Jan Douwes Visser

# Pediatric Orthopedics

## Symptoms, Differential Diagnosis, Supplementary Assessment and Treatment

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Word processing: Hendrika Schotanus and Yvonne Werink. Translation: Prof. Peter H. Robinson, MD., Ph.D. Drawings: Douwe Buiter

ISBN 978-3-319-40176-8 ISBN 978-3-319-40178-2 (eBook)
DOI 10.1007/978-3-319-40178-2

Library of Congress Control Number: 2016960534



Printed on acid-free paper

This Springer imprint is published by Springer Nature
The registered company is Springer International Publishing AG
The registered company address is: Gewerbestrasse 11, 6330 Cham, Switzerland

# Preface

In practice, we do not tend to think in terms of differential diagnosis. We often consider the most probable diagnosis based on the patient's complaints. Only when the most probable diagnosis is not supported by physical examination and/or supplementary tests, do we further consider the other possibilities. In this book, *Pediatric Orthopedics: Symptoms, Differential Diagnosis, Supplementary Assessment and Treatment*, we are working towards a system for differential diagnosis based on the principal complaints and findings. In some cases there are more than one different kinds of complaint. As an example, in Osgood-Schlatter, there are complaints of pain and swelling. In those cases we consider the most preveland complaint which in Osgood-Schlatter is pain. In other cases, for instance a meniscal tear, there may be several individual complaints or a combination of these such as pain medially or laterally combined with limited extension and/or swelling. A torn meniscus appears in several differential diagnoses. A system has been established for every diagnosis as described here: (a) explanatory notes, (b) advice on supplementary tests, (c) advice as to which problems can be treated in primary care, (d) when to refer for secondary care treatment, and (e) what the options are for secondary care treatment. Generalized neurological disorders and neuromuscular disorders such as in spasticity, spina bifida, and acute traumatic lesions have been left out. There are many diverse treatment possibilities available. However, many pediatric orthopedic treatments are not evidence based or sometimes have a low or extremely low value from the literature. The author, advisers, and the publisher are not responsible for faults, omissions, or other implications as a result of the information given in this book. Application of the information given in this publication remains the responsibility of the clinician involved.

Groningen, The Netherlands — Jan Douwes Visser

# Acknowledgments

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# Abbreviations

| | |
|---|---|
| ANF | Antinuclear factor |
| C | Cervical |
| CRP | C-reactive protein |
| CT | Computed tomography |
| ESR | Erythrocyte sedimentation rate |
| Hb | Hemoglobin |
| HLA | Human leukocyte antigen |
| Ht | Hematocrit |
| L | Lumbar |
| MRA | Magnetic resonance arthrography |
| MRI | Magnetic resonance imaging |
| n. | Nerve |
| NSAID | Non-steroidal anti-inflammatory drugs |
| S | Sacral |
| T | Thoracal |
| WBC count | White blood cell count |

# Contents

# Chest Wall

# 1

## Chest Wall Deformity

Complaint: there is a deformity of the chest wall.

Assessment: deformity of the chest wall.

Differential diagnosis:
**pectus carinatum (pigeon breast)**
**pectus excavatum (funnel chest)**
**fusion disorder of the sternum**
**absent ribs**
**Poland syndrome**

Explanatory note: **pectus carinatum.** The sternum and the adjoining cartilage and bone are anteriorly prominent (Fig. 1.1). The abnormality is most noticeable in the first year of life, but can also develop during puberty (Fig. 1.2). As a rule, pigeon breast is only a cosmetic issue. Respiratory difficulties may occur when the sternum shifts strongly forward, causing the thorax to be in a continuous inspiratory position.

**Pectus excavatum** There is an indentation on the front side of the chest. The deepest area lies at the level of the distal part of sternum and the xiphoid process (Fig. 1.3). The abnormality is usually present at birth, but can also develop later. A funnel chest seldom causes physical complaints, the issue tends to be only cosmetic. The abnormality is common in Marfan syndrome.

**Fusion disorder of the sternum** Partial fusion disorders of the cranial area are the most common. In the distal area they are very rare. It is also possible that the sternum is split along the entire length (total sternal fissure). This abnormality is accompanied by cardiac anomalies and/or defects in the diaphragm.

**Absent ribs** In this abnormality several ribs are usually absent on one side of the chest wall. There may also be sternal and vertebral abnormalities such as hemivertebrae and block vertebrae. As a result there is a flail thorax, which can lead to a shortness of breath.

**Poland syndrome** In Poland syndrome[1] there is a unilateral absence of the pectoralis minor muscle and the sternal part of the pectoralis major muscle (Fig. 1.4). Males are affected in 70% of cases. It is generally combined with abnormalities of the hand on the same side, such as small hands (hypoplasia) with absent fingers, webbed fingers (syndactyly) and/or shortened fingers (brachydactyly). The combination of hypoplasia of the hand, syndactyly and brachydactyly is sometimes called symbrachydactyly. There may also be absence of the forearm flexor muscles and the entire arm may be under-developed. Associated anomalies may be the Klippel-Feil syndrome[1] (short neck), Möbius syndrome[1] (paralysis of the facial muscles), a Sprengel deformity (elevated shoulder blade) and pectus excavatum.

[1] See Appendix.


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_1

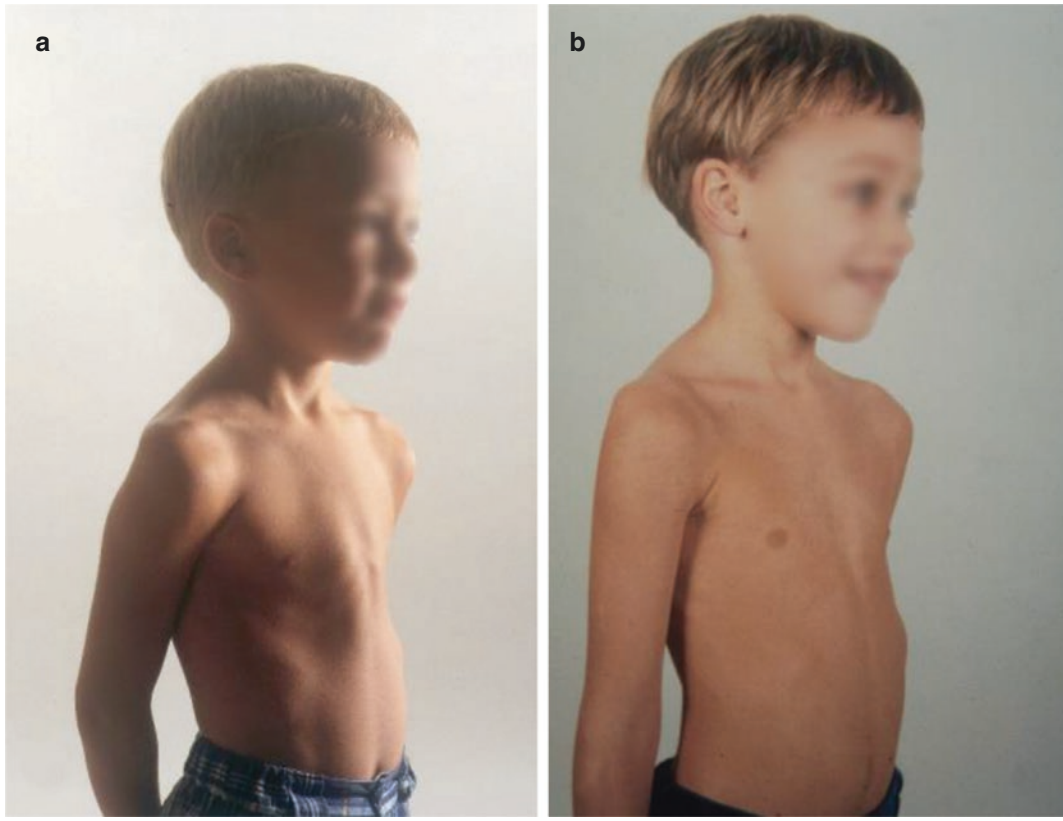


**Fig. 1.1** (**a**) Boy, about 5 years old, with a pectus carinatum (pigeon breast). (**b**) After treatment with a trunk orthosis with a pressure pad

Supplementary assessment: in a fusion disorder of the sternum, cardiac abnormalities or defects in the diaphragm should be ruled out.

Primary care treatment: none.

When to refer: the child should be referred as quickly as possible when a shortness of breath accompanies absent ribs. If treatment of the pigeon breast is desired for cosmetic reasons, it is mostly carried out starting at the first year of age. Funnel chest treatment is done starting at the age of 6 years. Infants with a fusion disorder of the sternum should be referred as quickly as possible. Referral is not indicated for the chest deformity in Poland syndrome.

Secondary care treatment: **pectus carinatum.** Spontaneous resolution of the pigeon breast may occur before the first year of age. If the pigeon breast is still present after the first year of age, it can be slowly pushed back with the help of a trunk orthosis with a pressure pad at the level of the prominent thoracic area (Fig. 1.5). A period of 1 year is usually sufficient. The intention is to wear the trunk orthosis day and night, and after correction the trunk orthosis should still be worn for some time at night. This may also be an effective treatment for some cases when the anomaly develops during puberty. The assessing physician must be able to manually press the most prominent portion a little posteriorly. If he cannot do that, there is no point in treating with a trunk orthosis. Treatment with a trunk orthosis in adolescents usually has to be continued until full growth has been achieved. A stiff pectus carinatum may be treated operatively (Fig. 1.6).

**Pectus excavatum** Starting at the age of 6 or 7 years, a pediatric or thoracic surgeon can carry out a Nuss procedure under thoracoscopic guidance.

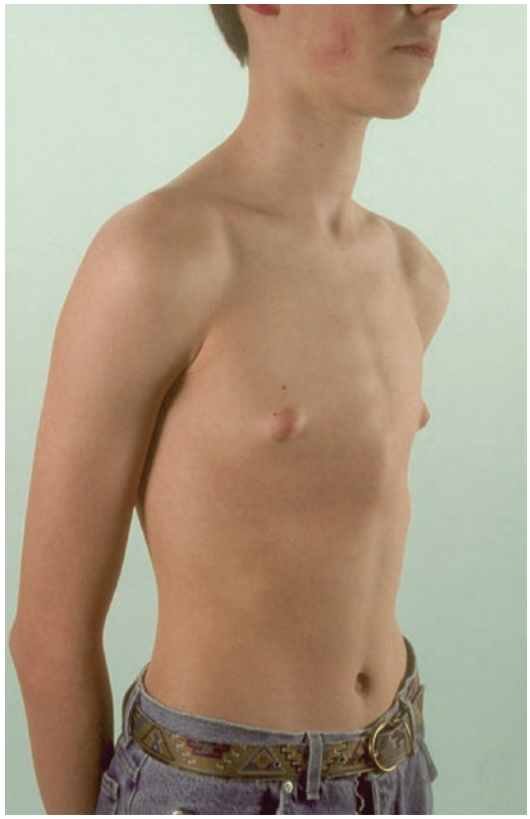

**Fig. 1.2** A pectus carinatum (pigeon breast) can also develop during puberty

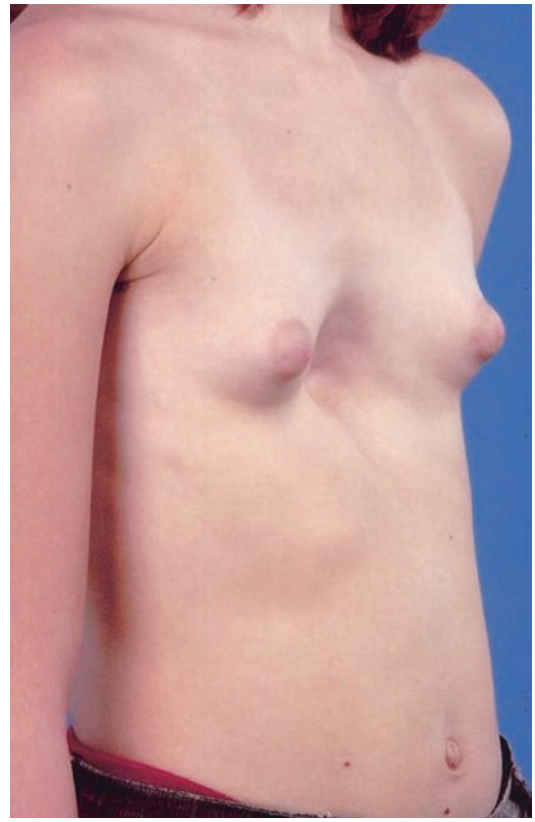

**Fig. 1.3** Pectus excavatum (funnel chest)

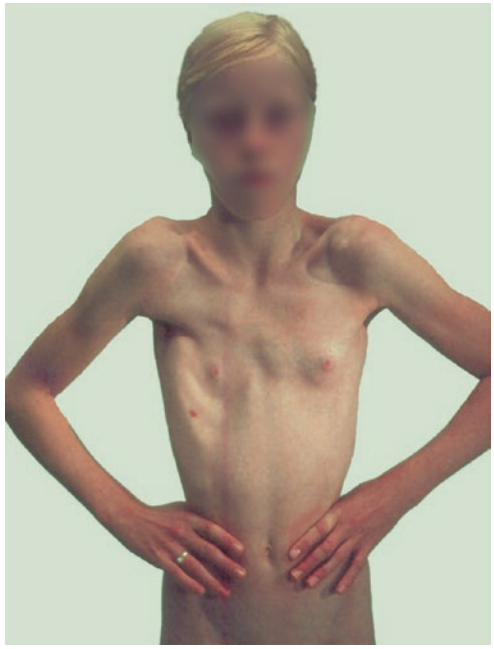

**Fig. 1.4** Poland syndrome. On the right side there is a unilateral absence of the pectoralis minor muscle and the sternal portion of the pectoralis major muscle

An incision is made on each side of the chest wall and a forward-curved bar is inserted which acutely pushes the indentation forward (Figs. 1.7 and 1.8). The bar is removed after a few years.

**Fusion disorder of the sternum** Operative closure of the defect should be carried out as soon as possible after birth.

**Absent ribs** Early operative correction in the neonatal period gives the best results.

**Poland syndrome** Treatment of the chest wall abnormality is not indicated.

## Chest Wall Pain

Complaint: the child complains of pain in the front side of the chest wall.

Assessment: there is pain on pulpation at the level of the sternum-rib junction.

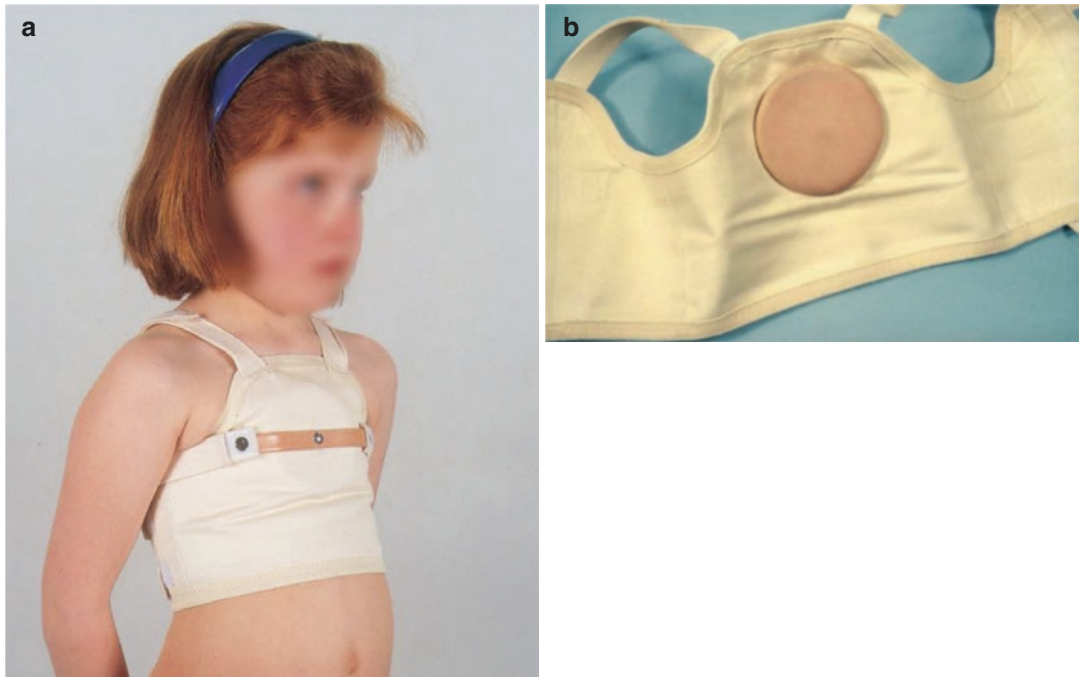


**Fig. 1.5** (**a**) Starting at the first year of age, a pectus carinatum (pigeon breast) can be treated using a trunk orthosis with a pressure pad for the prominent thoracic area. (**b**) The pressure pad on the inner side of the trunk orthosis

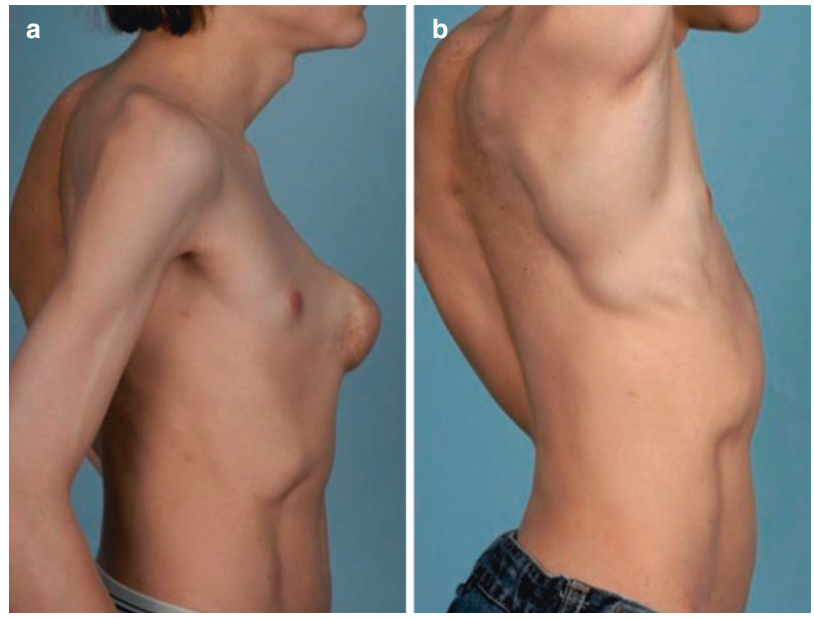


**Fig. 1.6** An extensive pectus carinatum (pigeon breast), (**a**) before and (**b**) after operative correction. There is also a considerable scoliosis (Images received from Prof. D.C. Aronson, M.D., Ph.D., The Netherlands)

Differential diagnosis:
**Tietze syndrome**
**costochondritis (costosternal syndrome, costosternal chondrodynia)**
**bone tumor**

Explanatory note: **Tietze syndrome.** In this syndrome, there is a painful unilateral swelling of the rib cartilage at the level of the costosternal junctions. It usually involves the second or third rib. This is a suspected

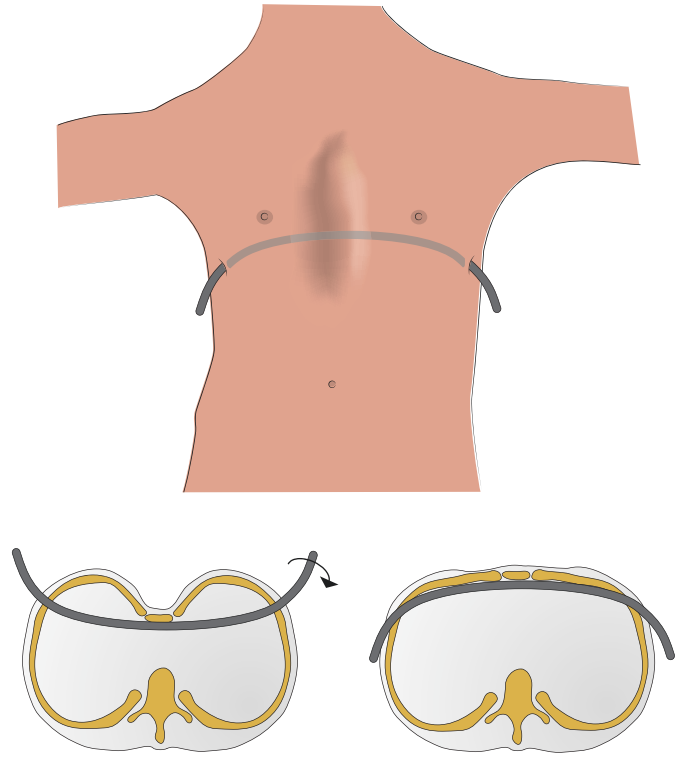

**Fig. 1.7** Pectus excavatum (funnel chest): the Nuss operation can be performed starting at the age of 6 or 7. Under thoracoscopic guidance, an incision on each side of the chest wall is made and a curved metal bar is inserted which pushes the indentation forwards. The bar is removed after a few years

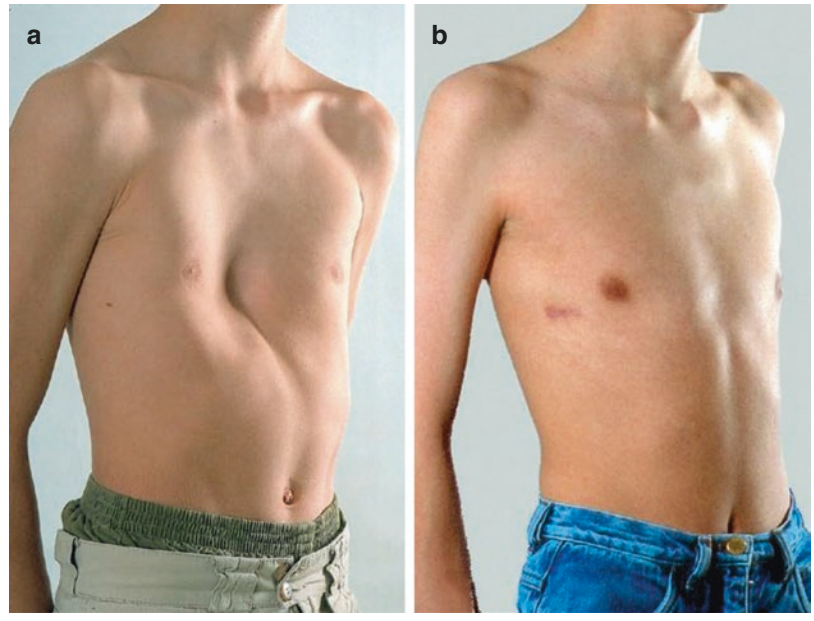


**Fig. 1.8** Pectus excavatum (funnel chest), (**a**) preoperatively. (**b**) after the Nuss procedure (Images received from Prof. D.C. Aronson, M.D., Ph.D., The Netherlands)

perichondritis with an unknown cause. Symptoms tend to disappear spontaneously after several weeks to months.

