finally, the artery of Adamkiewicz, which supplies the anterior sections of the spinal cord can also be compressed, resulting in a motor paraplegia.

During *operations on the major vessels and the heart* the temporary loss of peripheral perfusion can cause the blood supply to the spinal cord to fall below the required level, potentially resulting in permanent complete or incomplete lesions. These spinal cord injuries can be differentiated symptomatically from the neurological signs in myelomeningocele by the fact that a clearly identifiable neurological level is usually present. These paraplegias also tend more towards spasticity, particularly if the lesion of the spinal cord occurs in later childhood. The orthopaedic problems and the principles of the neuro-orthopaedic and rehabilitative measures are identical to those applicable to myelomeningocele (► Chapter 4.7.4.1).

#### 4.7.4.3 Tethered cord

##### ➤ Definition

A tethered cord is considered to be present if the medullary cone is below the level of L2 after the neonatal period. Fixation of the spinal cord prevents the upward physiological shifting of the sacral end of the spinal cord during growth.

##### Pathogenesis

The fixation of the spinal cord may be caused by a thickened, and thus excessively rigid terminal filum. Alternatively, malformations of the spinal canal and sacral agenesis, intraspinal lipomas or deformities of the spinal cord

#### 4.7.4 · Spinal cord lesions

itself (syrinx, diastematomyelia, hydromyelia, diplomyelia etc.) can also lead to a tethered cord [6, 26, 32, 43, 46, 49]. The spinal cord can also become adherent as a result of scarring after trauma or surgery [39, 44]. Or an intraspinal dermoid can produce the same effect. Since these structures fix the spinal cord they place the cord under tension during growth, thereby producing a deterioration in neurological function.

##### **Clinical features and diagnosis**

In a patient with a primary (not postoperative or post-traumatic) tethered cord, skin changes over the spine (lipomatous subcutaneous tumors, atypical hairs and nevi) are very common [6, 56]. Clinical signs and symptoms are urinary incontinence, pains in the back or legs and foot or spinal deformities [6, 7, 26, 51].

The symptoms may not appear until adulthood, and one case of initial onset at the age of 70 has even been described [3, 21]. The adhesions are usually at the lumbosacral, although cervical adhesions also occur [55]. In patients with existing neurological deficits and no progressive underlying neurological condition (e.g. with a myelomeningocele or posttraumatic paraplegia) the possibility of a tethered cord must be considered if the neurological situation deteriorates. The easiest way to confirm the diagnosis is by an *MRI scan*. A sonogram may also be considered for neonatal patients [6, 32, 49, 51].

##### **Treatment**

Surgical release of the adhesions of the spinal cord can lead to a (partial) recovery of the neurological syndrome [31, 49], although recurrences are not infrequent [7]. Functional deficits are managed according to the principles outlined in ► Chapter 4.7.4.1.

#### 4.7.4.4 Other spinal cord lesions

Other pathological changes of the spinal cords such as the formation of cavities (*syringomyelia*), *tumors* or *inflammation* can lead to neurological deficits. The cause of the underlying condition must first be treated. Any residual neurological deficits that result in dysfunction are managed as for myelomeningoceles in terms of neuro-orthopaedic and rehabilitative measures.

#### 4.7.4.5 Poliomyelitis

##### **► Definition**

Viral infection of the spinal cord, particularly of the anterior horn cells, that causes neurological symptoms in 1–2% of affected patients. Thanks to vaccination campaigns, the disease has largely been eradicated, although the *post-polio syndrome* in particular is still relevant today.

##### **Acute phase**

##### **Clinical features, diagnosis**

Viral infection with an incubation period of 3–20 days, with an initially uncharacteristic febrile peak during the first stage of the illness, followed by a latency period and another febrile main phase. Signs and symptoms during the main phase include headache, meningism and a feeling of illness, together with progressive paralyses. Pain and tenderness of the muscles can occur, although sensory problems do not occur in poliomyelitis. *Differential diagnosis*: One other diagnosis that must be ruled out is *Guillain-Barré syndrome*.

##### **Prognosis**

In patients with bulbar involvement the prognosis is poor, with mortality rates of up to 50%. Otherwise, the paralyses only recover partially, leaving motor pareses with muscle atrophy and areflexia. Spasticity does not occur in the post-polio syndrome, nor do the paralyses progress.

##### **Post-polio syndrome**

While the cause of the viral infection has almost been completely eradicated in the developed world, the *sequelae* are still having to be managed. These typically include flaccid pareses. In functional terms, the legs (foot muscles, particularly the plantar flexors, knee extensors and hip extensors) and trunk (spinal muscles) are especially affected.

The patients develop *compensatory* and adaptive mechanisms for their disability. Surgery for severe *scolioses* and stiffening of the spine may therefore be contraindicated for reasons related to independence and the ability to perform everyday tasks. Thus, for example, whereas patients with a scoliotic, but mobile, spine are able to tie their shoelaces, this simple task may become impossible after a procedure which results in a stiffened spine. Patients thus become dependent on helpers. A functional trial with a plaster corset is therefore advisable preoperatively, as this will show the patient what the situation will be like postoperatively.