**Costochondritis** In contrasts to Tietze syndrome several costosternal junctions are involved usually the third, the fourth and the fifth. There is no swelling. The cause is unknown. Pain from costochondritis resolves within a year.

**Bone tumor** There is painful swelling at the level of the ribs. It is not possible to manualy press the bony swelling posteriorly (see Table 1.1).

Supplementary assessment: If there is any doubt of a tumor X-rays, MRI or CT scan may be requested.

Primary care treatment: In case of Tietze syndrome or costochondritis pain medication may be given if necessary.

When to refer: if a tumor is suspected.

Secondary care treatment: **bone tumor.** A bone tumor should be treated in a medical center specialized in bone tumors. Treatment depends on the nature of the tumor.

**Table 1.1** Tumours at the level of the chest cavity. The tumours identified with the § sign are rare

| Benign bone tumors | Malignant bone tumors |
|---|---|
| Osteochondroma | Ewing sarcoma |
| Enchondroma | |
| Fibrous dysplasia | |
| Eosinophilic granuloma | |
| Aneurysmal bone cyst § | |
| Osteoid osteoma § | |

Based on Adler and Kozlowski (1993)

**Differential Diagnosis Chest Wall Deformity**

| | |
|---|---|
| Prominence of the anterior area of the chest wall. | **Pectus carinatum (pigeon breast)** |
| Indentation of the anterior area of the chest wall. | **Pectus excavatum (funnel chest)** |
| Split at the level of the sternum. | **Fusion disorder of the sternum** |
| Flail thorax. | **Absent ribs** |
| One-sided absence of the pectoralis minor and the sternal part of the pectoralis major muscles. | **Poland syndrome** |

**Diagnosis: Chest Wall Pain**

| **Chest wall pain** | |
|---|---|
| Painful swelling of the costosternal junction usually the second or third rib. | **Tietze syndrome** |
| Pain without swelling of the costosternal junctions of the third, fourth and fifth rib. | **Costochondritis** |
| Painful swelling at the level of the ribs. | **Bone tumor** |

# Neck

# 2

## Neck Deformity

### Neck Flexion Deformity

Complaint: flexion deformity of the neck.

Assessment: instead of a normal cervical lordosis there is a kyphosis.

Diagnosis: **cervical kyphosis**

Explanatory note: **cervical kyphosis.** A cervical kyphosis in a neutral posture should be considered abnormal. Cervical kyphosis can be caused by one or two under- developed vertebral bodies or an operation in which a laminectomy was performed, or it can be part of a syndrome (Table 2.1). If it is one of the symptoms of a syndrome, the child's other abnormalities are usually so impressive that the practitioner has not noticed the kyphosis in the neck. It is however important to identify the cervical kyphosis because even during infancy compression of the spinal cord can already occur, resulting in lifelong paraplegia or even death.

Supplementary assessment: anteroposterior and lateral X-rays as well as a CT-scan and a MRI of the cervical spine.

Primary care treatment: none.

When to refer: all cervical kyphoses should be considered pathological and be referred as quickly as possible.

Secondary care treatment: **cervical kyphosis.** In milder kyphosis without neurological abnormalities a wait-and-see approach may be taken. One should wait until the age of 18 months before performing a spondylodesis for more severe kyphosis without neurological abnormalities.

Neurological abnormalities necessitate earlier intervention. In addition to the cervical spondylodesis, the spinal cord must be decompressed.

### Short Neck

Complaint: short neck.

Assessment: the patient has a short neck (Fig. 2.1). In 50 % of cases there is a triad:

- short neck
- limited mobility of the cervical spine
- growth of cranial hair on the back

In less than 20 % of cases extra skin can be seen on the lateral side(s) of the neck with muscular fascial tissue in the form of a wing (pterygium colli), known as "webbed neck". This extra tissue

**Table 2.1** Syndromes with a cervical kyphosis. See Appendix for features of syndromes

| |
|---|
| Campomelic (camptomelic) dysplasia |
| Conradi-Hünermann syndrome |
| Larsen syndrome |
| Neurofibromatosis (Von Recklinghausen disease) |


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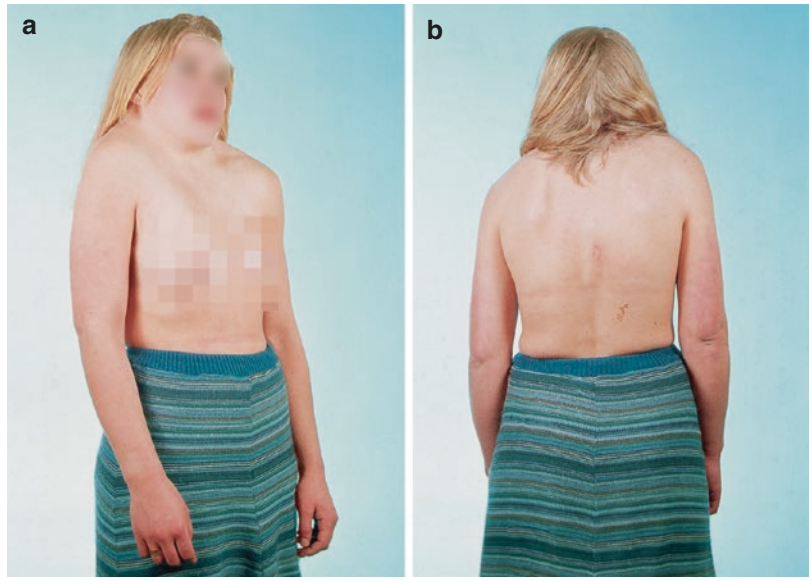



**Fig. 2.1** (**a**, **b**) Klippel-Feil syndrome. The short neck and the low cranial hair growth

is spread between the mastoid process and the acromion on both sides.

Diagnosis: **Klippel-Feil syndrome**[1] **(congenital brevicollis)**

Explanatory note: **Klippel-Feil syndrome.** The clinical picture was described by the French physicians Klippel and Feil in 1911 as "l'homme sans cou" (man without a neck). The short neck is caused by vertebrae that have fused together. There may also be a unilateral unsegmented bony connection, hemivertebrae, absence of the posterior elements (arches and spinal processes), a basilar impression, an occipitoatlantal synostosis, an atlanto-axial subluxation and a hypoplasia of the dens. In 20 % of cases the abnormality is accompanied by a torticollis, in 60 % there is a kyphoscoliosis at the thoracic level caused by the hemivertebrae and unilateral and unsegmented bony connections. In 30 % of cases there is an elevated posture and underdevelopment of the shoulder blade, known as a Sprengel deformity. In about 25 % of cases there are cardiac and renal abnormalities, and in 30 % deafness. Neurological complications in adults are possible as a result of compression of the myelum or of the exiting spinal nerves.

Supplementary assessment: X-rays as well as a CT-scan and a MRI of the cervical and thoracic spines.

Primary care treatment: if the patient has no pain or neurological symptoms there will be a wait-and-see policy. If there is pain, it is advisable to limit the activities and a cervical collar can be prescribed.

When to refer: in the early phase to the pediatrician before detecting any other anomalies. Referral should be made to the orthopedic surgeon for additional anomalies such as muscular torticollis and scoliosis, and to the neurosurgeon if neurological complications occur at an older age.

Secondary care treatment: **Klippel-Feil syndrome.** A torticollis based on a shortened sternocleidomastoid muscle can be treated by dividing or lengthening the muscle. Neurological symptoms are caused mostly by an atlantoaxial subluxation, a basilar impression or a congenital occipitoatlantal synostosis (for treatment, see pp. 15, 18, 19).

[1] See Appendix.

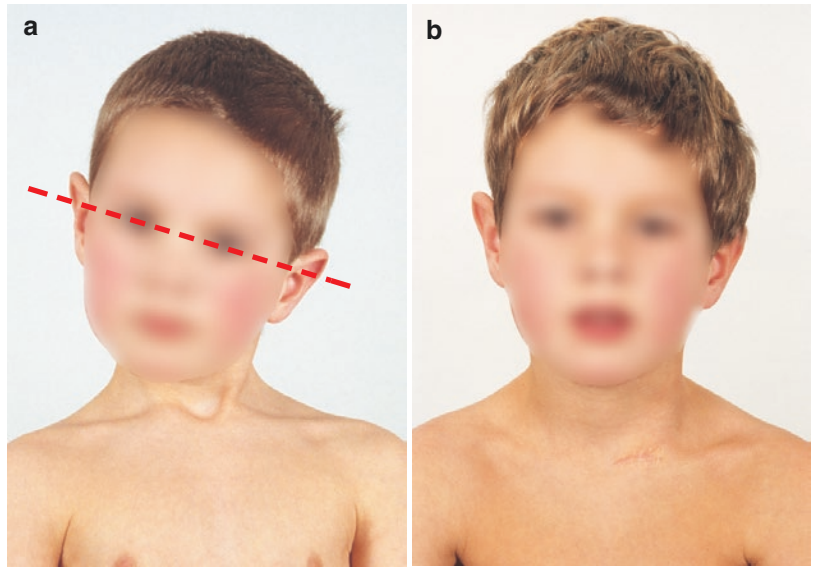



**Fig. 2.2** (**a**) Torticollis. Contracture of the left sternocleidomastoideus muscle. An easy way to determine on which side the abnormality lies is to draw an imaginary line between the pupils. The shoulder this line points to is the affected side. (**b**) Situation 2 years after dividing the left sternocleidomastoid muscle origin

## Wry Neck

Complaint: a wry neck is usually present at birth, but sometimes it appears at an older age.

Assessment: lateral flexion of the head towards the affected side, the chin is turned towards the non-deviated side. An easy way to determine on which side the deviation lies is to draw an imaginary line between the pupils. The shoulder that this line points to is the affected side (Fig. 2.2).

Differential diagnosis:

**torticollis**
- infantile muscular torticollis
- (congenital muscular torticollis)
- juvenile muscular torticollis
- osseous torticollis
- ocular torticollis

Explanatory note: **torticollis.** The cause of the infantile as well as juvenile muscular torticollis is unknown.

*Infantile muscular torticollis* In 20% of cases a swelling in the trajectory of the sternocleidomastoid muscle is visible and palpable between 10 days and 4 weeks after birth (Fig. 2.3). The swelling reduces gradually 4 weeks after it has appeared. By the age of 4–6 months the swelling has disappeared. In 80% of cases the swelling is not observed or not recognized, and the

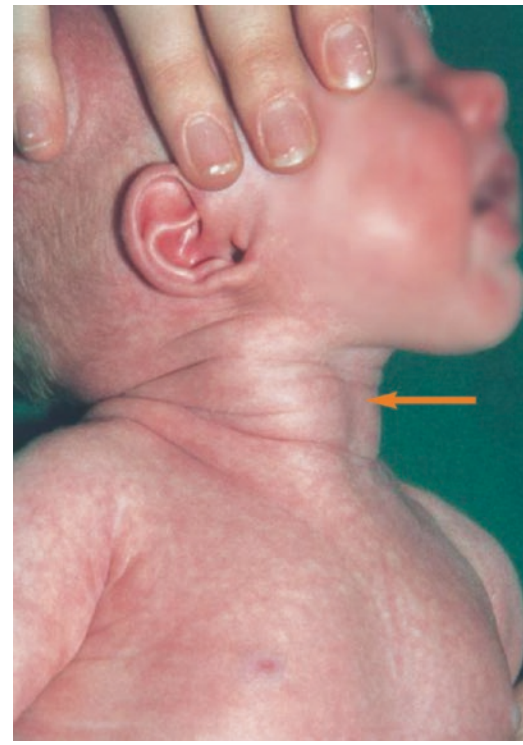


**Fig. 2.3** Swelling in the trajectory of the right sternocleidomastoid muscle (*arrow*)

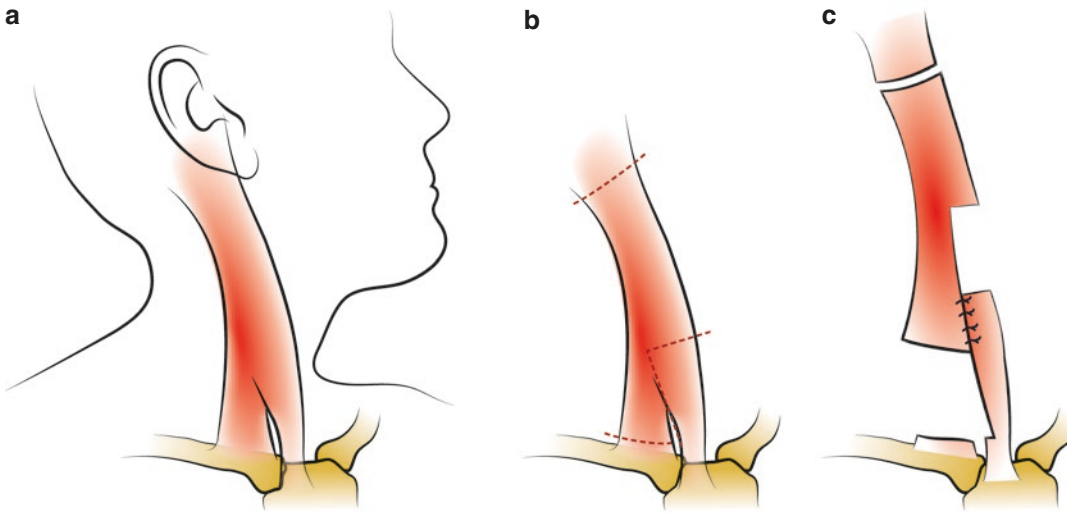



**Fig. 2.4** (**a**) The sternocleidomastoid muscle has a sternal and a clavicular origin. The shared muscle belly inserts into the mastoid process and occiput just behind the ear. The sternocleidomastoid muscle can operatively be lengthened or divided at the level of the clavicle and sternum. If there is a recurrence the insertion at the level of the mastoid process and occiput may also have to be divided. (**b**, **c**): In this case the sternocleidomastoid muscle is divided at the mastoid process and occiput. The origins of the clavicular and sternal head are lengthened with a Z-plasty (Redrawn from: Ferkel RD, Westin GW, Dawson EG, Oppenheim WL. Muscular torticollis. A modified surgical approach. J Bone Joint Surg Am. 1983;65-A:894–900)

abnormality is later on identified on the basis of a contracture of one or both heads of the sternocleidomastoid muscle.

The sternocleidomastoid muscle has a sternal and a clavicular origin. The joint muscle belly inserts into the mastoid process and occiput just behind the ear (Fig. 2.4). The cause of the swelling in the sternocleidomastoid muscle is unclear. It used to be assumed that during a difficult birth (breech presentation or forceps delivery) there was bleeding into this muscle, followed by fibrosis of the hematoma. However, blood in the swelling has never been shown. It is now assumed that the fibrous tissue is already present before birth.

Spontaneous recovery of the torticollis occurs during the first year of life in 90 % of cases. If the abnormality persists, the face and the skull will also become deformed, with a flattening of the skull and the face on the side of the contracture (plagiocephaly). A developmental dysplasia of the hip is found in 20 % of children with an infantile muscular torticollis. If there is no contracture of the sternocleidomastoid muscle, the underlying causes are a congenital anomaly of the skeleton or an ocular abnormality.

*Juvenile muscular torticollis* Sometimes a muscular torticollis appears at childhood. In that case there is a contracture of both heads of the sternocleidomastoid muscle. This condition does not recover spontaneously.

*Osseous torticollis* Due to the presence of a hemivertebra (half vertebra) or one or several asymmetric nonsegmented bony connections, a wry neck can be present. The patient has Klippel-Feil syndrome[2] if the neck is also too short.

Goldenhar syndrome or oculoauriculovertebral dysplasia is a special form of osseous torticollis in which in addition to abnormal vertebrae there are also eye and ear abnormalities such as epibulbar dermoid cysts and preauricular skin anomalies.

*Ocular torticollis* This anomaly is present from birth, but is often only noticed around the age of 9 months, after the child has achieved sitting balance. Paresis of the extraocular muscles, generally the superior oblique muscle, causes crossed and double vision when the head is held horizon-

[2]See Appendix.

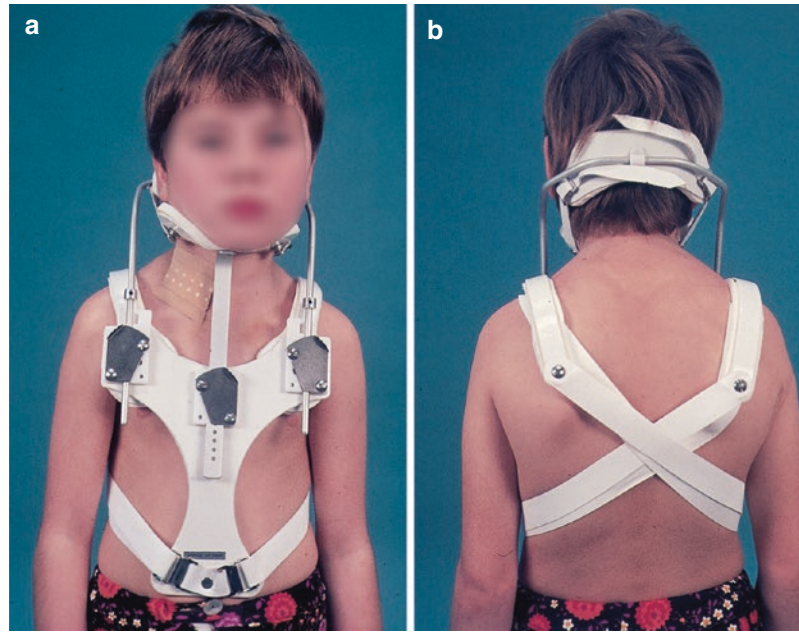



**Fig. 2.5** (**a**, **b**) Neck orthosis, in this case a SOMI orthosis (*S* sternum, *O* occiput, *M* mandibula, *I* immobilization)

tally. The child must hold his head slanted in the frontal plane to prevent double vision.

Supplementary assessment: radiological assessment of the cervical spine to reveal or rule out an osseous torticollis. In infantile or congenital muscular torticollis an ultrasound of the hips or an anteroposterior X-ray of the pelvis should be made to check for a developmental dysplasia of the hip.

Primary care treatment: a patient with infantile muscular torticollis is usually referred to a pediatric physiotherapist up to the age of 18 months. The parents do stretching exercises under the supervision of the pediatric physiotherapist. It is not certain whether these stretching exercises influence the natural history. Infantile muscular torticollis disappears spontaneously around the first year of life in 90 % of cases.

When to refer: children with a muscular or osseous torticollis should be referred to an orthopedic surgeon when the child is older than 18 months. If the sternocleido-mastoid muscle is not shortened and there are no osseous abnormalities the child should be referred to an ophthalmologist.

Secondary care treatment: **infantile muscular torticollis.** If the abnormality is still present at 1 year of age there is little chance of spontaneous recovery. In such cases, the sternocleidomastoid muscle should be operatively lengthened or divided at the clavicular and/or sternal origin (Fig. 2.4). The ideal age for operative treatment is in the third year of life. The asymmetry of the face and the skull restores completely after that. Operating before this age increases the chances of an ugly scar and retraction of the skin at the level where the sternocleidomastoid muscle has been lengthened or divided. Even after this age it is not too late to carry out the correction. Good cosmetic results can be achieved up to the age of 12. The sooner the treatment is implemented, the better the ultimate result will be with regard to facial symmetry.

Postoperative treatment involves the use of a neck orthosis day and night for 6 weeks (Fig. 2.5). In the case of a recurrence, in addition to dividing or lengthening the origin at the level of the clavicle and sternum the insertion at the level of the mastoid process should also be divided.