In addition to paresis of the paravertebral muscles, patients may develop a scoliosis during childhood for another reason after poliomyelitis: The pareses regularly result in a pronounced *leg length discrepancy*. If the hip muscles are also paretic on one side, a unilateral atrophy will develop, resulting in constant pelvic obliquity, even in a sitting position. In such cases, and in addition to a leg lengthening procedure, a cushion is required to compensate for the unilateral muscle atrophy and place the pelvis in a horizontal position.

If *paresis of the triceps surae muscle* is present an *equinus foot* produces a positive functional effect since it fixes the lower leg in relation to the foot. A weak triceps

muscle without contracture causes the lower leg to sink when the patient leans forward, resulting in a crouch gait. Contractures of the knee and hip flexors can then develop and further restrict the patient's walking ability. Such consequences can be particularly serious if the power of the knee extensors is also diminished. The patient eventually has to support the knee with his hand during walking. The hand is no longer free and the patient is unable to maintain an upright position.

A *footdrop* does not cause much functional disruption. Since the lack of dorsiflexion is compensated for by the leg shortening, patients do not need to circumduct. The gait pattern is unattractive from the esthetic standpoint however. One therapeutic measure is a posterior leaf spring ankle-foot orthosis, although this is rejected by most patients.

If *paresis of the knee extensors* is present, a slight hyperextension of the knee causes the knee to lock during the stance phase and thus allow upright walking without braces. The precondition for this is an active triceps surae or the stabilization of the ankles by a slight equinus foot position or a rigid ankle-foot orthosis. The lack of dynamic stability for the ankles will result, in turn, in a crouch gait (■ Fig. 3.436, ► Chapter 3.4.10).

If *paresis of the hip muscles* is present, the leg must be stabilized during walking, even in the initial stance phase, by extension. To achieve forward motion, the patients must swing the leg forward by means of compensatory movements at spinal level. It is amazing to see how patients with extensive paresis of the leg muscles are able to walk freely without braces thanks to these compensatory mechanisms. A precondition in these cases is a slight contracture of the triceps surae and slight hyperextension at the knee.

Since some *muscles are still innervated* – even in cases of partial paresis – they can be trained and thus produce functional improvement. Pain syndromes, even though these tend to occur during adulthood as a result of excessive stressing of the musculoskeletal system, can also be positively influenced by a muscle strengthening program with cardiovascular training [16]. *Joint dislocations* rarely occur in a post-polio syndrome even though the skeleton is fairly delicate and deformed. This is probably because sensation is not affected. Pronounced shortening of the affected extremity by 4–5 cm is typical however, particularly if the poliomyelitis was contracted during early childhood. This results in a shortening limp and a scoliotic spinal posture, which may become fixed.

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### 4.7.5 Nerve lesions outside the central nervous system

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#### ► Definition

Nerve lesions outside the central nervous system involve the spinal nerve roots, the peripheral nerves and the anatomical structures in plexus form located between the two. Since the lesions affect only the axons of the nerve cells the spontaneous recovery rates are good provided the key structures remain intact. The lesions are subdivided into plexus palsies and peripheral nerve injuries.

#### Etiology and pathogenesis

Peripheral neural structures in children can be damaged as a result of a variety of injuries. The *noxae* may produce a direct *sharp* (e.g. by cutting) or *blunt* injury (by pressure or, indirectly, by traction). *Accidents* are by far the most common cause of these lesions. Unfortunately, damage can also occur during birth or as a result of therapeutic measures. Nerves can be injured by conservative treatments such as plaster casts or dynamic splints (peroneal nerve palsy as a result of pressure exerted by a cast on the fibular head is a familiar example).

Damage to the peripheral nervous system can also occur during major treatments such as limb lengthening procedures or, during surgery, by positioning aids or surgical instruments. The lesion of nerve structures outside the CNS can vary according to the frequency and intensity of the noxa. A pathophysiological distinction is made between a neurapraxia, an axonotmesis and a neurotmesis.

In *neurapraxia* the neuronal structures show a functional deficit, but remain intact in terms of their continuity. The function subsequently recovers completely. In *axonotmesis* the axons are interrupted, although the key structures (myelin sheaths) remain intact. The axons regrow at a rate of 1 mm/day along the key structures from the proximal end. If the latter are interrupted or refixed with staggered alignment, defective innervation results. Proximal lesions are particularly prone to this risk. Motor fibers encounter e.g. incorrect muscle groups or even sensory end organs. Neurotmesis refers to the complete severance of the axons and key structures. No regeneration takes place. *Paralysis* may be complete or incomplete. An incomplete paralysis of a nerve is known as a *paresis*. In everyday clinical practice, however, these terms are not kept strictly separate and in some cases are used as synonyms.

#### Clinical features and diagnosis

Sensory and motor function are impaired or completely absent in the area of the affected nerve or nerve roots.