**Juvenile muscular torticollis** Usually this type is permanent and requires operative treatment.

**Osseous torticollis** Spondylodesis over a trajectory as short as possible should be carried out.

**Ocular torticollis** Treatment by an ophthalmologist.

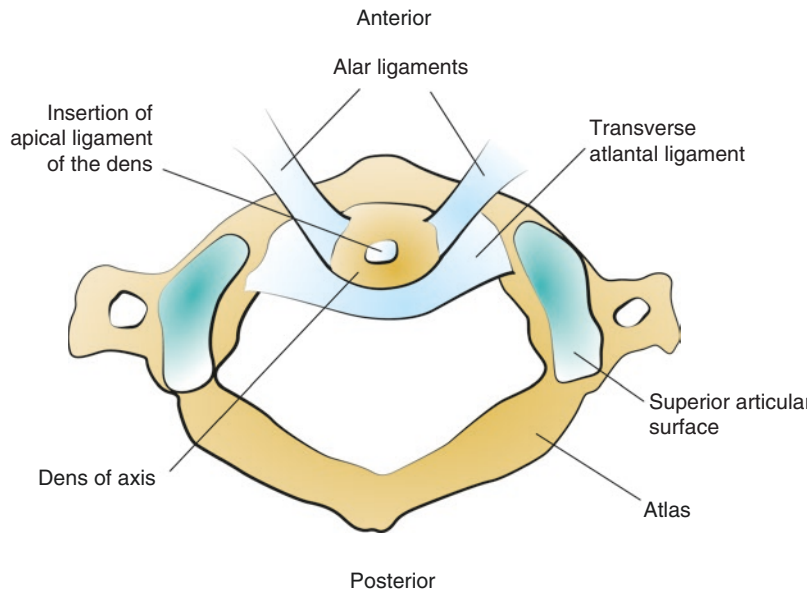



**Fig. 2.6** Ligaments at the level of C1 and C2

## Neck Pain

### Acute Onset of a Painful Stiff and Possibly Wry Neck

Complaint: the child has a painful stiff neck with a possible abnormal posture.

Assessment: there is no mobility in the cervical spine and there may be a wry neck.

Differential diagnosis:

**occipitoatlantal subluxation**
**atlantoaxial subluxation**
Grisel syndrome
Sandifer syndrome
**spondylodiscitis (discitis)**
**disc calcification**
**juvenile idiopathic arthritis**

Explanatory note: **occipitoatlantal subluxation.** An occipitoatlantal subluxation is very rare, and is generally caused by abnormal weakness in the ligaments between the dens and the back of the skull and these are the apical ligament of the dens and the alar ligaments (Fig. 2.6). This causes an occipitoatlantal subluxation and occurs in 60 % of cases of Down syndrome[3]. These children are mostly symptom-free.

**Atlantoaxial subluxation** An anterior atlantoaxial rotatory displacement can arise as a result of a congenital weakness or tear of the transverse atlantal ligament, which is the case in 10–20 % of children with Down syndrome. A posterior atlanto-axial rotatory displacement can occur as a result of an under-developed (hypoplasia) or absent (aplasia) dens (Fig. 2.7). Atlantoaxial subluxations also appear in dwarfism, such as spondyloepiphysial dysplasia[3] (40 %), often accompanied by hypoplasia of the dens, an os terminale or an os odontoideum (Fig. 2.8), and in mucopolysaccharidoses with Morquio syndrome[3] as its most common type and is often accompanied by a hypoplasia of the dens.

*Grisel syndrome* Grisel syndrome is characterized by the acute occurrence of a stiff and wry neck resulting from a throat infection, such as tonsillitis or a retrotonsillar abscess. The infection weakens the intervertebral ligaments and an atlantoaxial subluxation is the result.

*Sandifer syndrome* An acute painful stiff and possibly wry neck may occur as a result of

[3]See Appendix.

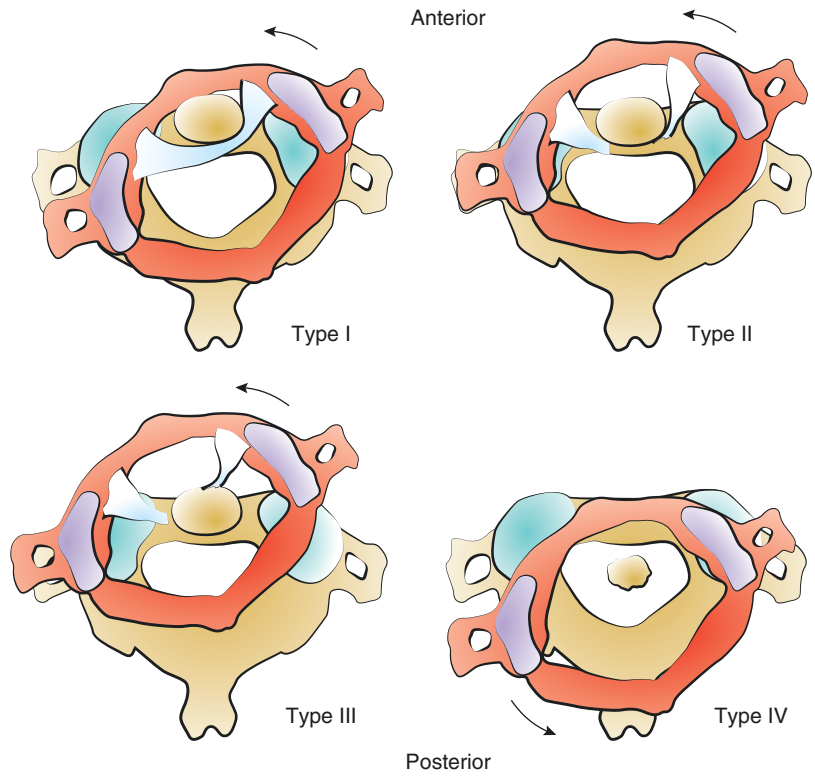



**Fig. 2.7** Atlantoaxial subluxation. *Type I*: Rotatory displacement, the transverse atlantal ligament is intact. No anterior shift. *Type II*: Rotatory displacement. One of the two atlantoaxial joints is subluxated. The transverse atlantal ligament is insufficient. Anterior shift of 3–5 mm. *Type III*: Rotatory displacement. Both atlantoaxial joints are subluxated. Insufficient transverse atlantal ligament. Anterior shift of more than 5 mm. *Type IV*: Posterior subluxation. There is hypoplasia or aplasia of the dens (Redrawn from: Fielding JW, Hawkins RJ. Atlantoaxial rotatory fixation. (Fixed rotatory subluxation of the atlanto-axial joint). J Bone Joint Surg Am. 1977;59-A:37–44)

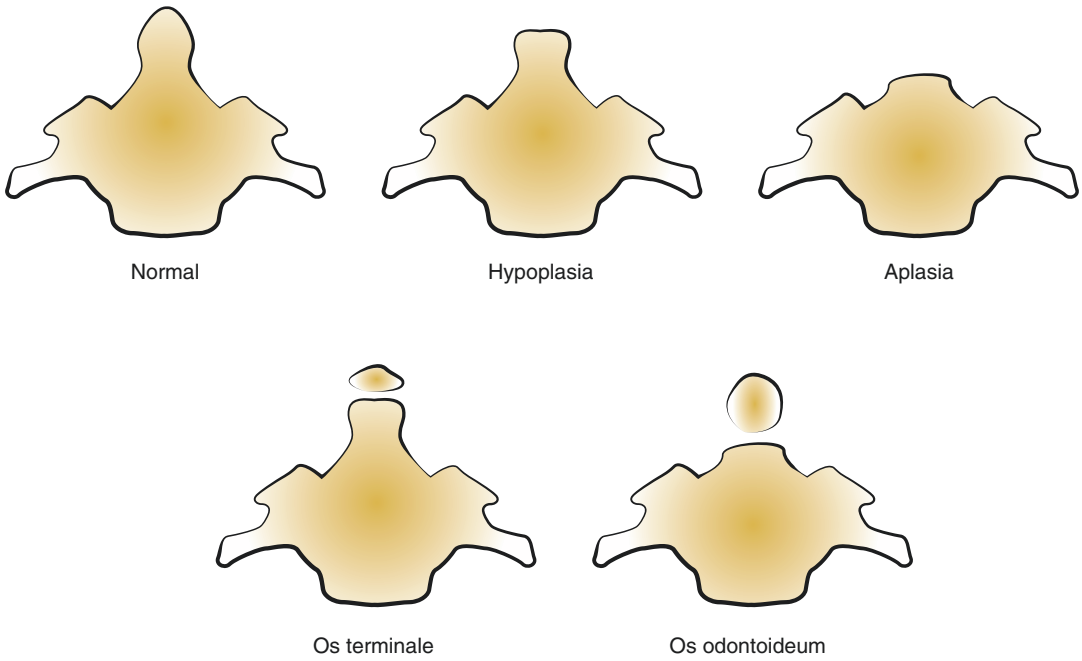



**Fig. 2.8** Dens variations

heartburn caused by gastroesophageal reflux in young children, during or right after a meal. There is no atlantoaxial subluxation.

**Spondylodiscitis** Older children usually have fever in addition to a painful stiff neck. Discitis is generally caused by staphylococcus aureus.

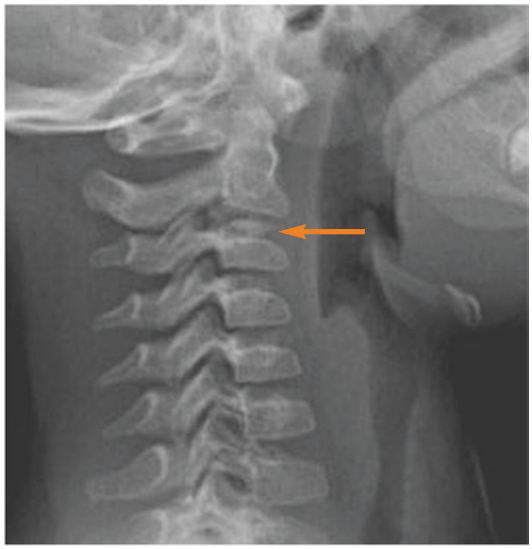


**Fig. 2.9** Lateral X-ray of the cervical spine. Disc calcification at the C2-C3 level (*arrow*)

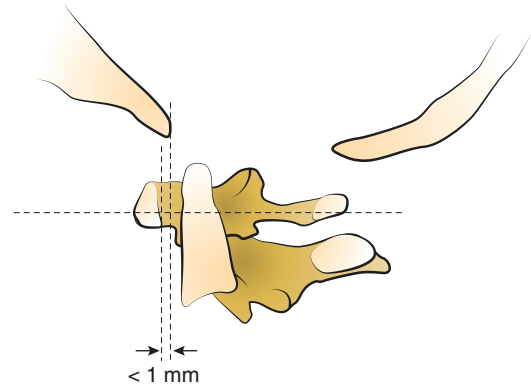



**Fig. 2.10** Wiesel-Rothman method for assessing occipitoatlantal instability. Lateral craniometry: a horizontal line is drawn that connects the anterior and posterior borders of the atlas. A perpendicular line is drawn touching the posterior border of the anterior arch. A second perpendicular line is drawn from the anterior border of the foramen magnum. In flexion and extension images the distance between the two perpendicular lines (distance from occiput with respect to the atlas) should not exceed 1 mm (Redrawn from: Wiesel SW, Rothman RH. Occipitoatlantal hypermobility. Spine. 1979;4:187–91)

The WBC count is normal or elevated, the CRP is normal in 60 % of cases, and the ESR is usually elevated. The blood culture is positive in 30 % of cases.

A bone scan will show increased local activity within a week. Radiological images show no abnormalities in the first instance. After 10–14 days X-rays show a narrowing of the intervertebral space with irregular end plates of the adjacent vertebral bodies. An MRI at an early stage shows an abnormal intervertebral disc before abnormalities can be seen on a bone scan.

**Disc calcification** The cause of disc calcification is unknown. Disc calcifications can occur in the entire spinal column and in several discs simultaneously. In 70 % of cases cervical (Fig. 2.9), mostly at the C6-C7 level and in 20 % in the thoracic spine and in 10 % at both levels. Lumbar manifestations are rare. In 60 % of cases the radiological manifestation of the disc calcification disappears spontaneously. In addition to a painful stiff neck, one quarter of children get a wry neck. In very exceptional situations there is a progressive neurological deterioration.

**Juvenile idiopathic arthritis** An acute stiff and a wry neck can be the first symptom in juvenile idiopathic arthritis, usually of the systemic or polyarticular type. Neck pain in the poly- or pauciarticular type is rare. A stiff and possible wry neck is rarely accompanied by pain. It is usually caused by destruction of the occipitoatlantal joints or as a result of atlantoaxial subluxation.

Supplementary assessment: in the case of a suspected occipitoatlantal or atlantoaxial subluxation, an anteroposterior and lateral X-ray of the cervical spine should be taken. A dens image (antroposterior X-rays of C1-C2 with an open mouth), and flexion and extension images of the cervical spine should be made. If abnormalities are seen on the X-rays additional CT-scans and MRIs should be taken. The Wiesel-Rothman method can be used to assess occipitoatlantal instability. On the flexion and extension images the distance from the occiput with respect to the atlas should not exceed 1 mm (Fig. 2.10). The atlantodental index (ADI) can be determined in the case of atlantoaxial instability. In children younger than age 8 this should be less than 4.5 mm and for older children it should be less than 2 mm (Fig. 2.11).

Spinal canals with an anteroposterior diameter (SAC = Space Available for Cord) of less than 13 mm have a stenosis of the spinal

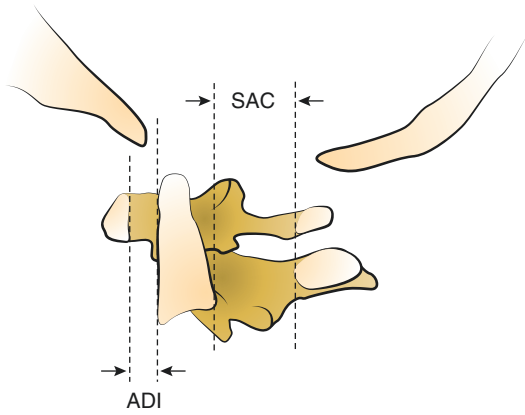



**Fig. 2.11** Atlantodental index for atlantoaxial instability. Lateral craniometry: the atlantodental index (ADI) is the distance between the posterior border of the anterior arch of the atlas and the anterior border of the dens. In children younger than 8 years this distance should be less than 4.5 mm and in children older than 8 years it should be less than 2 mm. Spinal canals with a anteroposterior diameter (*SAC* Space Available for Cord) of less than 13 mm have a canal stenosis with an increased risk of spinal cord compression (Redrawn from: Copley LA, Dormans JP. Cervical spine disorders in infants and children. J Am Acad Orthop Surg. 1998;6:204–14)

canal with an increased risk of spinal cord compression.

If a discitis is suspected it is prudent to do an MRI at an early stage in addition to determining the CRP and the ESR and taking blood cultures. Disc calcification can be easily recognized on lateral X-rays of the cervical spine. In juvenile idiopathic arthritis generalized abnormalities are often present shortly before or after the neck problems appear, so the diagnosis is not very difficult to make.

Primary care treatment: start by checking whether there is a throat infection (Grisel syndrome) or heartburn caused by gastro-esophageal reflux (Sandifer syndrome). These underlying causes must be treated first. In addition analgesics and a soft cervical collar are given for the causes mentioned above. The collar can be removed after disappearance of the symptoms. If X-rays show disc calcification this can also be treated with analgesics and a soft collar as long as symptoms are present. The worst complaints will disappear after 7–10 days. NSAID's can be given for juvenile idiopathic arthritis.

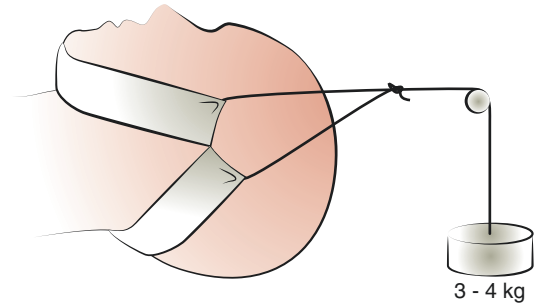



**Fig. 2.12** Glisson sling

When to refer: if there is no throat infection, no gastroeso-phageal reflux, no disc calcification or juvenile idiopathic arthritis, or if acute pain has lasted for more than a week.

Secondary care treatment: **occipitoatlantal and atlantoaxial subluxation.** Most patients whose symptoms have been present for less than a week can be treated with a soft collar. The soft collar is no longer required after the symptoms disappear. It is recommended to treat the subluxation by using traction with a Glisson sling if the symptoms have been present for more than a week, (Fig. 2.12). A CT-scan should be taken to assess whether the subluxation has been corrected. In this case, a follow-up treatment ensues with a soft collar for 6 weeks. The subluxation must be corrected non-operatively under anaesthetic, followed by immobilization for 3 months using a Minerva cast (Fig. 2.13) or a halo vest, if correction has not been achieved or the traction is poorly tolerated.

In occipitoatlantal subluxations a spondylodesis is almost never necessary. If the symptoms last for more than a month, then the chances of successful nonsurgical repositioning is slight and a spondylodesis must be carried out. If the atlantodental index (ADI) is between 5 and 10 mm, contact sports and diving must be avoided and even a prophylactic C1-C2 spondylodesis should be considered. In the case of an atlantodental index of more than 10 mm a C1-C2 spondylodesis is performed regardless of symptomatology.

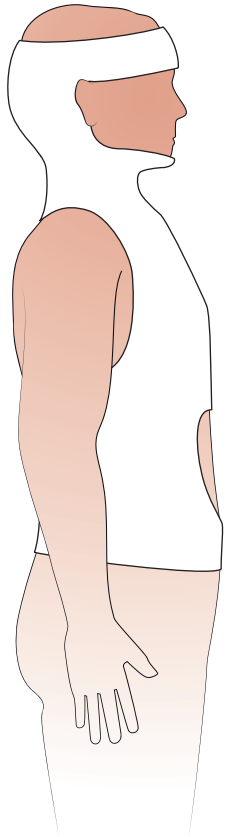


**Fig. 2.13** Minerva cast

**Spondylodiscitis** A discitis is treated with some form of immobilization and antibiotics. The staphylococcus aureus should in any event be sensitive to the administered antibiotics. This treatment is continued until CRP and BSE levels are normal. The process usually takes 6–8 weeks.

**Juvenile idiopathic arthritis** In severe abnormalities at the level of the craniocervical junction an occipitoatlantal or atlantoaxial spondylodesis or a combination of these can be considered.

## Slow Onset Painful Stiff and Possibly Wry Neck

Complaint: the mobility of the neck decreases gradually and at a certain point in time the patient will have a stiff neck. This may or may not be accompanied by a wry neck and pain.

Assessment: mobility of the cervical spine is severely limited or absent, and there may be a wry neck.

Differential diagnosis:

**metastases**
**primary bone tumor**
**primary spinal cord tumor**

Explanatory note: **metastases.** Metastases are the most frequent tumors (Table 2.2).

**Primary bone tumor** In children most primary bone tumors are benign (Table 2.3). All of these tumors may give compression, mostly to the spinal nerve roots and not the spinal cord.

**Primary spinal cord tumor** Most primary spinal cord tumors are malignant (Table 2.4). Spinal

**Table 2.2** Tumors that cause metastasis in the spine

| |
|---|
| Adenocarcinoma |
| Ewing sarcoma |
| Neuroblastoma |
| Rhabdomyosarcoma |

From: Freiberg AA, Graziano GP, Loder RT, Hensinger RN. Metastatic vertebral disease in children. J Pediatr Orthop. 1993;13:148–53

**Table 2.3** Primary spinal column tumors. The tumors identified with the § sign are rare

| Benign | Malignant |
|---|---|
| Aneurysmal bone cyst | Ewing sarcoma |
| Chordoma | Lymphoma § |
| Endochondroma | Osteosarcoma § |
| Eosinophilic granuloma | |
| Osteoblastoma | |
| Osteoid osteoma | |

Based on Adler CP, Kozlowski K. Primary bone tumors and tumorous conditions in children. Springer Verlag; 1993

**Table 2.4** Primary spinal cord tumors

| Benign | Malignant |
|---|---|
| Lipoma | Astrocytoma |
| Neurofibroma | Ependymoma |
| Spinal cyst | Oligodendroglioma |

**Table 2.5** Tumors with an increased risk of spinal cord compression in children

| Spinal cord compression |
|---|
| Astrocytoma |
| Lymphoma |
| Neuroblastoma |
| Sarcoma (in particular Ewing sarcoma) |

From: Conrad EU. Pediatric spine tumors with spinal cord compromise. J Pediatr Orthop. 1992;12:454–60

cord tumors at the cervical level are usually accompanied by muscle weakness and coordination disorders (Table 2.5). Sometimes they can be very slow-growing and only cause a stiff or possible wry neck as the sole symptom at the moment of assessment.

Supplementary assessment: in addition to X-rays, a CT-scan and MRI of the cervical spine and brain should be carried out.

Primary care treatment: the primary care provider, usually the general practitioner, must suspect that a single symptom such as only a slow onset stiff neck may be a sign of a serious abnormality.

When to refer: every patient with a slow onset stiff neck should be referred.

Secondary care treatment: **tumor.** Treatment of spinal column and spinal cord tumors must take place in a specialized medical center. The treatment is usually surgical, depending on the nature of the tumor and/or radiotherapy and/or chemotherapy may be necessary.

## Painful Stiff and Possibly Wry Neck with Neurological Symptoms

Complaint: painful, possibly wry neck accompanied by neurological symptoms.

Assessment: the most noticeable are symptoms of the neurological deterioration. There may be a nystagmus, cerebellar ataxia causing lack of ordered locomotion, swallowing disorders and symptoms of headache and dizziness.

Differential diagnosis:

**cervical kyphosis**
**basilar impression**
primary type
secondary type

**Arnold-Chiari malformation**
**stenosis of the foramen magnum**
**congenital occipitoatlantal synostosis**
**hypoplasia or aplasia of the dens**
**os odontoideum tumor**

Explanatory note: **cervical kyphosis** (see p. 8).

**Basilar impression** In a basilar impression the dens of the axis lies too high in the foramen magnum. As a result of this, in nearly 80 % of basilar impressions there is a short neck and in 70 % of cases a wry neck. Structures in the posterior cranial fossa (cerebellum and brainstem) have too little space because of the elevated position of the dens. There are two types of basilar impression: primary and secondary.

*Primary type* This is a congenital abnormality and is often associated with other anomalies of the neck vertebrae such as occipitoatlantal synostosis, an abnormal dens and the Klippel-Feil syndrome[4].

*Secondary type* This type is the result of weakening of the bony (base of the skull) structures as may be seen in rachitis and osteogenesis imperfecta[4]. In 85 % of basilar impression cases there is a paresis and paresthesia of the extremities. There may also be difficulties with swallowing resulting from compression of the cranial nerves IX, X and XII.

**Arnold-Chiari malformation** Arnold-Chiari malformation is caused by a caudal displacement of the tonsils of the cerebellum into the foramen magnum. This deformity is accompanied by a basilar impression in half of all cases. Symptoms tend to arise between the ages of 10 and 30. In addition to neck pain there is often a forced neck posture, accompanied by cerebellar and vestibular

[4]See Appendix.

symptoms such as lack of ordered locomotion, dizziness and nystagmus. Swallowing problems occur because of compression of the cranial nerves IX, X and XII. Arnold-Chiari malformation is very common in meningomyeloceles (spina bifida), and causes hydrocephalus in many of these children.

**Stenosis of the foramen magnum** In achondroplasia[5] one should consider a stenosis of the foramen magnum which can cause sleep apnea and even sudden death.

**Congenital occipitoatlantal synostosis** Congenital occipito-atlantal synostosis can be entirely symptom-free during one's entire life. Some patients can develop symptoms after a mild trauma. This abnormality can occur in isolated form, but also in addition in the Klippel-Feil syndrome[5].

**Hypoplasia or aplasia of the dens** Hypoplasia or aplasia of the dens (Fig. 2.8) is very common in children with spondylo-epiphysial dysplasia[5], Morquio syndrome[5] and Klippel-Feil syndrome[5]. A mild trauma can cause symptoms with neurological deterioration.

**Os odontoideum** Separation of the dens with respect to the axis can be the result of an accident (fracture), but can also be present as a congenital abnormality. Instead of the dens one speaks of an os odontoideum (Fig. 2.8). A relatively small trauma can cause symptoms that may vary from a stiff neck to complete quadriplegia.

**Tumor** There is a progressive movement limitation of the neck. In principle, all spinal column and spinal cord tumors can cause neurological symptoms. Some tumors can increase the risks for such symptoms (Table 2.5).

Supplementary assessment: in addition to anteroposterior and lateral X-rays, a CT-scan and MRI of the cervical spine are necessary.

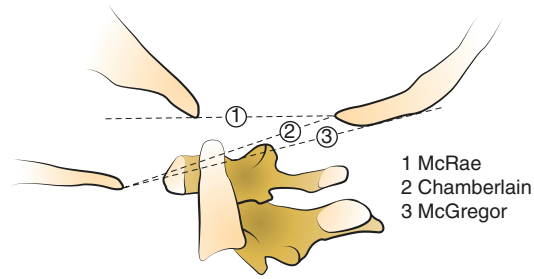



**Fig. 2.14** Lateral craniometry for basilar impression. The lines of McRae, Chamberlain and McGregor are drawn here. The McGregor line is the one that connects the anterior and posterior borders of the foramen magnum. If the dens lies distal to this line there will be no symptoms of a basilar impression. The Chamberlain line connects the posterior border of the foramen magnum with the most posterior border of the hard palate. In 50 % of children the top of the dens lies at the level of or distal to this line. If the dens projects more than 3 mm there may be a basilar impression. The McGregor line is drawn from the most posterior border of the hard palate towards the most caudal point of the occipital bone. In children the proximal part of the dens may project a maximum of 4.5 mm cranially. The McGregor line is the best method to screen for a basilar impression because at all ages the two points connected by the line are identifiable. Nevertheless, a lateral reconstruction of a CT assessment of the craniovertebral junction is often necessary in order to draw the lines correctly (Redrawn from: Copley LA, Dormans JP. Cervical spine disorders in infants and children. J Am Acad Orthop Surg. 1998;6:204–14)

A craniometry can be performed for a basilar impression on a lateral cranial X-ray or on a sagittal CT-scan; McGregor line is the best method to screen a basilar impression. The dens should not be more prominent than 4.5 mm above this line (Fig. 2.14). The os odontoideum is clearly visible on a dens image (anteroposterior X-ray of C1-C2 with an open mouth).

Primary care treatment: a severe abnormality should always be suspected if there are neurological symptoms.

When to refer: refer to a specialized center as quickly as possible.

[5] See Appendix.

Treatment in a specialized center: **cervical kyphosis.** (for treatment, see p. 8.)

**Basilar impression, Arnold-Chiari deformity, stenosis of the foramen magnum** The foramen magnum should be enlarged surgically in these cases.

**Congenital occipitoatlantal synostosis** If there is a congenital occipitoatlantal synostosis, head traction with a Glisson sling. Once the symptoms disappear a neck orthosis will be prescribed.

Operative treatment is indicated if the neurological symptoms persist. This involves carrying out a laminectomy and a C1-C2 spondylodesis. Morbidity and mortality for such an intervention is great.

**Hypoplasia or aplasia of the dens** A C1-C2 spondylodesis is indicated in a case with neurological problems.

**Os odontoideum** A halo vest may be given in the first instance for a fractured dens or a symptomatic os odontoideum. This immobilizes the head and neck. If symptoms persist or if they recur after removal of the halo vest a C1-C2 spondylodesis should be considered.

**Tumor** Treatment of benign as well as malignant tumors must take place in a specialized medical center. The treatment is mostly surgical and on indication radiotherapy and/or chemotherapy may be given depending on the nature of the tumor.

**Differential Diagnosis: Neck Deformity**

| **Neck flexion deformity** | |
|---|---|
| Instead of the lordosis there is a cervical kyphosis. | **Cervical kyphosis** |
| **Short neck** | |
| Short neck, limited mobility of the cervical spine, low hair growth into the back. | **Klippel-Feil syndrome (congenital brevicollis)** |
| **Wry neck** | **Torticollis** |
| Shortening of the sternocleidomastoid muscle from birth onwards. | Infantile muscular torticollis (congenital muscular torticollis) |
| Shortening of the sternocleidomastoid muscle appears at childhood. | Juvenile muscular torticollis |
| Abnormal vertebrae. | Osseous torticollis |
| Paresis of the extraocular muscles. | Ocular torticollis |

**Differential Diagnosis: Neck Pain**

| | |
|---|---|
| **Acute onset of a painful stiff and possibly wry neck** | |
| Down syndrome. | **Occipitoatlantal subluxation** |
| Down syndrome, spondyloepiphysial dysplasia, Morquio syndrome. | **Atlantoaxial subluxation** |
| Throat infection. | **Grisel syndrome** |
| Gastroesophageal reflux. | **Sandifer syndrome** |
| Fever . | **Spondylodiscitis (discitis)** |
| No fever. | **Disc calcification** |
| Generalized complaints, often shortly before or after the neck problems. | **Juvenile idiopathic arthritis** |
| **Slow onset painful stiff and possibly wry neck** | |
| Mobility decreases slowly. | **Metastasis** |
| | **Primary bone tumor** |
| | **Primary spinal cord tumor** |
| **Painful stiff and possibly wry neck with neurological symptoms** | |
| Instead of the lordosis there is a cervical kyphosis in: Campomelic dysplasia, Conradi-Hünermann syndrome, Larsen syndrome, neurofibromatosis. | **Cervical kyphosis** |
| Paresis and paresthesia of the extremities. There may also be swallowing disorders. | |
| | **Basilar impression** |
| Often associated with other anomalies of the neck such as occipitoatlantal synostosis, an abnormal dens and the Klippel-Feil syndrome. | Primary type |
| Result of weakening of the bony (base of the skull) structures as may be seen in rachitis and osteogenesis imperfecta. | Secondary type |
| Disordered locomotion, dizziness, nystagmus, swallowing disorders in: | |
| Spina bifida. | **Arnold-Chiari malformation** |
| Achondroplasia. | **Stenosis of the foramen magnum** |
| After a mild trauma in: Klippel-Feil syndrome. | **Congenital occipitoatlantal synostosis** |
| After a mild trauma in: | |
| Spondyloepiphysial dysplasia, Morquio syndrome, Klippel-Feil syndrome. | **Hypoplasia or aplasia of the dens** |
| After a mild trauma. | **Os odontoideum** |
| Progressive movement limitation of the neck. | **Tumor** |

# Back

# 3

## Back Misalignment

### Lateral Curvature(s) of the Back

Complaint: curved back.

Assessment: assessment shows one or two lateral curvatures in the spinal column.
Ask the patient to bend forward until the trunk is horizontal, hanging the arms down loosely with the palms facing each other (forward bending test). If the lateral curvature disappears and there is no hump, then there is a non-structural scoliosis. In a structural scoliosis the curve remains visible during the forward bending test, and any hump will also be more clearly visible (Fig. 3.1).

Differential diagnosis:

nonstructural scoliosis:
- **postural scoliosis**
- **compensatory scoliosis**

structural scoliosis:
- **congenital scoliosis**
- **idiopathic scoliosis**
  - infantile idiopathic scoliosis
  - juvenile idiopathic scoliosis
  - adolescent idiopathic scoliosis

Explanatory note: **scoliosis.** The anteroposterior projection of a normal spinal column is straight in the frontal plane. If the lateral curvature measured with the Cobb method is 10° or less (Fig. 3.2), we speak of a physiological asymmetry. A scoliosis entails one or more lateral curvatures of the spinal column with a Cobb angle greater than 10°.

*Nonstructural scoliosis* In a nonstructural scoliosis no gibbus is observable in the forward bending test. Manifestations of a non-structural scoliosis are postural and compensatory scoliosis.

**Postural scoliosis** If present, it is usually seen at the end of the first decade of life. There is a slight, usually left convex thoracic curve. It disappears when the child lies down and can be actively corrected when standing. When bending forwards there is no hump, and the lateral curve disappears completely (Fig. 3.3). This curvature never becomes structural. The lateral curvature disappears spontaneously as the child grows.

**Compensatory scoliosis** A difference in leg length causes a compensatory scoliosis only when standing. The convex side is on the side of the short leg. There is no torsional component. The scoliosis disappears when sitting, when lying down, also after correcting the difference in leg length, and also during the forward bending test (Fig. 3.4). This curvature never becomes structural either.

*Structural scoliosis* Manifestations of a structural scoliosis are congenital, idiopathic and neuromuscular.


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_3

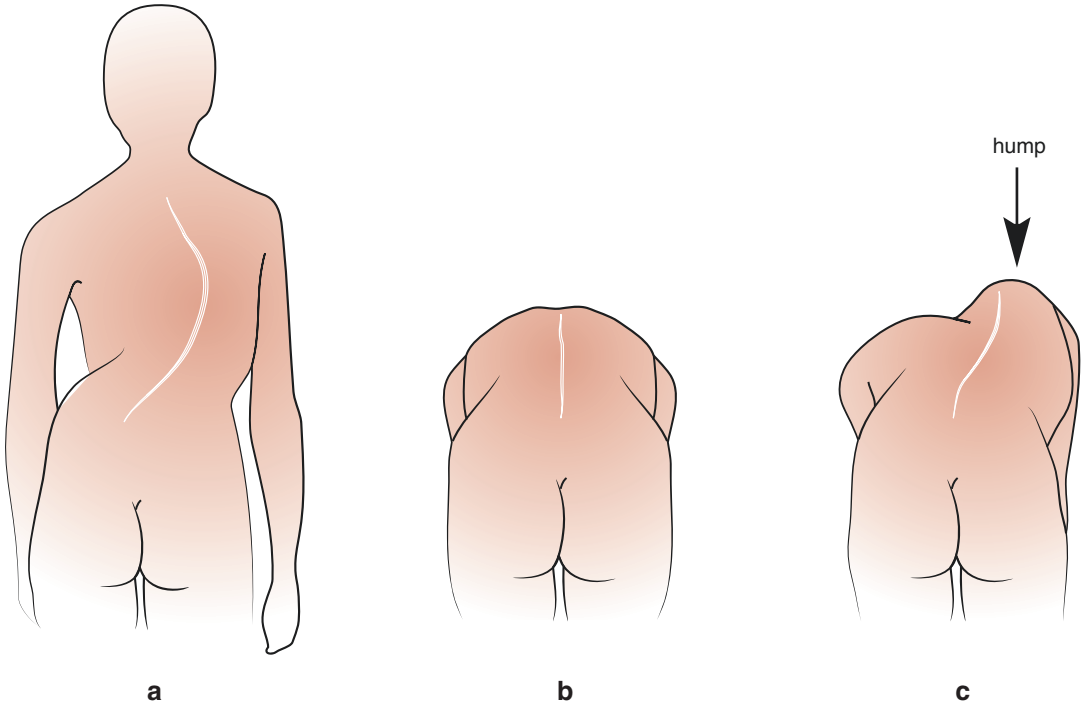



**Fig. 3.1** (**a**) Right convex thoracic scoliosis. (**b**) Nonstructural scoliosis. In the forward bending test there is no hump: on the contrary, the scoliosis disappears entirely. (**c**) Structural scoliosis. In the forward bending test a thoracic prominence (rib hump) is more visible

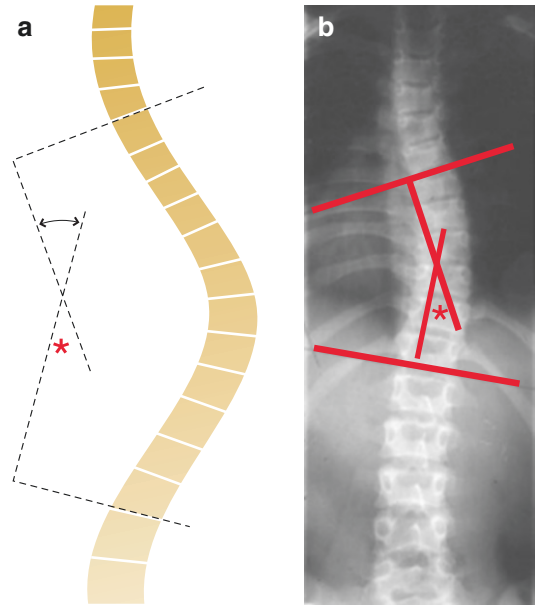



**Fig. 3.2** (**a**) The degree of lateral curvature (scoliosis) is measured according to the Cobb method, by drawing lines parallel to the superior endplate in the top vertebra and the inferior endplate of the lowest vertebra in the curvature. Then drawing perpendicular lines to these lines. The angle between these perpendicular lines, known as the Cobb angle (*), indicates the degree of the lateral curvature in degrees. (**b**) Scoliosis means one or more lateral curvatures of the spinal column, where the lateral curvature measured according to the Cobb method is 10° or more

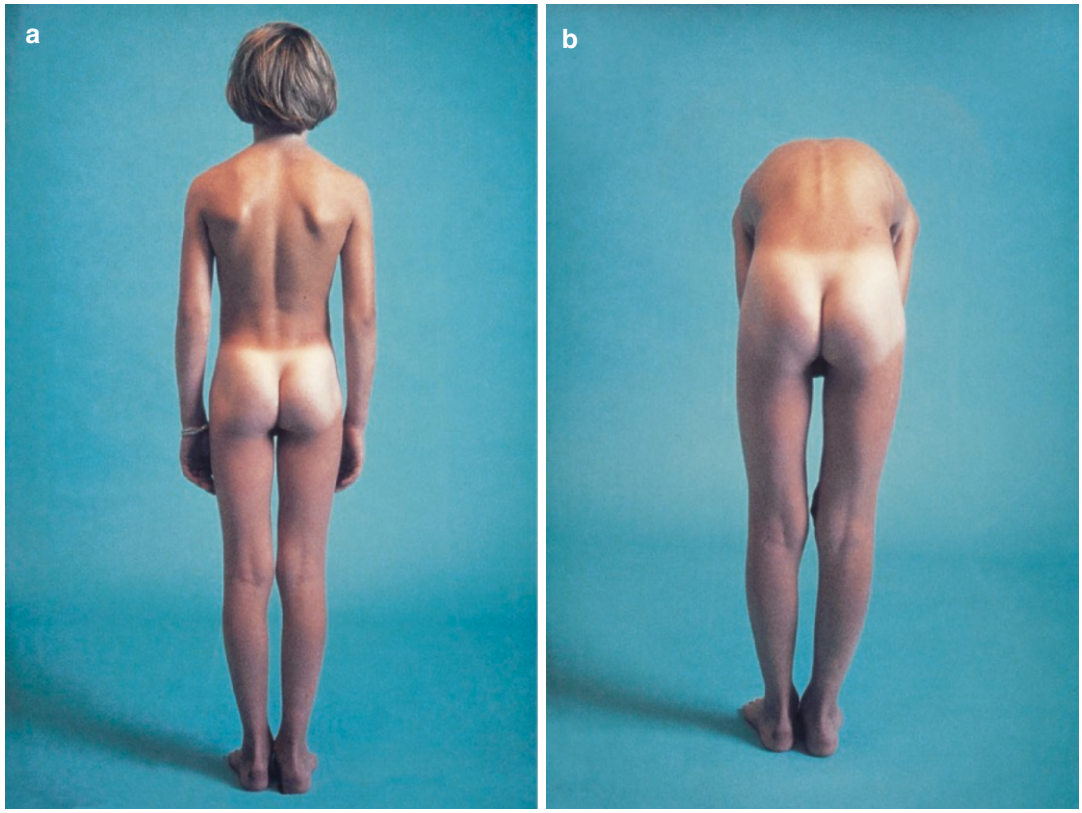



**Fig. 3.3** (**a**) Postural scoliosis, left convex thoracolumbar curvature. (**b**) The scoliosis disappears during the forward bending test

**Congenital scoliosis** This scoliosis occurs due to abnormal vertebrae such as hemivertebrae and unilateral nonsegmented bony connections (Fig. 3.5). In 75 % of these cases there is a progressive scoliosis of 5° per year, in 14 % there is slight progression, and only 11 % are non-progressive (Table 3.1). The abnormality occurs usually at the level of the thoracic spine. On physical assessment the scoliosis cannot be distinguished from a progressive infantile or juvenile idiopathic scoliosis (see below). Radiological assessment provides information. Children with a congenital scoliosis often have other abnormalities too (Table 3.2). The most dangerous accompanying abnormalities are congenital cardiac anomalies (5–10 %) and abnormalities of the urogenital system (20 %). In 10 % of cases there is a spinal dysraphia (Table 3.2).

**Spinal dysraphias** These are developmental disorders in the median line of the back. The most common is spina bifida. Diastematomyelia (Fig. 3.6), tethered cord (Fig. 3.7) and intraspinal lumbosacral lipoma (Figs. 3.8 and 3.9) are also spinal dysraphias. In half of all cases physical assessment shows signs that point to a spinal dysraphia. These can be skin abnormalities in the lower back in the midline such as enhanced hair growth (lumbar hypertrichosis), pigmented or depigmented naevi, port-wine stains, a sacral dimple, (Fig. 3.10) a fistula or a subcutaneous lipoma.