The typical areas of skin and muscle supplied by the relevant nerve allow a topical diagnosis to be made (► Chapter 2.1.2). *Sensitivity* may be completely absent (anesthetic), reduced (hypesthetic) or even increased (hyperesthetic). The *muscle dysfunction* can also manifest itself in the form of weakness or complete paresis. The *pareses* are always flaccid, and spasticity never occurs. The presence of sensation and/or motor function rules out a complete nerve lesion. But if complete lesions are present, only the course of the condition will show whether the neuronal structures are actually interrupted or not.

The *electromyogram* can show fine motor impulses that are not clinically detectable and thus enable a more accurate diagnosis to be made. The EMG can also be used for monitoring regeneration. The neural structures can be depicted directly on an MRI scan. Moreover, in the case of plexus injuries, the roots can be shown in their pouches by myelography or MRI.

#### Treatment and prognosis

Measures for nerve lesions are basically curative or palliative.

A *curative* procedure is suturing of the damaged nerve, with or without interposition. The prognosis is better for early than for late interventions and better for the upper limbs than for the lower. On a proximal extremity, a success rate of 80–90% can be expected after early reconstruction. The results are not so good for a lower extremity, particularly if the peroneal nerve is involved [1, 6, 10]. For secondary procedures, the results are worse by 10–20% [1, 7, 8].

*Prognostically negative factors* in relation to the results after reconstructive operations:

- proximal lesion,
- increasing age of the patient,
- increasing time to the intervention,
- extension of the lesion,
- certain nerves (e.g. sciatic nerve, peroneal nerve).

Contractures and movement restrictions in the affected section of the extremity must be prevented so that the muscles can be reused in the optimal manner after any re-innervation. Consequently, physical and/or occupational therapy are indicated during rehabilitation.

Reconstructive procedures on nerves are not always possible or successful. The patients may remain functionally impaired as a result of *muscle deficits*. If severe functional deficits are present, a muscle transfer is a possible palliative measure, but a complete recovery is never possible. We know that a muscle loses some of its power when it is transferred [2, 11]. But modern microsurgical anastomosis techniques now provide the option of a free muscle transfer in which a power generator is transferred from a remote site (where the deficit does not matter so much) to the desired location.

## 4.7.6 · Muscle disorders

Basic requirements for an optimal functional result [2, 4]:

- The muscle anatomy of the transferred muscle must match that of the replaced muscle as closely as possible so that it can exert its full power at a length at which the muscle is used in everyday life.
- Normal innervation of the transferred muscle and good motor coordination.
- Correct phasic activity of the transferred muscle (this is easier to achieve for a former synergist than for a former antagonist [3, 15]).
- Adequate power of the transferred muscle.
- Skeletal deformities compromise the results of muscle transfers
- Redirections over which the muscle operates after its transfer are undesirable since the muscle loses some power as a result.
- After the transfer the muscle should remain under tension even at rest so that it does not have to adapt to any overlength by fiber shortening, as happens after tendon lengthening procedures, with a corresponding loss of power.
- Scar adhesions can hinder the functioning of the transferred muscle despite ideal preconditions, and are particularly common where a tendon passes through fascia or bone channels. Early mobilization should therefore be attempted subject to adequate stability of the reinsertion.
- The gain in function must compensate for the loss at the site of origin of the transferred muscle.

Another therapeutic option involves the optimal adjustment of the existing active range of motion for everyday functions. Osteotomies are suitable for this purpose.

In addition to functional motor deficits, *sensory deficits* can also remain, but these are usually tolerated by the patient. However, *pain syndromes* can develop in very proximal injuries or root avulsions. In these cases neurotization can prove helpful. In this procedure, a nerve with a less important function is attached to the distal stump or stumps. A functional gain is extremely unlikely however [12, 13].

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### 4.7.6 Muscle disorders

R. Brunner

#### General information

##### ➤ Definition

The myopathies comprise a group of diseases that exclusively affect the muscles. Characteristic signs and symptoms are a slowly progressing muscle weakness, rapid fatigability and diminished reflexes. Many myopathies tend to run in families, although they can also occur concomitantly with other underlying illnesses (e.g. metabolic or hormonal disorders). Sensory perception remains normal.

#### Classification, clinical features and diagnosis

The *classification* involves dystrophic muscle disorders, contraction disorders, inflammatory conditions and muscle symptoms associated with other underlying diseases. An overview is presented below. The key feature of muscle disorders is *muscle weakness* of varying severity and progression. Rapid fatigability during sport and generally during physical exertion is also typical. At the same time reflexes are diminished. *Sensory perception*, however, is fully preserved. The muscle involvement is usually symmetrical. In some types, the ability to walk is delayed or, in severe forms, does not even develop.