**Spina bifida** (Latin for split spine). This is a birth defect with an incomplete closing of the spine. There are three types: spina bifida occulta, meningocele and myelomeningocele. The most common location is the lumber spine. The spina bifida occulta has no or only skin

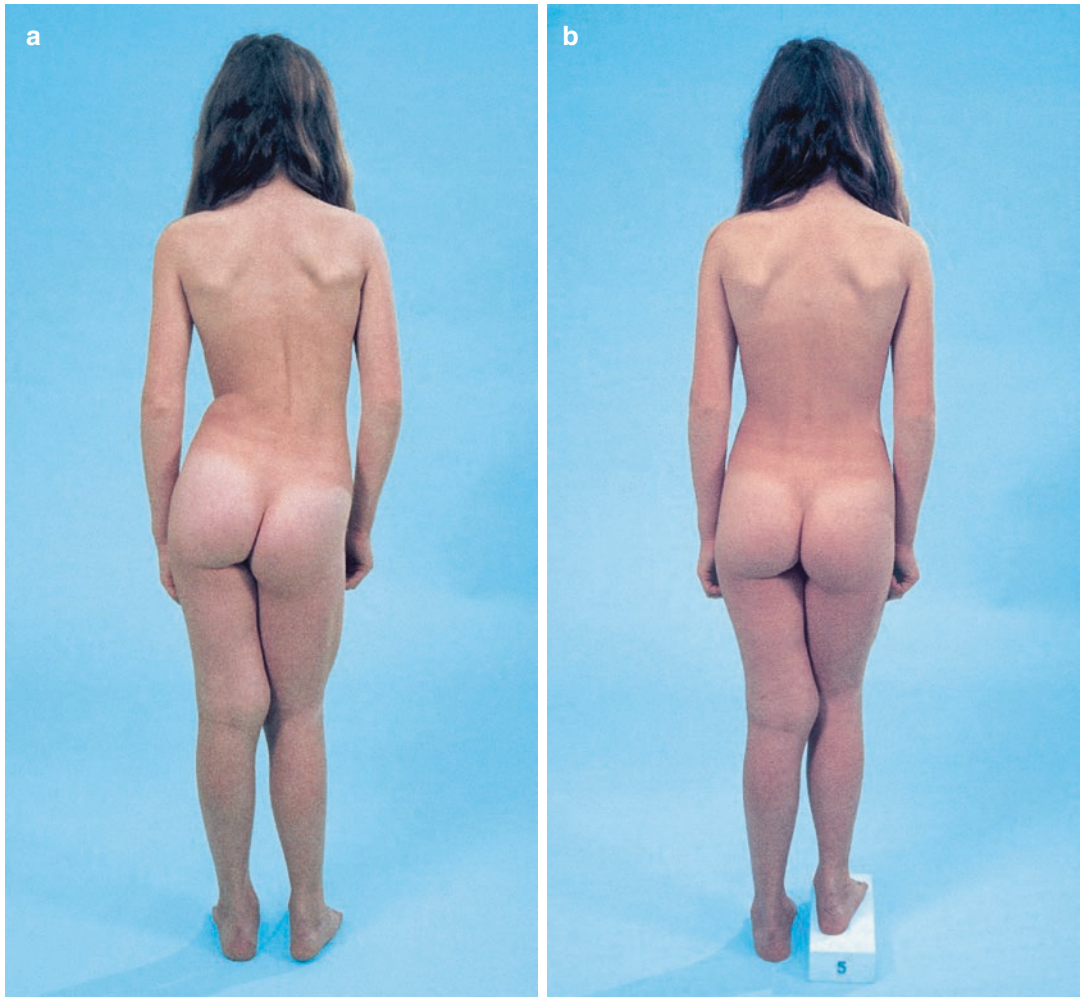



**Fig. 3.4** (**a**) Compensatory scoliosis, the convex side points to the leg that is too short. (**b**) The same patient as in Fig. 3.4a. The scoliosis disappears after correction of the leg length difference

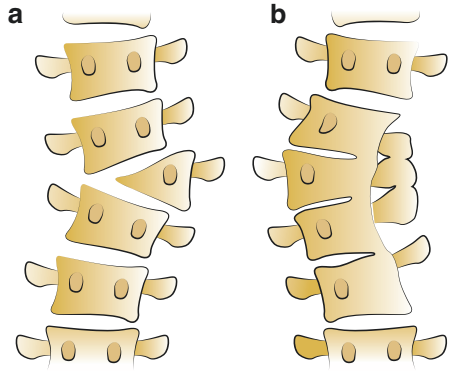



**Fig. 3.5** Congenital scoliosis. (**a**) Hemivertebra. (**b**) Non-segmented bony connections

**Table 3.1** Progression of the different scolioses

| | |
|---|---|
| Congenital scoliosis | 75 % strongly progressive |
| | 14 % lightly progressive |
| Infantile scoliosis | 15 % progressive |
| Juvenile scoliosis | 90 % progressive, especially after age 10 |
| Adolescent scoliosis | Depending on age and size of the curve at the time of discovery. See Table 3.2. |
| Neuromuscular scoliosis[a] | Nearly always progressive, pelvic obliquity |

[a]Neuromuscular scolioses are not covered in this book

signs at physical examination. A meningocele causes a sac of fluid at the gap in the spine. A myelomeningocele is the most severe form

associated with hydrocephalus, tethered cord, anomalies of the lower extremities, walking problems and problems with bladder and bowel control.

**Diastematomyelia** In a diastematomyelia (diastema is Greek for in-between space) there is a bony, cartilaginous or fibrous partition in the middle of the spinal canal (Fig. 3.6). As a result of traction on the spinal cord, which is fixed at the position of the partition in the spinal canal, pain and progressive neurological symptoms such as urinary problems, gait disorders and foot deformities occur in growing children.

**Table 3.2** Abnormalities associated with a congenital scoliosis

| Abnormalities |
|---|
| Abnormalities of the chest wall |
| Absence of lower extremity or parts of it |
| Clubfoot |
| Congenital heart abnormalities 5–10 %[a] |
| Congenital urinary tract abnormalities 20 %[a] |
| Cranial or head abnormalities |
| Ear abnormalities |
| Klippel-Feil syndrome[a] |
| Leg length inequality |
| Pelvic obliquity |
| Rib anomalies (extra or absent ribs, deformed or fused ribs) |
| Spina bifida occulta |
| Spinal dysraphia 10 % (spina bifida, diastematomyelia, tethered cord or lumbosacral lipoma) |
| Sprengel deformity |

[a]Most dangerous anomalies

**Tethered cord** Tethered cord may also described as an attached low medullary cone or a fixed myelum. The lowest cone-shaped end of the spinal cord (medullary cone) normally lies at the level of L3 in newborns and at the L1 or L2 in adults (Fig. 3.7). The distal end of the medullary cone is attached to the distal end of the spinal cord canal by a fibrous band (terminal filum). During growth the lower end of the myelum (spinal cord) does not shift cranially in a tethered cord. Adhesion of the cone often also occurs and the the filum is terminally shortened. During growth progressive neurological symptoms can develop as a result of traction on the spinal cord.

**Lumbosacral lipoma** The lipoma can usually be clearly seen as a subcutaneous swelling (Fig. 3.8). The extraspinally located part of the lipoma is often connected with a part located interspinally (Fig. 3.9).

**VATER or VACTERL association** In a congenital scoliosis one should also consider a VACTERL association (V stands for vertebral abnormalities, A for anal atresia, C for cardiac anomalies, T and E for tracheoesophageal fistula, R for renal anomalies such as urethral atresia and renal agenesis, and L for limb defects such as radial aplasia[1]).

**Idiopathic scoliosis** This is a scoliosis in which the cause is unknown. The curvature may consist of a single or double curvature (S-shaped). The position and direction of the lateral curvature

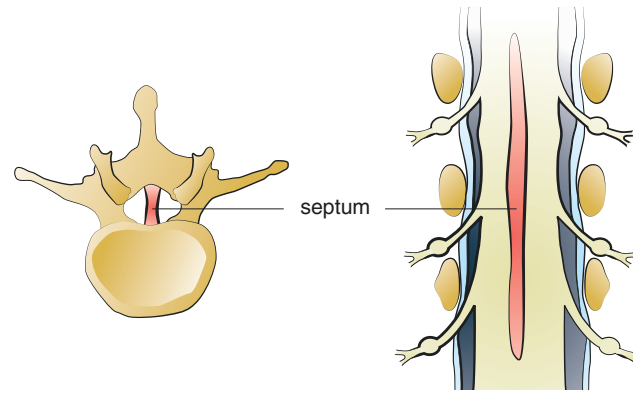



**Fig. 3.6** Diastematomyelia. A bony, cartilaginous or fibrous partition in the middle of the spinal canal

[1]See Appendix.

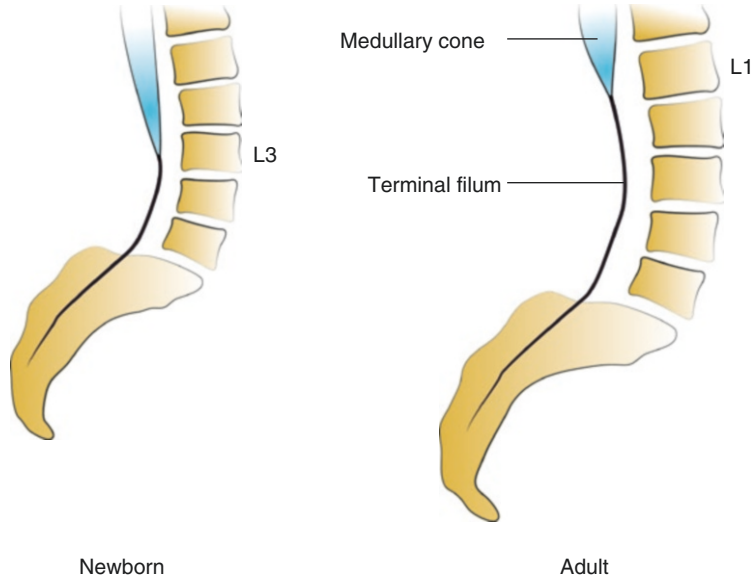



**Fig. 3.7** Tethered cord: the distal part of the medullary cone is attached to the distal end of the spinal cord canal with a fibrous strand (terminal filum). In newborns the distal end of the spinal cord (medullary cone) is at the level of L3. During growth the medullary cone ends up cranially. In adults at level LI-L2. In a tethered cord the medullary cone does not shift cranially during growth

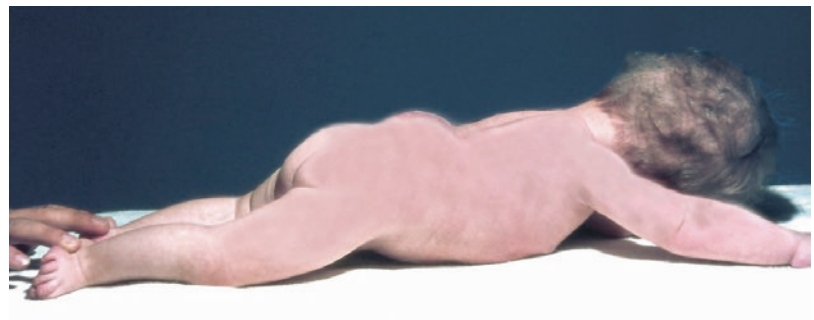


**Fig. 3.8** Lumbosacral lipoma

remains unchanged in the course of time, but if the anomaly increases several additional vertebrae may become involved. In single curvatures compensatory curvatures tend to appear, so that the spinal column remains in balance. If there is a loss of balance, misalignment can be assessed using a perpendicular line. Compensatory curves are in principle not structural but can become so with the passage of time.

In an idiopathic scoliosis there is a torsion abnormality in the transverse plane, in addition to an abnormality in the frontal plane. At the level of the thorax the torsion deformity manifests itself with prominence of the ribs on the convex side of the curve, known as a rib hump (Fig. 3.11). The torsion at the lumbar level causes a unilateral prominence of the back muscles. Idiopathic scoliosis is always accompanied by a torsion in the spinal column. The scoliosis can be classified according to the position of the lateral curves (Fig. 3.12). Idiopathic scoliosis affects 2–3 % of children, especially starting from 10 years of age. There is a strong congenital component. A prevalence of scoliosis is found in 18 % among first-degree relatives.

A classification has been made with the following three age groups because idiopathic scoliosis can occur at different ages:

- infantile idiopathic scoliosis from ages 0–3
- juvenile idiopathic scoliosis from 3 to about 10 years of age
- adolescent idiopathic starting from around the age of 10

Some scoliosis experts doubt whether juvenile scoliosis consists of a separate group and claim that this scoliosis may be a late-diagnosis infantile

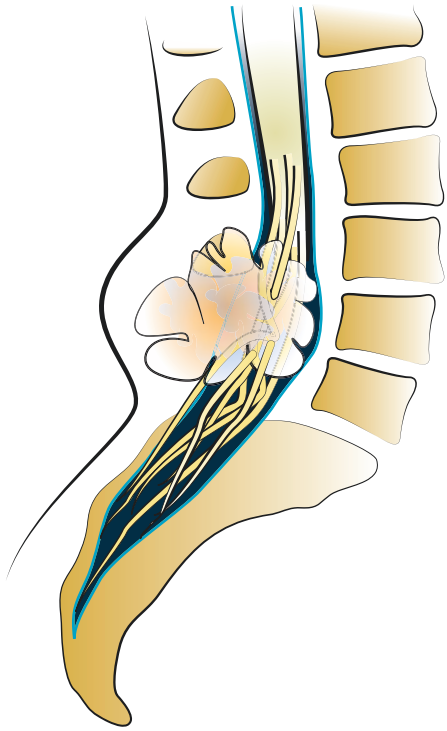


**Fig. 3.9** Lumbosacral lipoma: an extraspinally located part of the lumbosacral lipoma is often connected with an intraspinal part. This fixes the conus at a lower level and neurological symptoms can also occur

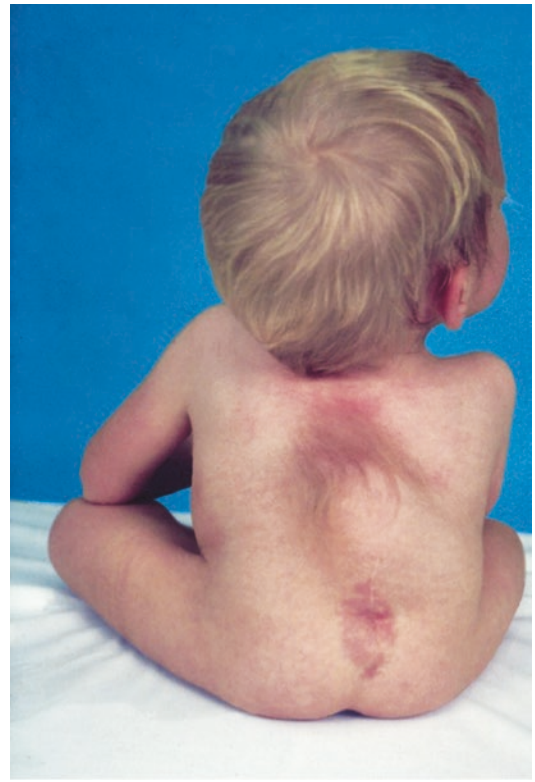


**Fig. 3.10** Hypertrichosis and a fistula

or an early-diagnosis adolescent scoliosis. In recent years these groups have been put into two groups, early onset and late onset. Some set the boundary at 5 years, therefore merging juvenile and adolescent scolioses. Other experts set the boundary at age of 8, merging infantile and juvenile scolioses.

*Infantile idiopathic scoliosis (ages 0–3)* This is rare and comprises 1 % of all scolioses. It affects boys more often than girls (3:2). The lateral curvature is convex to the left and thoracically localized (Fig. 3.13). Accompanying anomalies may be facial and cranial asymmetry (plagiocephaly), torticollis or developmental dysplasia of the hip. In 85 % of cases the curvature fortunately disappears before the first year of age. Spontaneous disappearance of the curvature never occurs if the curvature is more than 35°. In 15 % of cases the curvature does not disappear spontaneously and a severe deformity is the result (Table 3.1). It is important to make a distinction between nonprogressive and progressive infantile scoliosis. This is best achieved by requesting an anteroposterior X-ray of the spinal column and measuring the ribvertebral angle difference at the top of the curvature (RVAD or Metha angle). The angle between the spinal column and the rib, on both the left and the right sides, is measured. If the difference is more than 20° we are dealing with a progressive infantile idiopathic scoliosis in 90 % of cases. In 10 % of cases the scoliosis will disappear spontaneously (Fig. 3.14). If the RVAD is less than 20° spontaneous disappearance occurs in 90 % and in the remaining 10 % will become progressive. It is prudent to follow-up the child every 4 months radiologically from age 1 to age 2, to determine non-progression or progression.

*Juvenile idiopathic scoliosis (ages 3–10)* This involves 10–20 % of all idiopathic scolioses. This type of scoliosis is equally common among boys and girls. It usually involves a right convex thoracic scoliosis (Fig. 3.15) or a double curvature: thoracic convex to the right and lumbar

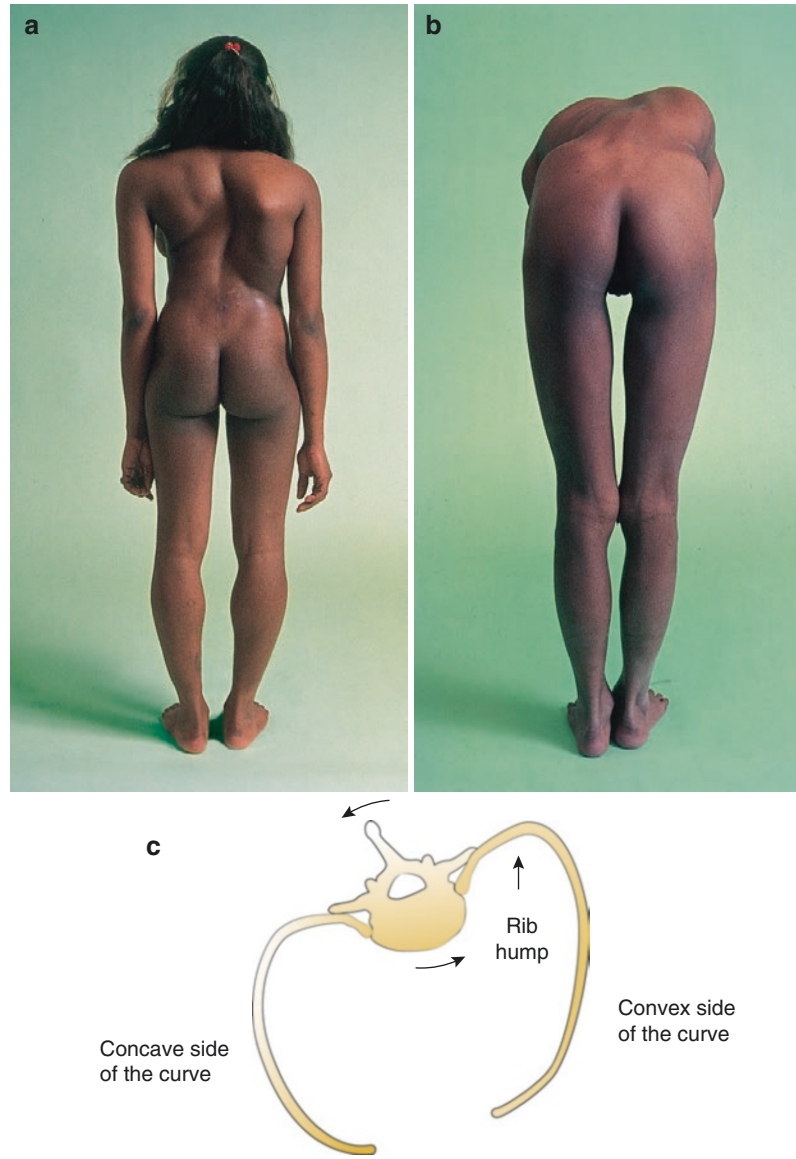



**Fig. 3.11** (**a**) Right thoracic scoliosis. (**b**) A thoracic prominence of the ribs (rib hump) is clearly visible during the forward bending test. (**c**) A rib hump occurs because an idiopathic scoliosis is always accompanied by a torsion in the spinal column. At the chest level this manifests itself by a bulge in the ribs on the convex side, which becomes clearer when bending over forward. Upon physical assessment the scoliosis seems less severe than on the X-rays because the spinous processes rotate toward the concave side in the arch

convex to the left. In girls between 6 and 10 years of age the curvatures are more severe than in boys. If there is a convex thoracic curve to the left at this age, intraspinal pathology should be ruled out with an MRI. This also applies to curvatures of more than 20° because in 25 % of such cases other spinal pathology will be found. The curvatures never disappear spontaneously as is the case in infantile idiopathic scolosis. About 90 % of juvenile lateral curvatures, are progressive and require treatment especially after 10 years of age (Table 3.1). If the rib-vertebral angle difference (RVAD) is less than 10°, there is generally no progression in the curvature. Progressive curvatures are mostly those with a rib-vertebral angle difference (RVAD) greater than 10°. This is particular so in the case of curvatures with an apex of the curvature between T8 and T10. This is also the case when the angle is more than 45° at the first consultation with a thoracic kyphosis of less than 20° (normal range 20–50°). Juvenile idiopathic scoliosis must be checked radiologically every 4 months to see if there is any progression.