### Classification of myopathies

1. Muscular dystrophies
  - Progressive forms:
    - Pelvic girdle form: Duchenne type
    - Pelvic girdle form: Becker type
    - Facioscapulohumeral form
    - Emery-Dreifuss type
    - Limb-girdle type
  - Curschmann-Steinert myotonic dystrophy
  - Other
2. Congenital muscular dystrophies
  - Central core disease
  - Minicore myopathy
  - Mitochondrial myopathies
  - Other
3. Spinal muscular atrophy SMA
  - Severe SMA (Werdnig-Hoffmann)
  - Intermediate SMA
  - Mild SMA (Kugelberg-Welander)
4. Hereditary motor and sensory neuropathy (peroneal muscular atrophy, Charcot-Marie-Tooth)
  - HMSN 1
  - HMSN 2
  - HMSN 3
5. Myotonias
6. Myasthenia gravis pseudoparalytica
7. Secondary myopathies

Myopathies usually progress over several years or decades. Spontaneous pain is rarely present, except in inflammatory and necrotic conditions. With the loss of muscle function, the patients also lose active control of their joints, although sensory perception remains intact. However, the joints and muscles can still be moved into extreme positions. Healthy individuals protect the joint capsules and muscles in these positions by contracting the muscles before the joint is overstretched. Since stretching of the soft tissues may elicit pain if muscle activity is lost, simply testing the patient's range of motion can prove painful.

Additional diagnostic tools include the *measurement of muscle enzymes* and the *EMG*. Ultimate diagnostic confirmation is provided by *molecular genetic investigation* or a *muscle biopsy*. If there is evidence to suggest the presence of some other underlying condition (inflammation, metabolic disorder, etc.), corresponding investigations must be arranged.

### Treatment and prognosis

Since a *causal treatment* for muscular dystrophies is not currently available, the treatment must be limited to the secondary effects. With the primary dystrophic muscle disorders, in particular, there is a substantial risk that

the patients will suffer functional impairment in their everyday lives.

The ability to *walk, stand*, and ultimately even to *sit* may be lost in severe forms. As their muscle weakness progresses, patients increasingly spend their time in a seated position in order to be able to participate in everyday life. The inability to extend the joints in the lower limbs involves the risk of flexion contractures of the hips and knees, which further interferes with the ability to stand. The residual muscle power is no longer able to counteract the force of gravity. When the joints are fully extended, this function can be taken over by the capsules and ligaments. Contractures therefore lead to a premature loss of abilities. This results in the development of secondary scolioses which, in turn, exacerbates the patient's disability. It is important therefore for patients to be supported at trunk level and to retain full extension of their knees and hips. In this way, they can lock these joints while standing by slight overextension, thus enabling an erect position to be achieved without substantial muscle work.

The only deformity that provides the patient with a functional benefit is an equinus foot. The shortening of the triceps surae muscle prevents dorsiflexion at the ankles and thus a forward dropping of the lower leg while standing. As a result, the patients do not need to stand in an energy-consuming crouch position. *A slight equinus foot position (5°–10°) must be left as is since it serves as a functional replacement for the missing triceps activity.*

In muscular dystrophy patients with a poor prognosis (particularly in patients with Duchenne type muscular dystrophy), every effort should be made to prolong the ability to walk and stand in order to postpone the development of a scoliosis. For this reason, early surgical treatment at the age of 6–8 years has been proposed [22]. In the corresponding procedure, the knee flexors, foot dorsiflexors and hip abductors undergo aponeurotic lengthening, and a section of the iliotibial tract is resected. This represents an extensive operation in a child with few symptoms who is usually still able to walk freely at this stage [6, 29]. Although statistics indicate that walking ability can be extended by 1–2 years as a result of this procedure [6, 9], in our experience neither the parents nor the children themselves can be persuaded to agree to such an operation at this point. For some years now we have been increasing our conservative efforts in cases where only a few degrees of extension of hip or knee are lost. In these patients functionally relevant deformities have been reliably prevented, thus reducing the need for functional surgical corrections to occasional cases only.

For all those involved the need for surgery is more readily appreciated when contractures occur and when these start to restrict the mobility and everyday activities of the patient [28]. While the ability to walk and stand can still be prolonged at this stage, the effect is slightly weaker than after an early operation. Every surgical procedure,

## 4.7.6 · Muscle disorders

however, involves a temporary loss of power, which must be countered during a rehabilitation phase.

But even after such a rehabilitation phase, the outcome is not always certain in muscular dystrophy patients: If the procedure is left too late, the patient will only lose additional functions. Moreover, soft tissue lengthening procedures for contractures may subsequently be required in order to enable the patient to perform everyday tasks [28].

The loss of the ability to walk and stand usually also involves a *loss of trunk control*. Since the muscles have largely lost their activity by this stage of the illness, the spine very rapidly collapses under the effect of gravity (■ Fig. 4.123). For this reasons, incipient scolioses must be



■ Fig. 4.123. Female patient with *spinal muscular atrophy* that behaves like a severe muscular dystrophy in relation to the *scoliosis*. The spine has completely collapsed in just under two years



■ Fig. 4.124. Patient with *Duchenne muscular dystrophy*. He stands with slight equinus feet, largely extended knees and hips and lumbar hyperlordosis. This is typical of a standing position that does not require much muscle power

monitored very closely (every 4 to 6 months), and even slight deformities with a scoliosis angle of 10–20° may constitute an indication for surgery [20, 28]. Although an early operation can hinder everyday functions [7], it will prolong the patient's survival time [24], improve quality of life and facilitate nursing care [16, 27].

! In severe muscle disorders, early operations are required to prolong the ability to walk and stand. Late operations serve to correct deformities, whether in the form of scolioses or disabling joint contractures.

Patients must compensate for the lack of muscle activity by functional means. They often develop equinus feet, producing the effect of a tenodesis, and these can be beneficial provided they are not excessive since they stabilize the lower leg in relation to the foot during walking and standing. Slight hyperextension at the knee and hip also allows patients to stand and walk without quadriceps activity, since the ligaments stabilize the joints passively. Since the pelvis tilts forward during hyperextension at the hip, the lumbar spine adopts a lordotic posture by way of compensation. Because of the lack of any counteracting muscle tension, the spine collapses into a position of extreme lordosis. Thus, despite the unfavorable posture (■ Fig. 4.124), the patient is still able to stand and walk largely without muscle strength.

### Important clinical conditions

#### 4.7.6.1 Duchenne muscular dystrophy

##### ➤ Definition

Duchenne muscular dystrophy is an x-linked hereditary disorder. The clinical signs and symptoms start during the first 5 years of life. The ability to walk is lost around the age of 10 and life expectancy is between 25 and 30 years. These days, heart failure rather than pulmonary insufficiency is the limiting factor.

##### **Historical background, etiology and pathogenesis**

This form of muscular dystrophy was first described by Duchenne in 1861 and occurs in 30/100,000 neonatal males. New mutations are involved in a third of cases [28]. Duchenne dystrophy involves a gene defect that leads to an underproduction of a protein that is important for muscle function known as dystrophin. This protein is found in skeletal muscle, cardiac muscle, smooth muscle and the brain, and plays an important role in the excitation of the muscle cell [18, 23].

##### **Clinical features and diagnosis**

Duchenne muscular dystrophy only affects boys. The onset of walking is delayed or fails to occur. The disease manifests itself as muscle weakness, usually during the first five years of life. Striking features include an abnor-



mal gait (waddling with hyperlordosis as compensation for the prevailing muscle weakness) and frequent falls. The muscle weakness is more pronounced in the legs than the arms. As the condition progresses, patients are no longer able to rise from the floor without help and use their arms to climb up the legs and stand up (Gowers sign). A pseudohypertrophy typically develops on the calves (■ Fig. 4.125).

The ability to walk and stand is lost around the age of 10 or 11, and the children then become confined to a wheelchair. In this phase the leg muscles are no longer adequately stretched and contractures occur. The trunk muscles also become increasingly weaker, involving the risk of rapidly progressing scolioses (■ Fig. 4.126) [4, 19].

Towards the end of the teens, the muscular dystrophy leads to progressive pulmonary insufficiency. The involvement of the cardiac muscle is difficult to manage and constitutes a life-limiting factor. In addition to the striated muscle, the *smooth muscle* also appears to be affected by the disease. Intestinal motility disorders can occur. Patients with muscular dystrophy are also associated with an increased bleeding tendency during surgeries, even though coagulation is primarily normal. One possible explanation is the involvement of vascular muscle and the

lack of reactive vasoconstriction. A dystrophin deficiency has also been detected in the brain, which explains the deterioration in patients' cognitive abilities [13].

*Female carriers* can, in rare cases, also show muscle symptoms in the form of calf cramps, calf hypertrophy and muscle weakness in the legs, and sometimes even cardiomyopathy [17, 19].

*Laboratory tests* reveal a marked elevation in the creatinine kinase level. The dystrophin deficiency can also be confirmed in the laboratory [3, 10]. The *EMG* shows signs of myopathy while the sonogram of the muscles appears hyperechoic. The *muscle biopsy* is the decisive diagnostic factor. Histological examination reveals a picture with simultaneous degeneration and regeneration. The muscle fibers show differing diameters, and the proportions of fatty and connective tissue are increased [4]. The muscle fibers ultimately die. The (tonic) postural work is increasingly taken over by connective tissue, which becomes correspondingly hypertrophic.

In terms of the *differential diagnosis*, Duchenne muscular dystrophy must be differentiated from the Becker type and from spinal muscular atrophy. *Spinal muscular atrophy* greatly resembles Duchenne muscular dystrophy, but can affect both sexes and appears at an earlier age, with the result that patients are not even able to walk. The course of *Becker muscular dystrophy* is similar to that of Duchenne muscular dystrophy but the former progresses more slowly and has a better prognosis.