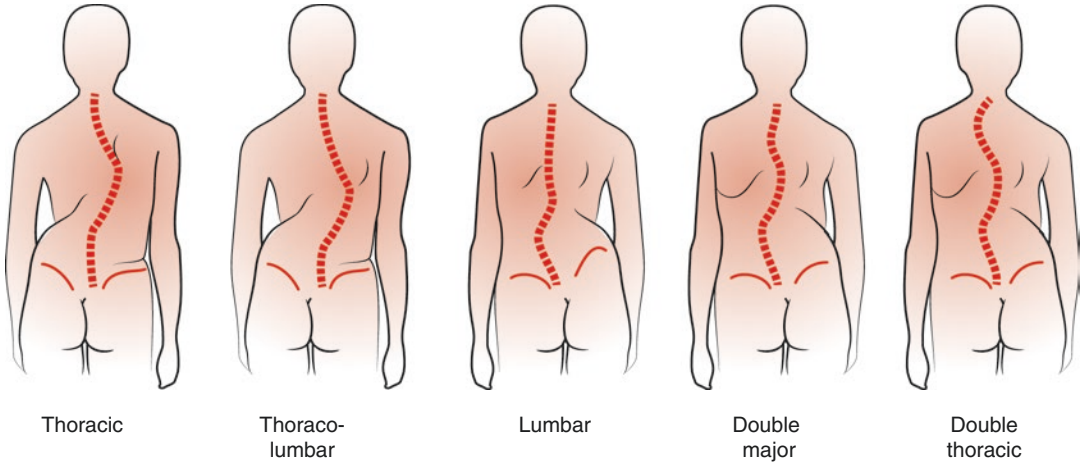



**Fig. 3.12** The scoliosis can be classified on the basis of the localization of the lateral single or double curvature

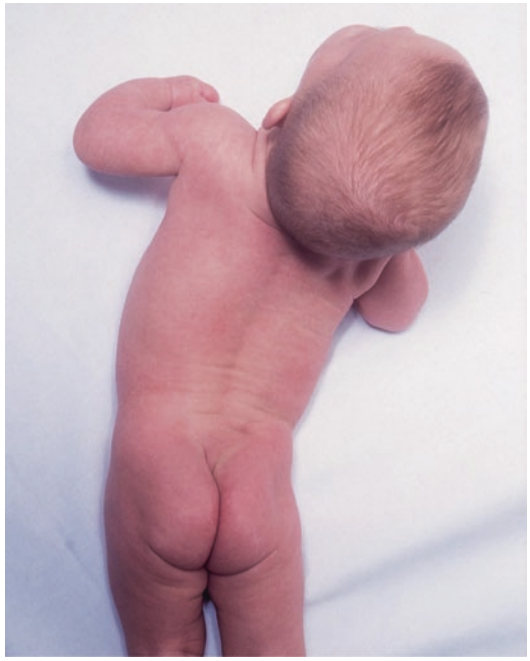


**Fig. 3.13** Infant with infantile idiopathic scoliosis, left convex lateral curvature

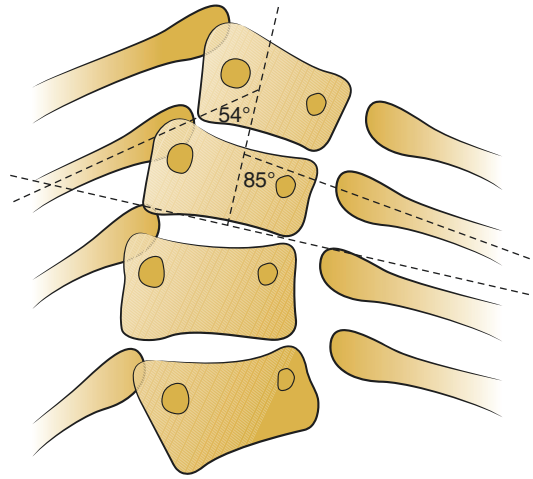



**Fig. 3.14** Rib-vertebral angle difference (RVAD or Metha angle). At the top of the curve the angle was measured between the spinal column and the rib, on both the left and the right side. If the difference is more than 20°, then we are most likely dealing with a progressive idiopathic scoliosis. In this case it is 85°−54° = 31°, therefore most likely progressive (Redrawn from: Metha MH. Rip-vertebra angle in the early diagnosis between revolving and progressive infantile scoliosis. J Bone Joint Surg Br. 1972;54-B:230–43)

*Adolescent idiopathic scoliosis (from the age of 10)* Scoliosis in adolescence is the most common type. In the age group between 10 and 15 years, 2 % have a lateral curvature between 10° and 20°, with a male:female ratio of 1:2. A curvature between 20° and 30° occurs in 0.3–0.5 % in this age group, with a male:female ratio of 1:5. A curvature of more than 30° occurs in 0.1–0.3 % of adolescents, with a male:female ratio of 1:10. The most common curvature is a right convex thoracic curvature (Fig. 3.16), followed by a double curvature thoracic to the right and lumbar to the left and in the third place a curvature with a left or right convex lumbar scoliosis. If there is a left convex thoracic curvature at this

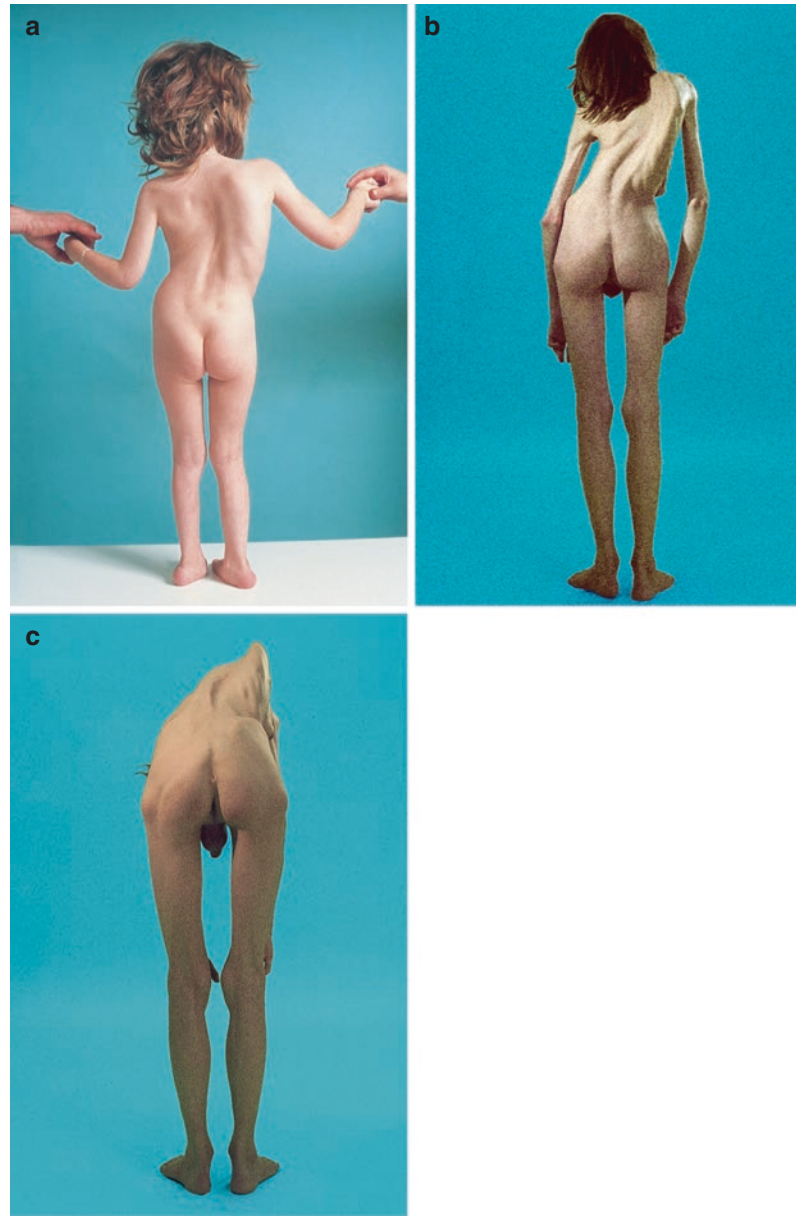



**Fig. 3.15** (**a**) Five-year-old girl with juvenile idiopathic scoliosis. The thoracic curvature is convex to the right. (**b**, **c**) Untreated juvenile idiopathic scoliosis

age, intraspinal pathology must be ruled out with an MRI. The first complaint is not always the lateral curvature of the spinal column but an elevated position of the shoulder with a crease on the concave side of the body or that one breast is positioned slightly more anteriorly than the other. Asymmetry of the air silhouettes between arms and trunk is often noticeable. The lateral curve may not be progressive. In some cases it is stationary. As a rule of thumb, the following applies: the greater the curvature at the moment it is identified, the greater the chances are of progression, especially in children between the ages of 10 and 12 (Table 3.3). On average, one can say that in children between the ages of 10 and 15, with curvatures less than 20° the chances of progression are 20 %, and in curvatures more than 20° the chances of progression are 70 %. It is impossible to predict which curvatures will remain stationary and which won't, although thoracic curvatures

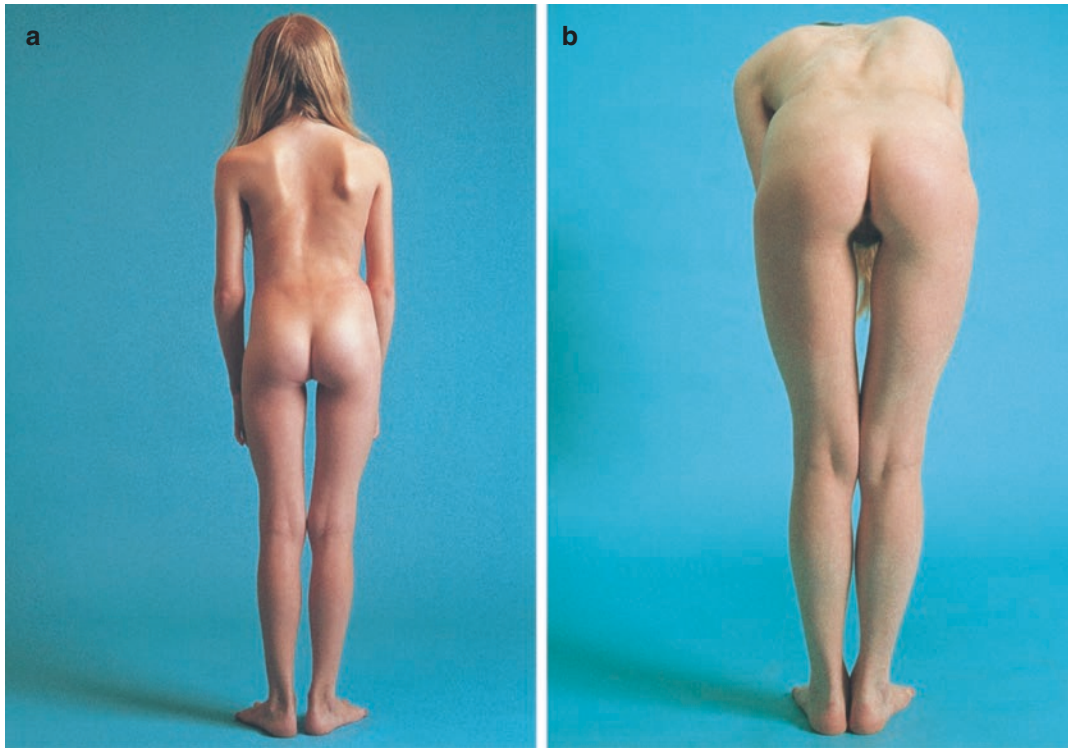



**Fig. 3.16** (**a**) Adolescent idiopathic scoliosis with a right convex thoracic curvature. (**b**) A rib hump is visible during the forward bending test

**Table 3.3** Progression of an idiopathic adolescent scoliosis related to the size of the lateral curvature and age at the first X-ray

| Size of the curve | Chances of progression at the age of | | |
|---|---|---|---|
| | 10–12 (%) | 13–15 (%) | 16 (%) |
| <19° | 25 | 10 | 0 |
| 20°–29° | 69 | 40 | 10 |
| 30°–59° | 90 | 70 | 30 |
| >60° | 100 | 90 | 70 |

From Nachemson AL, Lonstein JE, Weinstein S. Report of the prevalence and natural history committee of the Scoliosis Research Society. Denver: Read at the annual meeting of the Scoliosis Research Society; 1982

have greater chances of progression than lumbar ones. Curvatures less than 30° increase little during the rest of their lives after the children have grown up. A curvature between 30 and 50° increases 10–15 % after growth. However, if the scoliosis is more than 50° the lateral curvature increases due to premature degenerative changes in the intervertebral joints and the intervertebral discs. At the thoracic level this increase can even amount to 30° at an older age, with an average increase of 1° per year. In lumbar curvatures there is less of an increase, but at this level more pain is experienced with these curvatures than with thoracic curvatures. In curvatures less than 50° the chances of back problems is not greater than in individuals without a scoliosis. Cardiopulmonary problems only occur with curvatures more than 60–70°. A reduced life expectancy is only the case with curvatures of 100° or more.

Supplementary assessment: in a structural scoliosis, an anteroposterior and lateral X-ray of the entire thoracic and lumbar spine should be made. In addition an anteroposterior X-ray should be taken of the whole vertebral column (a so-called scoliosis X-ray). In idiopathic scoliosis, in addition to the lateral curvature there is also a torsion of the spinal column. This torsion can be detected based on the position of the pedicles (Fig. 3.17). In single curvatures

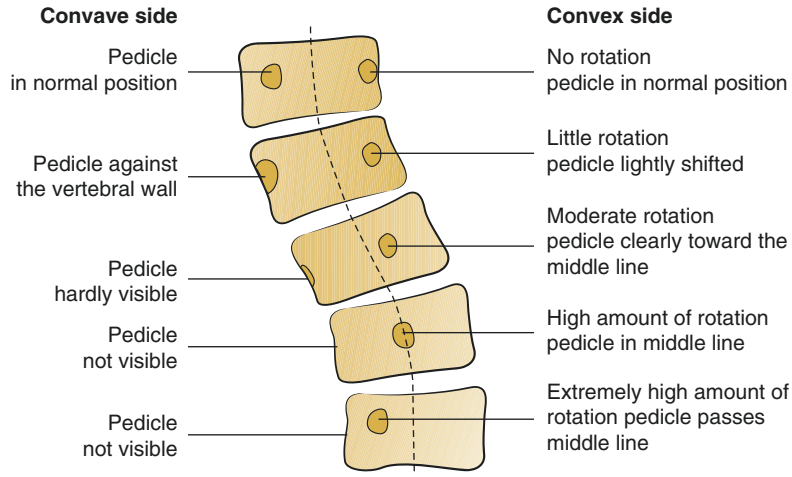



**Fig. 3.17** In a structural scoliosis there is torsion of the spinal column in addition to a lateral curvature. This rotation can be observed based on the position of the pedicles on an anteroposterior X-ray of the spinal column

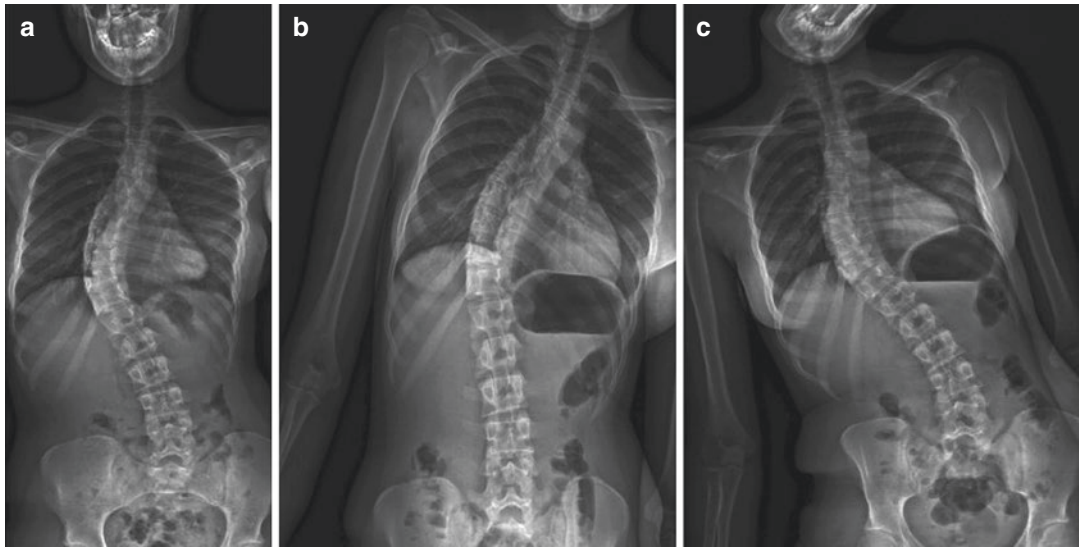



**Fig. 3.18** (**a**) In single curvatures a compensatory curvature can occur. Lateral flexion images of the spinal column to the left (**b**) and (**c**) to the right give insight into the suppleness of the arch. (**b**) The secondary or compensatory curvature (in this case lumbar) tends to completely corrected. (**c**) The primary or structural thoracic curvature cannot be corrected. The secondary lumbar curvature increases

a compensatory curvature may also be present. Lateral flexion images of the spinal column to the left and to the right give insight into the suppleness of the curve. A secondary or compensatory curvature tends to be completely straight (Fig. 3.18). If there is a congenital scoliosis, a CT-scan and MRI should also be taken. In infantile and juvenile scoliosis the rib-vertebral angle difference (RVAD) can be calculated (Fig. 3.14). A MRI should be requested to rule out intraspinal pathology if a left convex thoracic scoliosis is present in juvenile or adolescent patients. The same applies to juvenile scoliosis with a Cobb angle greater than 20°.

Primary care treatment: a lateral curvature due to a postural deviation will disappear

spontaneously. A compensatory scoliosis due to a difference in leg length can be corrected with an insole up to 1½ cm or a complete sole elevation up to 2 cm (for larger differences in leg length, see Chap. 15).

When to refer: a congenital scoliosis should always be referred to a pediatrician and an orthopedic surgeon specialized in spinal column surgery. In the case of an infantile idiopathic scoliosis that is still present at 1 year of age and a juvenile and adolescent scoliosis with a curve measured according to Cobb of 10° or more should be referred to an orthopedic surgeon. A compensatory scoliosis resulting from a difference in leg length of more than 2 cm is also an indication for referral, in order to assess whether operative correction of the leg length difference is required.

Secondary care treatment: **congenital scoliosis.** In a progressive congenital scoliosis a spondylodesis, combined if necessary with a hemivertebra resection at the level of the congenital abnormalities, is performed as soon as possible. A non-progressive congenital scoliosis should be monitored every 4 months for a few years, to be on the safe side.

**Idiopathic scoliosis** Scolioses with a lateral curve between 10° and 20° are checked once every 4 months by an experienced practitioner to assess possible progression in the lateral curve. If these check-ups show that the curvature is increasing, or if the practitioner has doubts about it, new X-rays must be taken.

*Infantile idiopathic scoliosis* In the case of progression a plaster cast or orthosis treatment is indicated. In the case of progression one should delay carrying out a spondylodesis if possible until after 10 years of age.

*Juvenile idiopathic scoliosis* This is usually progressive, especially after 10 years of age. The first step is to treat with a orthosis. The majority of these scolioses can undergo a spondylodesis after the age of 10.

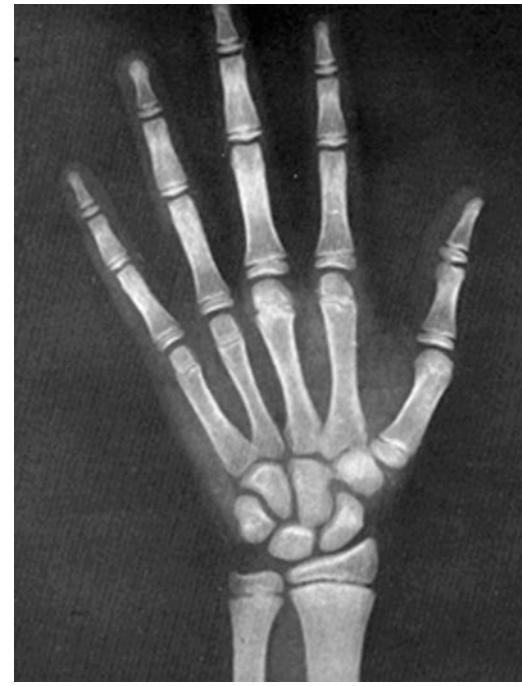


**Fig. 3.19** Hand skeleton X-ray. Skeletal age is determined based on an X-ray of the left hand. It is possible to establish the skeletal age by comparing the size and shape of the epiphyseal growth plates according to the atlas of Greulich and Pyle. The skeletal age based on this X-ray is 12½ years (Redrawn from: Greulich WT, Pyle SI. Radiographic atlas of skeletal development of the hand and wrist. 2nd ed. Stanford University Press; 1959)

*Adolescent idiopathic scoliosis* Progression in an adolescent idiopathic scoliosis usually occurs in early puberty, when armpit and pubic hair become manifest and external sexual characteristics develop, such as growth of the breasts and the testicles. In a progression of more than 5° a orthosis will be prescribed. The orthosis should be worn day and night, preferably for 23 h a day. The scoliosis and the orthosis are checked every 4 months. This treatment is continued until growth is complete. In girls growth stops at around the age of 14, in boys around the age of 16½. One gets and impression when growth is completed by measuring body height at each checkup. If the body height remains constant on two consecutive checkups, in girls aged 12½ or older and in boys aged 14½ or older, a skeletal image of the left hand can be made to see if growth has really stopped (Fig. 3.19).

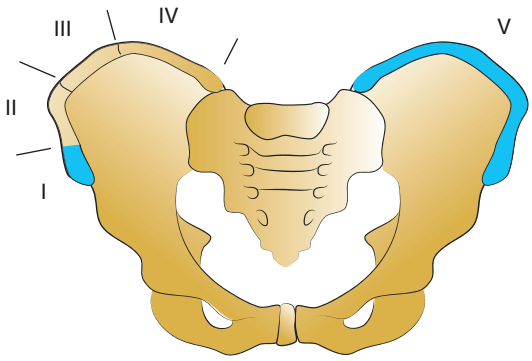



**Fig. 3.20** The Risser sign determines the degree of osseous maturity of the apophysis on an anteroposterior X-ray of the pelvis. The drawing shows stages I-IV on the right iliac crest, with a constant 25 % increase of ossification per stage. The ossification begins on the interior side of the pelvis. Risser I means that 25 % of the pelvis is ossified; this indicates the beginning of puberty. Risser IV means 100 % ossification, but no full fusion yet. Risser V (left pelvic half) shows full fusion of the apophysis with the ilium (Redrawn from: Risser JC. The iliac apophysis, an invaluable sign in the management of scoliosis. Clin Orthop Relat Res. 1958;11:111–9)

Another method is to establish the Risser stage, where the degree of ossification of the apophysis of the iliac crests is determined on X-ray (Fig. 3.20). If on the first appointment a curvature between 20° and 50° is found then orthosis treatment should be started immediately. A orthosis will not improve the scoliotic angle. The orthosis does prevent progression in about 80 % of cases.

The most common orthosis used is the TLSO (thoracolumbosacral orthosis) or a Boston orthosis (Fig. 3.21). This orthosis is effective for curvatures smaller than 35°. An alternative to the Boston orthosis is the Triac orthosis (Fig. 3.21). Both can only be used when the apex of the curvature lies at T8 or more distally. An extension to the chin is generally needed in curvatures where the apex lies cranially to T8. This is the case with the Milwaukee orthosis (Fig. 3.22). However this orthosis tends to be poorly tolerated. Besides this problems may occur with the mandibular heads. Fortunately, curvatures with an apex cranial to T8 are not common. The orthosis should be worn daily 22–23 h, and removed for sports.

In cases with a Cobb angle up to 50° the treatment is in fact cosmetic. If the Cobb angle is more than 50° the patient usually has pain or will have pain in the future. If the scoliotic curve is more than 50° it is advisable to perform a spondylodesis over the scoliotic trajectory, in girls preferably from the age of 12 and in boys from the age of 14. There are many methods to fix the spinal column, from posteriorly as well as anteriorly (Fig. 3.23). The purpose of a spondylodesis is to halt progression and diminish the size of the curvature whatever system has been chosen. Complications are infection, material fractures, material breakouts and pseudarthrosis. Reoperation is needed in 5–15 % of cases because of these complications. The most disastrous complication is irreversible spinal cord damage. However the risk is very small using the current techniques (about 1 in 1000 operated patients).

## Abnormal Rounded Back

Complaint: abnormal rounded back.

Assessment: at the thoracic level there is an enhanced posteriorly convex curvature of the spinal column. A normal convex curvature of the spinal column is known as kyphosis, an enhanced kyphosis is known as hyperkyphosis.

On physical examination a non-structural enhanced dorsal curvature is easily distinguishable from a structural one. The patient is observed from the side; you ask the patient to straighten the back by moving the shoulder backwards and lift the chin. A patient with an extremely rounded back due to a postural deformity can correct it himself in a standing position. Next, the child is asked to bend over forwards (side view, forward bending test). A rounded back in a postural deformity will show a symmetric contour (Fig. 3.24). The child is then asked to lie on his abdomen on the assessment table and must lift his head, chest and legs from the table. In the case of a postural anomaly the hyperkyphosis would disappear due to hyperextension of the back (Fig. 3.25). With small children it is best to place them on

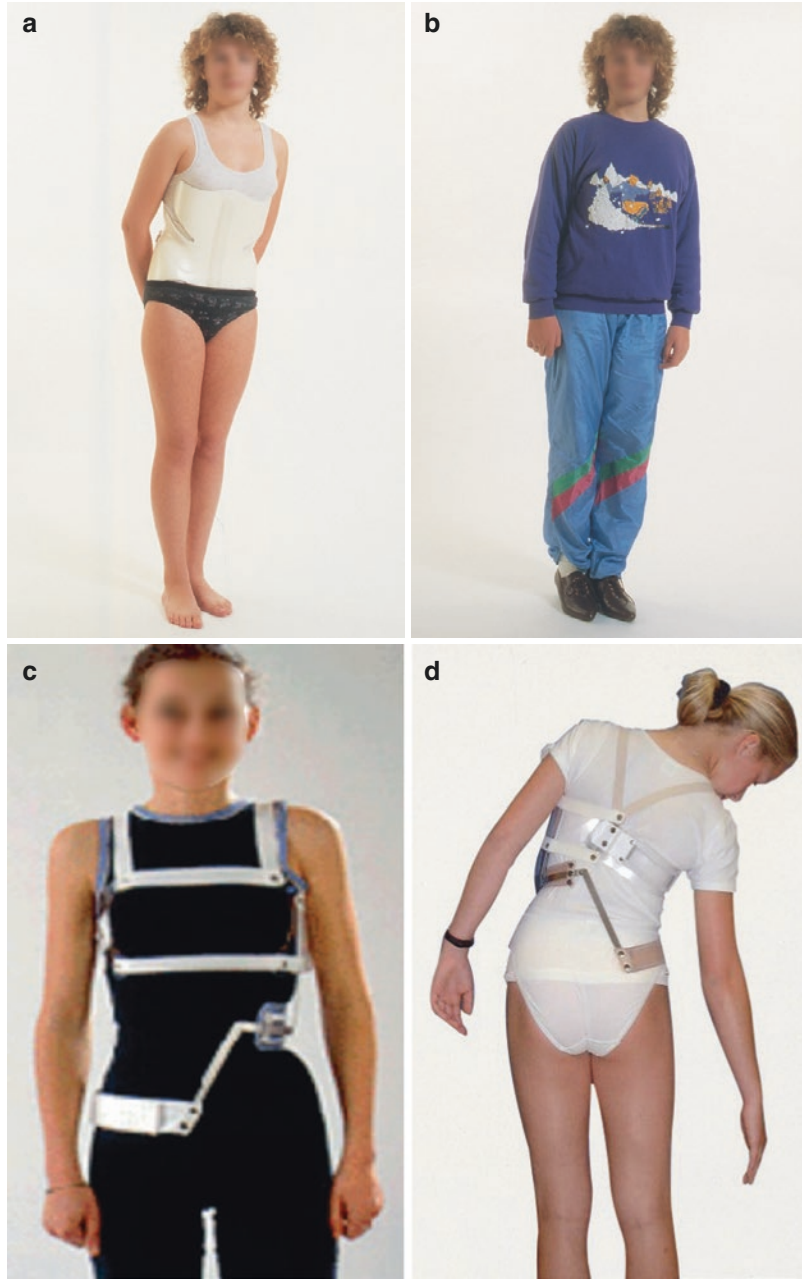



**Fig. 3.21** (**a**) Boston orthosis. (**b**) The orthosis is not noticeable when dressed. (**c**) Triac scoliosis orthosis. (**d**) This orthosis allows movements, in this case sideways to the right

their abdomen and extend the back by lifting the head and legs.

In a structural hyperkyphosis the patient is not able to actively correct the hyperkyphosis after carrying out the abovementioned tests. When the child bends over forward (side view, forward bending test) a kink in the contour of the spinal column becomes visible.

Differential diagnosis:

nonstructural hyperkyphosis
**postural hyperkyphosis**
structural hyperkyphosis
**congenital hyperkyphosis**
**Scheuermann disease**
**idiopathic hyperkyphosis**

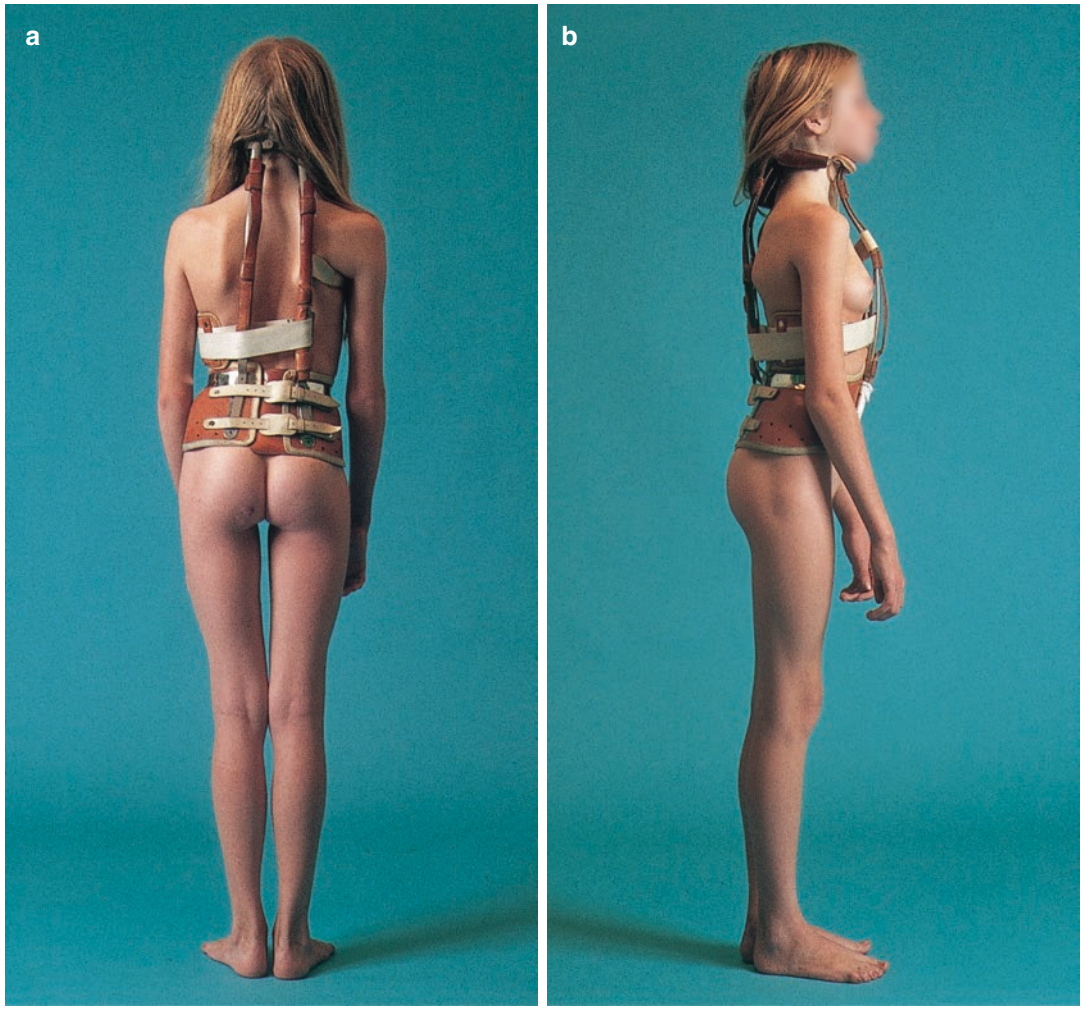



**Fig. 3.22** (**a**, **b**) In curvatures were the apex lies proximal to T8, extension of the orthosis to the chin is necessary: this is known as the Milwaukee orthosis

Explanatory note: **hyperkyphosis.** We speak of a hyperkyphosis when a thoracic kyphosis of more than 50° is present

*Nonstructural hyperkyphosis* In a nonstructural hyperkyphosis a smooth contour in the spinal column is visible on a side view in the forward bending test.

**Postural hyperkyphosis** This usually involves girls around the age of puberty with beginning breast development. They can get really worried about this probem. There are also boys and girls who think they are too tall. The postural anomaly can be actively corrected. The forward bending test shows a normal back contour. After puberty the hyperkyphosis usually resolve spontaneously.

*Structural hyperkyphosis* In a structural hyperkyphosis a forward angulated pattern in the spinal column contour is visible during the forward bending test (Fig. 3.24).

**Congenital hyperkyphosis** This can develop in two ways: either due to a absent part of a vertebral body or due to failing segmentation on the anterior side of the vertebrae (Fig. 3.26). Severe types can already be recognized at birth. The less obvious deformities tend to be detected later. These abnormalities will never resolve

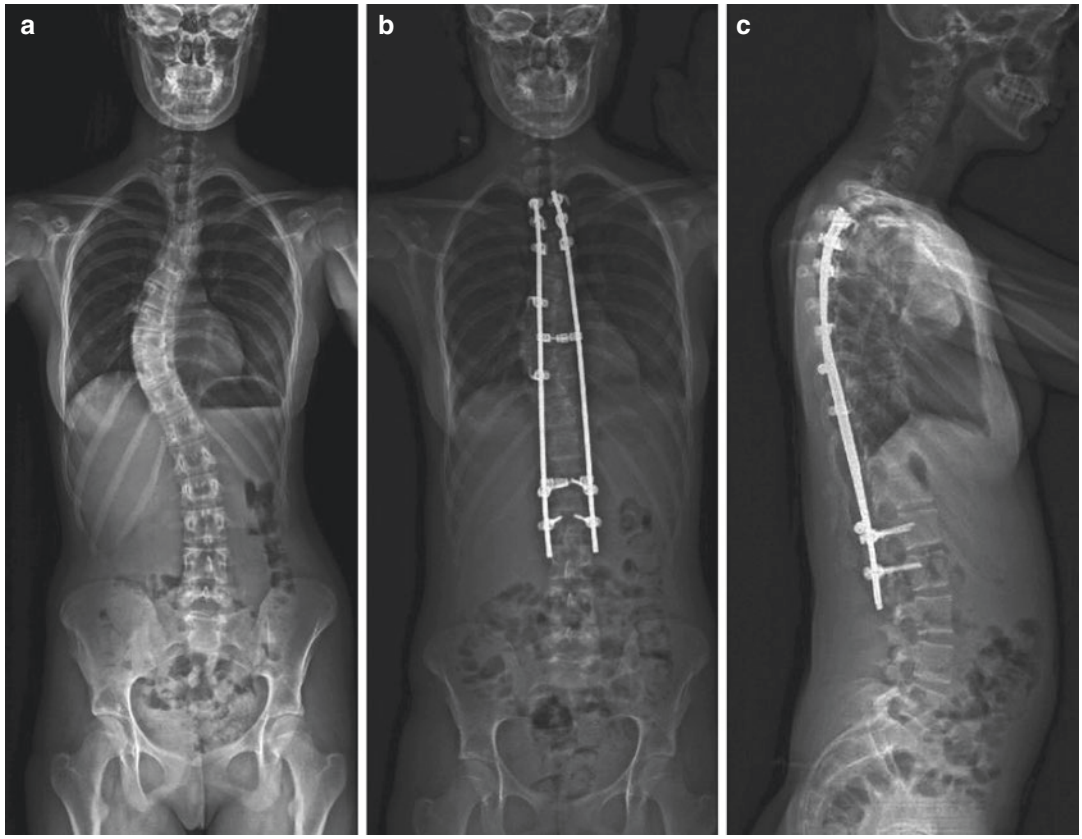



**Fig. 3.23** (**a**) Idiopathic adolescent thoracolumbar scoliosis. (**b**, **c**) The same patient as in Fig. 3.23a after a posterior spondylodesis.

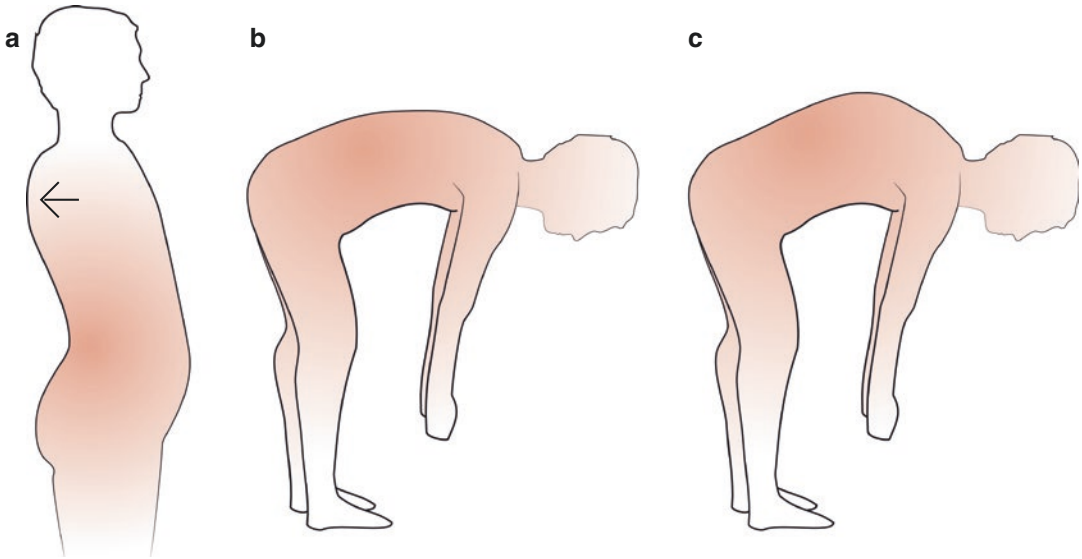



**Fig. 3.24** (**a**) Thoracic hyperkyphosis. (**b**) Nonstructural hyperkyphosis. When bending over forward (forward bending test) the back shows a smooth contour. (**c**) Structural hyperkyphosis. An angular apical kyphosis is visible in the contour of the spinal column when bending over forward

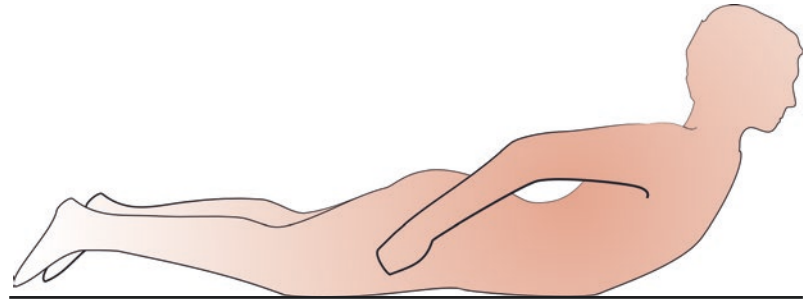


**Fig. 3.25** By lifting the head, chest and legs from the examination table the back is extended and the hyperkyphosis disappears in a nonstructural hyperkyphosis. It remains in a structural hyperkyphosis

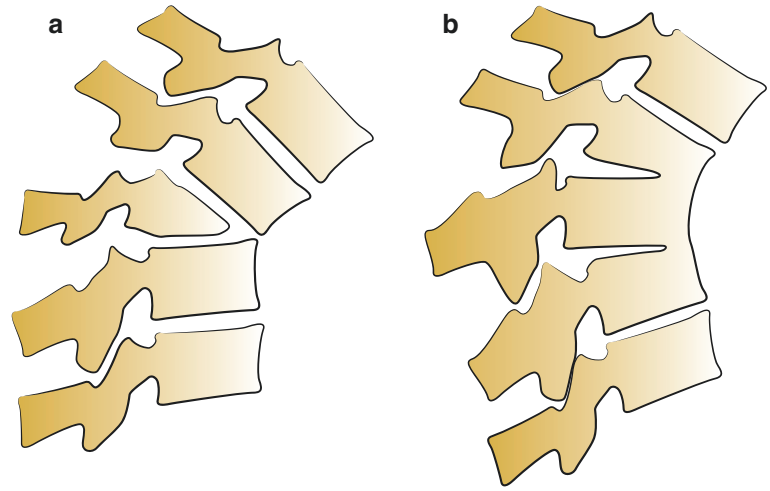



**Fig. 3.26** A structural hyperkyphosis can occur due to congenital anomalies of the spinal column. (**a**) A part of the vertebral body is absent on the anterior side. (**b**) The vertebral bodies on the anterior side are not segmented. The posterior side keeps growing, but the anterior side remains the same, which results in a structural hyperkyphosis

spontaneously. They will be progressive until growth stops. Compression of the spinal cord can occur causing neurological symptoms, especially if the anomalies are localized at the thoracic level.

**Scheuermann disease** Scheuermann disease is a relatively common deformity with a prevalence between 4 and 8%. It is as common in both boys and girls, and is usually identified between the ages of 12 and 15. Forward-hanging shoulders are visible (protraction) (Fig. 3.27). In addition to a forward angulated pattern in the contour of the spinal column during the forward bending test, the patient is generally unable to touch the ground with his fingers when bending forward due to shortened hamstrings. The cause of Scheuermann disease is unknown. On X-rays one can see that the anterior side of several adjacent vertebral bodies is irregular at the level of the endplates. As the disease progresses wedge-forming of the vertebral bodies occurs (Fig. 3.28). A common definition of Scheuermann disease is also: an increased thoracic kyphosis involving at least three consecutive vertebrae, each with a kyphotic angle of 5° or more. The apex of the hyperkyphosis is mostly localized at the level of T9. In 30% of cases there is also a mild scoliosis. Children sometimes complain of back pain. This usually disappears in adulthood. The pain persists only in very few cases.

**Idiopathic hyperkyphosis** Adolescents may show a hyperkyphosis without the radiological characteristics of Scheuermann disease. This is generally a familial form.