### Treatment and prognosis

No *causal treatment* exists for Duchenne muscular dystrophy. Transplantation of normal myoblasts has proved ineffective [15], and gene therapy is still in the experimental stage [12]. The *orthopaedic treatment* is aimed at preserving the ability to walk and stand for as long as possible and preventing contractures. Early surgical muscle and tendon lengthening procedures have been proposed as the statistics indicate that the loss of the ability to stand and walk can thereby be deferred [22, 30, 31]. The time gained is between 1 and 3 years, which is a significant period for these patients [28]. Like several authors [28] we also consider that these procedures, which combine lengthening of the hip abductors, knee flexors and triceps surae muscle, are indicated only when contractures start to interfere with everyday functions and the need for surgery becomes apparent even to critical parents. Braces and corsets are often required not only for postural correction, but also to replace the lost muscle function [28].

The correction of abnormal spinal posture and spinal deformities is useful even for small angular deviations. Once they appear, spinal deformities (with a scoliosis angle of approx. 30°) are progressive [1, 20], and any postponement of surgery can lead to a more difficult procedure in a patient with a worse state of general health. The surgical stabilization of the spine improves the quality



■ Fig. 4.125. Typical *pseudohypertrophy* of the calves in Duchenne muscular dystrophy



■ **Fig. 4.126a, b.** Patient with *Duchenne muscular dystrophy*. **a** Posterior view: The severe scoliosis can no longer be corrected by conservative means and only allows the patient to stand for short periods. **b** Anterior view: The rib cage sits on the iliac crest. The pelvis is oblique. The pronounced muscular atrophy, the lack of facial expression and the oversized tongue are additional visible signs of the muscular dystrophy

of life and, in particular, the ability to sit and facilitates nursing care [16]. Corset treatments are not very effective [28] and useful only in exceptional cases when operations are no longer possible or are declined.

From the *pediatric standpoint*, any *pulmonary and cardiac insufficiency* must be treated. The pulmonary insufficiency manifests itself in the form of hypoxemic phases (particularly at night), which also affects mental performance during the day. A home ventilation program (assisted nocturnal ventilation) can significantly improve these symptoms. More difficult is the treatment of the heart failure, which is likewise progressive and a life-limiting factor. In the context of surgery, patients with Duchenne muscular dystrophy are not only associated with an increased bleeding tendency but also an increased risk of malignant hyperthermia and hypercalcemia [5].

#### 4.7.6.2 Becker muscular dystrophy

##### ➤ **Definition**

Becker muscular dystrophy is an x-linked hereditary disorder. It resembles the Duchenne type, but the signs and symptoms are less pronounced and progress less rapidly.

##### **Occurrence, etiology**

Becker muscular dystrophy has an incidence of 3/100,000 and is thus 10 times rarer than the Duchenne type. A genetic defect of varying severity is also present in the Becker type and the formation of dystrophin is also affected [11].

##### **Clinical features and diagnosis**

The disease manifests itself between the age of five and adolescence. In some cases, it does not appear until adult-

hood. Problems occur with walking and climbing stairs. Muscle cramps are present during and after physical exertion. The muscle weakness tends to occur proximally and, as with the Duchenne type, the patient develops calf hypertrophy, together with a waddling gait and lordosis. Since the Becker dystrophy progresses much more slowly, patients are still able to walk even after puberty [4]. Life expectancy is determined by the degree of respiratory insufficiency and is between 40 and 60 years. The orthopaedic and cardiac findings resemble those for Duchenne muscular dystrophy, but are less pronounced. The incidence of scolioses is not known, but spinal deformities must be expected in severe cases [28].

From the diagnostic standpoint, the *muscle enzymes* are raised, and the *EMG* shows signs of myopathy. As in Duchenne muscular dystrophy, the *muscle biopsy* shows a picture of degeneration and regeneration. The abnormal dystrophin level can be confirmed by laboratory tests and differentiated from that for the Duchenne type [3, 10, 11].

##### **Treatment and prognosis**

No treatment currently exists for the underlying condition. The *orthopaedic measures* are essentially no different from those for Duchenne muscular dystrophy (see above), but are only required later in life.

#### 4.7.6.3 Other forms of muscular dystrophy Facioscapulohumeral muscular dystrophy

This type of muscular dystrophy is an autosomal dominant condition that affects the muscles of the face and shoulder girdle. The onset of signs and symptoms can occur from early childhood right through to adulthood. The condition manifests itself as a weakness of the aforementioned muscles, and the pelvic muscles can also be af-

affected in some cases. Deafness and changes in the fundus oculi may also be present [4].

The *muscle enzymes* are normal or slightly elevated, and the *EMG* may also be normal. The *muscle biopsy* shows a variable picture with focal fiber atrophy and dystrophic sections with proliferation and regeneration and increased amounts of connective and fatty tissue. The *course* and *prognosis* differ, varying from a very slow progression and a normal life expectancy to a loss of walking ability in early adulthood. The aim of *treatment* is to preserve mobility in the upper extremities. Fixation of the scapula to the thorax can prove helpful [2, 14, 28]. As the dystrophy progresses, the muscle power is no longer capable of stabilizing the trunk against gravity and the patient risks developing a scoliosis. Corresponding clinical and radiological follow-up is indicated.

### Emery-Dreifuss muscular dystrophy

This X-linked recessive form of muscular dystrophy primarily affects older children, adolescents and adults. The signs and symptoms consist of walking difficulties and cardiac arrhythmias. The spine stiffens and fixed deformities occur in the extremities. The muscular dystrophy is characterized by slow progression. As *laboratory parameters* the muscle enzymes are slightly to moderately elevated, and the *EMG* shows myopathic changes. The *muscle biopsy* shows a dystrophic histological pattern, partially mixed with muscular atrophies [4, 19, 21]. The condition can become life-threatening if it affects the heart. The *orthopaedic management* aims to preserve walking ability and prevent and treat skeletal deformities. The cardiac and pulmonary situation must be monitored. If life-threatening arrhythmias are present a pacemaker will need to be inserted [26, 28].

### Limb-girdle dystrophy

Limb-girdle dystrophy is an autosomal-recessive form of muscular dystrophy. The disease can appear between childhood and adulthood. Patients experience problems with walking and climbing stairs and suffer from muscle cramps. The gait pattern is abnormal and produces a compensatory lordotic posture. The muscle weakness is of varying severity, and the prognosis is not uniform. The *laboratory tests* show elevated muscle enzymes of varying degree, and myopathic changes are visible on the *EMG*. Dystrophic changes are observed on the *muscle biopsy* [4, 19]. The picture resembles that of Duchenne or Becker muscular dystrophy but the patient has a normal dystrophin level [28]. The *orthopaedic management* aims to preserve the ability to walk and prevent musculoskeletal deformities.

### Congenital forms of myopathy

These forms include:

- congenital muscular dystrophy, De Lange type,
- congenital muscular dystrophy, Batten-Turner type,

- central-core myopathy,
- nemaline myopathy,
- myotubular myopathy,
- fiber type disproportion,
- other forms.

Whereas the *De Lange myopathy* is a malignant condition, with death often occurring even during infancy, the *Batten-Turner type* is benign, often with slow or absent progression, even though the overall clinical picture matches that of a progressive muscular dystrophy [4]. Under histological examination, the cell nuclei in *central-core myopathy* are located centrally in the muscle fibers. The children are flaccid even at birth and tend to develop skeletal deformities (flatfoot, clubfoot: 21%, hip dislocations: 19%, scolioses: 37% and contractures: 15%), which require early orthopaedic treatment [8, 28].

The manifestation of *nemaline myopathy* (recessive or dominant) is highly variable: some patients die during the neonatal period, whereas others are able to walk normally and only show slight muscle weakness. The orthopaedic problems arise from the development of scoliosis and lumbar hyperlordosis [28].

The *myotubular myopathy* is variable with differing modes of inheritance. The rare X-linked recessive variant often leads to death during the neonatal period. The more common autosomal recessive type tends to produce clubfeet, lordoscolioses and winged scapula in the second decade of life [28]. *Fiber type disproportion* is histologically characterized by a predominance of excessively small type I fibers and excessively large type II fibers. This myopathy is not associated with a consistent clinical picture, but involves histological and functional changes of the muscles as a result of myopathies, neuropathies or CNS disorders. The severity of the signs and symptoms is highly variable. Spinal deformities may require surgical correction [28].

Other rare forms of congenital myopathy exist, including *minicore myopathy*, the *mitochondrial myopathies* and other forms that will not be elaborated on here.

### Curschmann-Steinert myotonic dystrophy

The illness usually occurs during early adulthood with the main symptoms of weakness and stiffness. One striking clinical feature is spontaneous myotonia after sustained clenching of the fist. The facial muscles are also weak and ptosis is present. Cataracts and delayed intellectual development are additional findings. The prognosis depends on any accompanying cardiomyopathy and respiratory problems. The *EMG* shows a myotonia with myopathy, while the *ECG* shows conduction disorders and arrhythmias. The muscle biopsy reveals dystrophic changes with central cell nuclei [4]. The *orthopaedic measures* are aimed at preventing deformities and preserving motor skills.

#### 4.7.6.4 Spinal muscular atrophy

##### Definition

The hereditary sensorimotor neuropathies are a heterogeneous group of diseases involving degeneration of the anterior horn cells of the spinal cord, resulting in progressive muscle weakness. The distribution is usually symmetrical.

— *Synonym: Hereditary motor and sensory neuropathy (HMSN)*

##### Historical background

The various forms of spinal muscular atrophy were first described by G. Werdnig in 1891, J. Hoffmann in 1893 and E. Kugelberg and L. Welander in 1956.

##### Clinical features and diagnosis

The disease starts even within the uterus or during infancy. The principal signs and symptoms are hypotonia, muscle weakness and respiratory problems. Although these are usually very pronounced, they are not very progressive. Creatinine kinase and nerve conduction velocity are normal. The *muscle biopsy* shows typical signs of denervation.

##### Classification

Three forms are distinguished in the *classification according to Byers* [9]:

- Type I: Acute infantile (severe) form *Werdnig-Hoffmann*.
- Type II: *Chronic infantile form* (intermediate spinal muscular atrophy).
- Type III: Juvenile (mild) form *Kugelberg-Welander*.

##### Specific forms

###### Type I: Acute infantile form, Werdnig-Hoffmann disease

(severe form)

This is an autosomal recessive hereditary disorder. It manifests itself even within the uterus as deficient fetal movements or, in the first few months of life, as hypotonia and muscle weakness. The infant has difficulty in sucking and swallowing. Clinically, the infant shows marked muscle hypotonia and muscle weakness of the whole trunk and extremities. Breathing is hampered, leading to frequent respiratory infections. The children die during the first three years of life. Creatinine kinase and nerve conduction velocity are normal. The *EMG* shows signs of denervation, while atrophied fibers are noted on the *muscle biopsy* [4, 19]. The *orthopaedic measures* must replace the missing muscles (supportive corset, braces).

###### Type II: Chronic infantile intermediate form

The intermediate form is an autosomal-recessive condition. The onset of the disease occurs between the ages of 6 and 12 months. The children exhibit weakness of the leg

muscles that prevents them from walking and standing without assistance, although they may be able to sit up on their own. Fasciculations are apparent on the tongue, as is a tremor of the hands. The children are of normal intelligence. The muscle weakness does not usually progress, and changes in the patient's abilities are primarily growth-related and caused by weight gain. The pulmonary situation determines the long-term prognosis.

The *creatinine kinase* is normal to moderately elevated. Signs of denervation and reinnervation are observed on the *EMG*. The *sonogram* shows a characteristic picture with a hyperechoic muscle pattern and muscular atrophy [4, 19]. From the *orthopaedic standpoint* the deficient muscles must be replaced with a corset and braces in order to enable the child to develop normally without deformities. A scoliosis develops early on and will need to be managed accordingly with corsets and, at a later date, by surgical measures.

###### Type III: Juvenile form, Kugelberg-Welander disease (mild form)

The inheritance mode of the disease is usually autosomal-recessive, although rarer dominant or X-linked forms also exist. The signs and symptoms appear between the age of two and adulthood. Muscle weakness is the key sign. The disease is not usually progressive, and slight muscular atrophy is present. It initially manifests itself in the form of problems with running and climbing stairs and, at a later stage, in the form of restrictions in walking. The gait pattern is abnormal (waddling), and the children have to use their arms to climb up the legs and stand up (Gowers sign). The proximal muscle weakness affects the legs more than the arms. Tremor and fasciculations may be present. The disease can remain stationary or show slight progression. Life expectancy is not affected.

The creatinine kinase is normal to slightly elevated. *Sonography* shows a hyperechoic muscle pattern and muscular atrophy. Signs of denervation and reinnervation are observed on the *EMG*, while the *muscle biopsy* shows fiber group atrophy and normal, enlarged fibers [4, 19]. The *orthopaedic treatment* must replace the lost muscle activity with orthoses and thereby ensure optimal patient rehabilitation.

#### 4.7.6.5 Hereditary motor and sensory neuropathy (HMSN)

— *Synonyms: Charcot-Marie-Tooth disease, peroneal muscular atrophy*

This group of diseases comprises genetic disorders involving a muscle weakness on the distal arms and legs. *Three forms* are distinguished:

- a dominant demyelinating form (*HMSN 1*)
- a neuronal-dominant form (*HMSN 2*),
- a demyelinating recessive form (*HMSN 3*).



The determination of the nerve conduction velocity is crucial for the demyelinating forms. The patients must also be investigated in order to be able to establish the mode of inheritance. The patients usually learn how to stand and walk reasonable well. The main problem, apart from the muscle weakness, is posed by foot deformities (pes cavus). Major skeletal procedures on the feet can be avoided by early soft tissue operations [25]. However, we have occasionally observed pain syndromes after minor procedures, which we considered to be neurogenic in origin. The pain occurs well away from the operation area and can be very irksome. Treatment is the same as that for other types of neurogenic pain. In severe cases the hands are affected as well, causing weakness and deformities.

#### 4.7.6.6 Myotonia congenita (Thomsen disease)

Myotonia congenita (Thomsen disease) can be inherited as a dominant or recessive condition and occurs shortly after birth or in early childhood. Typical signs and symptoms are stiffness after rest and difficulties with opening the fist, releasing an object, etc. The muscle power and function are almost unchanged [4, 19]. Orthopaedic treatments are not required.

#### 4.7.6.7 Myasthenia gravis

A distinction is made between neonatal, infantile and juvenile forms. A feature common to all these forms is increased muscle fatigability, which mainly affects the eye and masticatory muscles. Other muscle groups over the whole body are affected at a later stage. The diagnosis is confirmed by the edrophonium (Tensilon) test [4, 19]. Myasthenia gravis is primarily a neurological disorder, and orthopaedic problems are rarely involved.

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