Supplementary assessment: in a structural hyperkyphosis, anteroposterior and lateral X-rays of the thoracic and lumbar spines and a lateral X-ray of the entire spinal column should be made. Supplementary CT-scan and MRI should be made in congenital hyperkyphosis

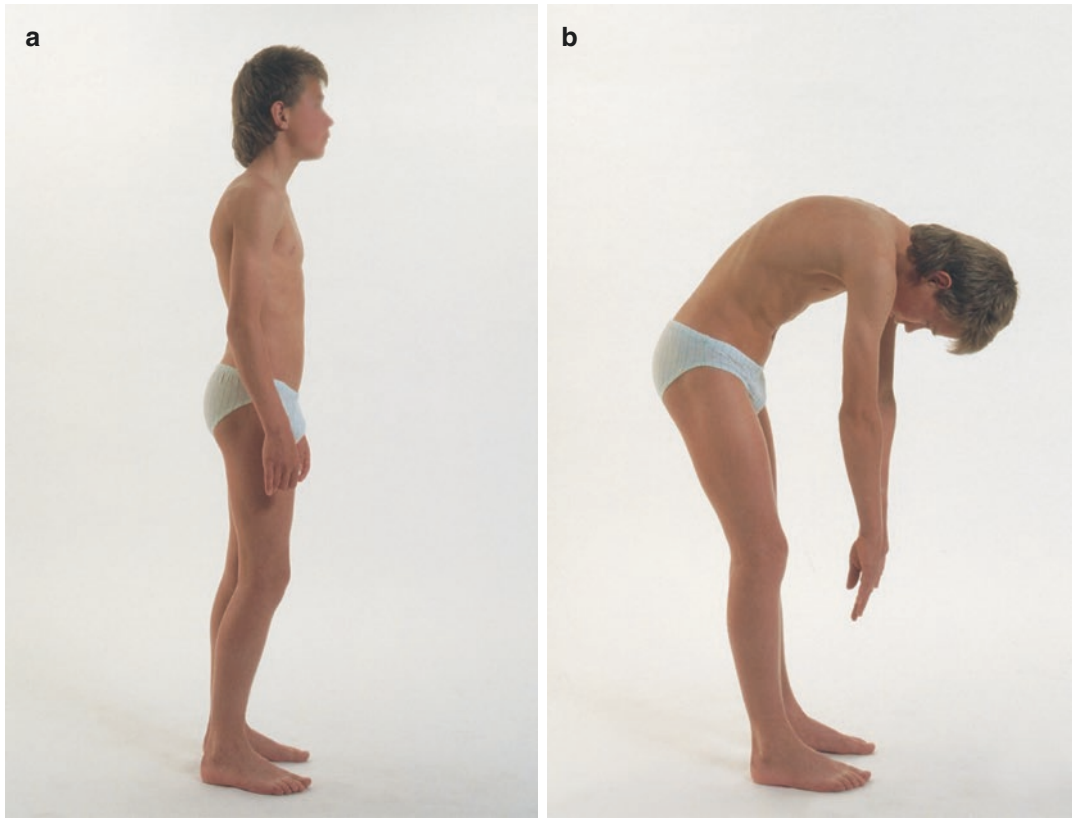



**Fig. 3.27** (**a**) A 14-year-old boy with Scheuermann disease. The forward-hanging shoulders are typical. (**b**) In addition to an angular pattern in the contour of the back when bending over forward (forward bending test), due to shortened hamstrings, the patient cannot usually touch the ground with his fingers

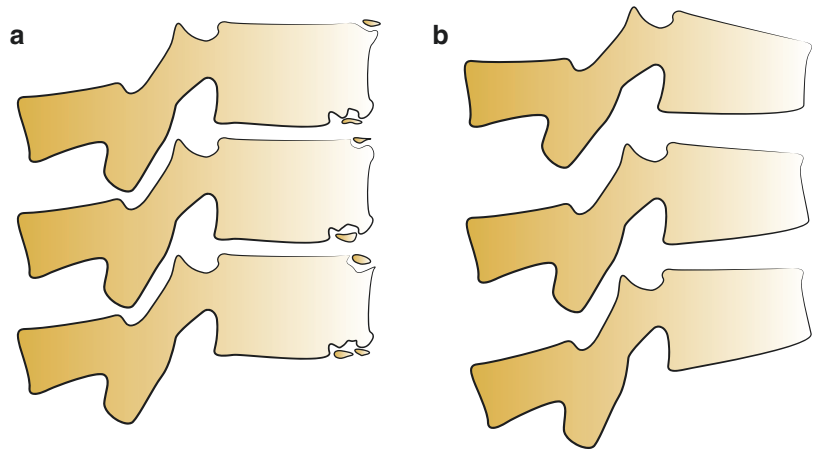



**Fig. 3.28** (**a**) In Scheuermann disease we see that in the initial stage the interior side of the vertebral bodies at the level of the endplates are irregular (apophyseal growth disorder). (**b**) Wedge formation of the vertebral bodies occurs if the disease is not treated

cases. The kyphotic angle can be measured on the lateral X-rays by calculating the Cobb angle. A thoracic kyphosis usually measures between 20 and 50°. If the kyphotic angle is greater than 50° there is a hyperkyphosis (Fig. 3.29). In the early stage of Scheuermann disease there are only irregular endplates on the anterior side of the vertebral bodies with an enhanced thoracic

kyphosis. In a more advanced stage there is also wedgeshape of the vertebral bodies. The anterior side of the vertebral body is less elevated than the posterior side. The intervertebral discs can also be narrow and there may also be vertical herniation of disc material (Schmorl nodes through the endplate).

Primary care treatment: a child with a postural abnormality must be encouraged to stand up straight. The child can also get instructions for exercises from a paediatric physiotherapist. There is no point in treating the abnormality with a plaster of Paris corset, a orthosis or a muscle stimulator. It is most important that the the physician can distinguish a postural anomaly from a structural hyperkyphosis.

When to refer: a structural hyperkyphosis should be referred.

Secondary care treatment: **congenital hyperkyphosis.** Exercises and orthosis therapy are pointless. Operative treatment is necessary. A posterior spondylodesis, if necessary combined with an anterior spondylodesis, should be performed over a trajectory as short as possible. If there are neurological symptoms an anterior decompression should be carried out.

**Scheuermann disease** Exercise therapy does not lead to improvement in this hyperkyphosis. An orthosis is required to prevent progression of the abnormality. A Milwaukee orthosis is provided in case of a curvature with the apex cranial to T8 (Fig. 3.22). A thoracolumbosacral orthosis with a sternal pressure pad is used for more caudal curvatures (Fig. 3.30). The orthosis should

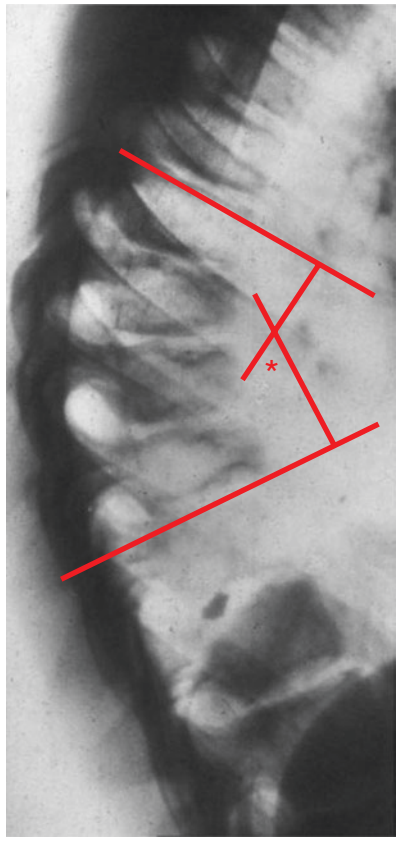



**Fig. 3.29** The thoracic kyphotic angle can be measured using the Cobb method (*). Normally there is a kyphosis of 20–50°. We speak of a hyperkyphosis if the kyphotic angle is larger than 50°. In this case the kyphotic angle is 65°

**Fig. 3.30** Thoracolumbosacral orthosis with sternal pressure pad, preventing forward bending

be worn daily 22–23 h, but should be removed during sports. The treatment should be continued until the patient stops growing. Operative treatment is recommended in very exceptional cases, with a curvature of more then 80°.

**Idiopathic hyperkyphosis** This is also treated with a orthosis.

## Abnormal Hollow Back

Complaint: abnormal hollow back.

Assessment: at the lumbar level there is too much ventral curvature in the spinal column.

Differential diagnosis:

nonstructural hyperlordosis
**postural hyperlordosis**
compensatory hyperlordosis
**spondylolisthesis**
**flexion contracture of the hip(s)**
structural hyperlordosis
**congenital hyperlordosis**

Explanatory note: **hyperlordosis.** Lumbar lordosis is a physiological antero-posterior curvature of the spinal column with an anterior convex curvature of 20–50°. An enhanced anterior convex curvature of the spinal column is called hyperlordosis.

*Nonstructural hyperlordosis* In a nonstructural hyperlordosis the hyperlordosis disappears during the forward bending test (Fig. 3.31). Non-structural hyperlordosis occurs as a postural anomaly or as a compensatory hyperlordosis with a spondylolisthesis or in flexion contractures of the hips.

**Postural hyperlordosis** This affects children with an enhanced forward-tilting pelvis, enhanced lumbar lordosis, sunken shoulders, a forward-tilting head and generally flatfeet. The entire posture changes immediately when the pelvic tilt is corrected. This can be achieved by making the child sink to its knees. The posture can also be easily corrected actively. The hyperlordosis disappears when viewed from the side during the the forward bending test (Fig. 3.31).

*Compensatory hyperlordosis* This is found in spondylolisthesis and in flexion contractures of the hip(s).

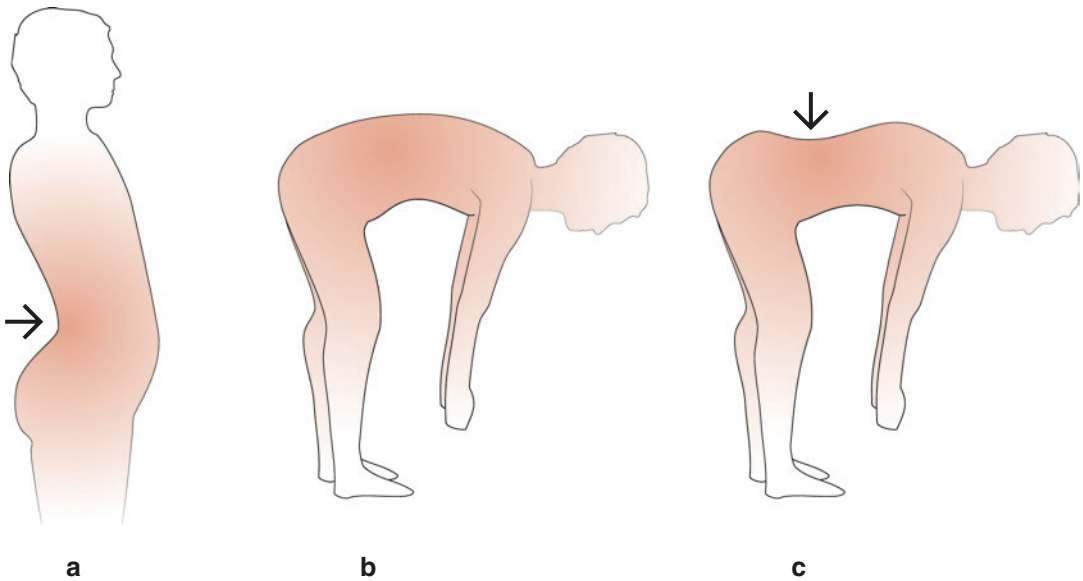



**Fig. 3.31** (**a**) A hyperlordosis. This is most common in children with a forward-tilting pelvis. (**b**) Nonstructural hyperlordosis. When bending over forward (forward bending test) the lordosis disappears. (**c**) Structural hyperlordosis. The lordosis remains when bending over forward

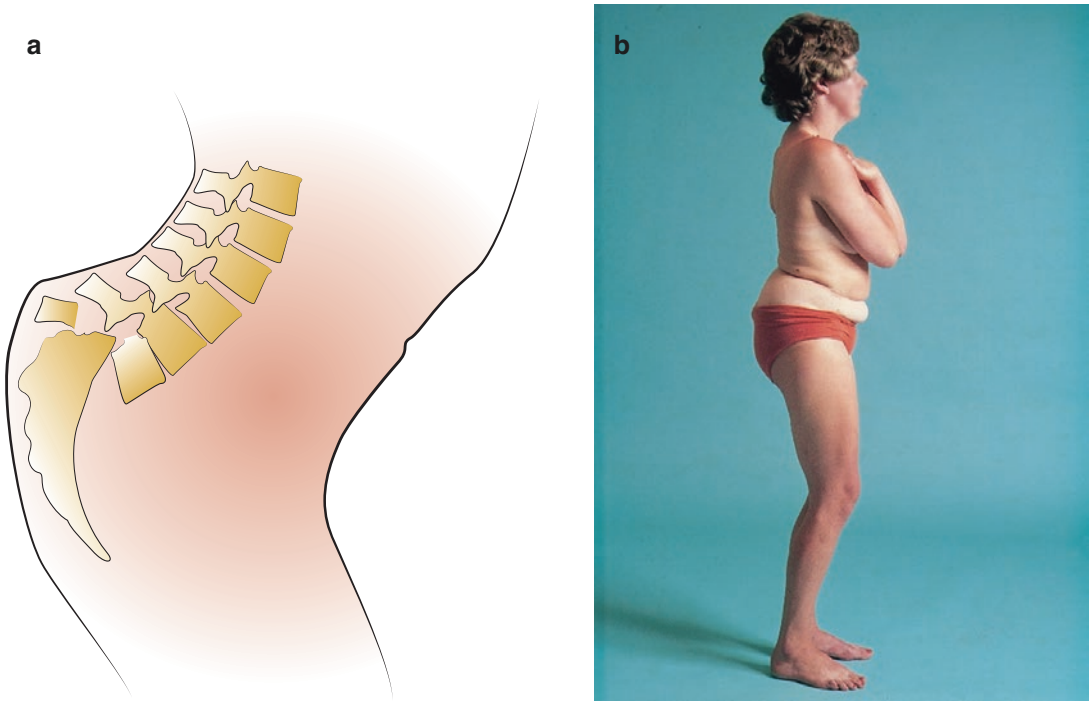



**Fig. 3.32** (**a**) A lumbosacral kyphosis occurs at the level of L5-S1 with a compensatory lordosis above it if the spinal column has subsided 50 % or more with respect to the anteroposterior distance from the vertebral body of S1. (**b**) The buttocks are flat and the knees bent. The back is relatively short, which causes a deep skin fold at the lumbar level

**Spondylolisthesis** A spondylolisthesis is an anterior displacement of the spinal column, mostly at the level of L5-S1 (spondylos is Greek for vertebra, olisthanein for sliding down). There is a lumbo-sacral kyphosis (at the L5-S1 level) with a compensatory lordosis if the spinal column has been displaced 50 % or more with respect to the anteroposterior length of the vertebral body at S1.

A compensatory lordosis does not disappear when sinking through the knees but does partially disappear during the forward bending test. The buttocks are flat and the knees are bent (Fig. 3.32). In 20–50 % of cases a spondylolisthesis is accompanied by a slight scoliosis. This condition may be painful (see pp. 45–48).

**Flexion contracture of the hip(s)** A compensatory hyperlordosis will occur (Fig. 3.33) in the case of a flexion contracture of one or both hips. Flexion contractures of the hips can be detected using the Thomas test (Fig. 3.34). In this case the child lies on his back with his or her legs flat on the underlying surface. The hyperlordosis will be visible. By flexing the hip or hips, the hyperlordosis will disappear. Next, by letting the leg with the hip flexion contracture drop down again the hyperlordosis appears at the moment the hip will not extend any further. The hyperlordosis also disappears during the sinking knees test and during the forward bending test. This happens in both cases because the hips are in flexion.

*Structural hyperlordosis* A structural hyperlordosis does not disappear during the forward bending test. This has a congenital or neuromuscular origin.

**Congenital hyperlordosis** This is the least common congenital spinal abnormality. It is caused because the vertebrae are not segmented on the posterior aspect (Fig. 3.35). A lordosis is caused by symmetrical bony connections in the intervertebral joints, but usually there is also a scoliosis due to asymmetry of the bony connections. The anomaly is always progressive.

**Fig. 3.33** Flexion contracture of both hips causes a compensatory hyperlordosis

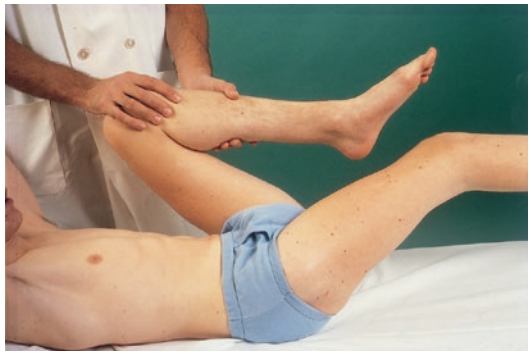


**Fig. 3.34** Thomas test. Flexion contracture of the right hip

Supplementary assessment: anteroposterior and lateral X-rays, a CT-scan and a MRI of the lumbar spine are made. An anteroposterior and a frog-leg lateral X-ray of the pelvis are made in the case of flexion contractures of the hips in order to rule out hip pathology.

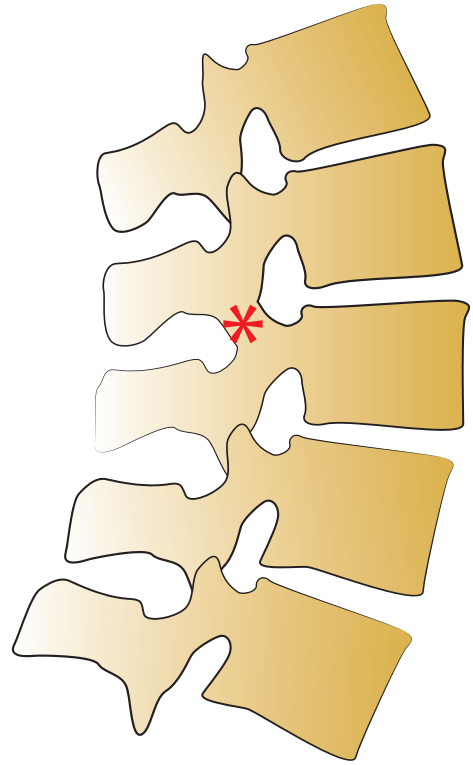


**Fig. 3.35** A congenital hyperlordosis is the least common congenital spinal abnormality. The cause is a symmetrical bony connection in the intervertebral joints (see*). Asymmetry of the bony connections often means that there is also a scoliosis. The anomaly is always progressive

Primary care treatment: the general practioner can prescribe pelvic tilt excercises in the case of a postural abnormality with an enhanced lumbar lordosis.

When to refer: a compensatory and a congenital hyperlordosis must be referred.

Secondary care treatment:**spondylolisthesis.** (see p. 50, 51).

**Flexion contracture of the hips** Treatment is dependent on the underlying pathology (see bilateral hip dislocation, Chap. 9).

**Congenital hyperlordosis** Treatment with a orthosis is not indicated. The treatment is operative in all cases.

An anterior spondylodesis can only be performed at an early stage. At a later stage a much

**Table 3.4** Neck and back abnormalities in different syndromes

| **Achondroplasia** | **Ehlers-Danlos syndrome** | **Osteogenesis imperfecta** |
|---|---|---|
| Stenosis of the foramen magnum | Scoliosis and hyperlordosis | Basilar impression |
| Lumbar hyperlordosis | **Marfan syndrome** | Scoliosis and hyperkyphosis |
| Spinal stenosis | Scoliosis and hyperkyphosis | **Spondyloepiphyseal dysplasia** |
| **Diastrophic dwarfism** | **Morquio disease** | Atlantoaxial subluxation |
| Scoliosis/hyperkyphosis | Aplasia/hypoplasia of the dens | Scoliosis and hyperkyphosis |
| Cervical spina bifida | Scoliosis and hyperkyphosis | |
| **Down syndrome (trisomy 21)** | **Neurofibromatosis (von Recklinghausen disease)** | |
| Occipitoatlantal subluxation | Scoliosis and hyperkyphosis | |
| Atlantoaxial subluxation | | |

See Appendix for characteristics of syndromes

### Differential Diagnosis: Back Misalignment

| **Lateral curvature(s) of the back** | **Scoliosis** |
|---|---|
| The scoliosis disappears during the forward bending test. | **Postural scoliosis** |
| Difference in leg length. | **Compensatory scoliosis** |
| The lateral curve does not disappear during the forward bending test. | **Congenital scoliosis** |
| Rib hump during forward bending. | **Idiopathic scoliosis** |
| Rib hump during the forward bending test, ages 0–3 years. | Infantile idiopathic scoliosis |
| Rib hump during the forward bending test, ages 3–10 years. | Juvenile idiopathic scoliosis |
| Rib hump during the forward bending test, from age 10 years. | Adolescent idiopathic scoliosis |
| **Abnormal rounded back** | **Hyperkyphosis** |
| The rounded back can be corrected actively. There is a normal back contour during the forward bending test. | **Postural hyperkyphosis** |
| There is an angular apical kyphosis in the back contour during the forward bending test, from birth. | **Congenital hyperkyphosis** |
| There is an angular apical kyphosis in the back contour during the forward bending test, ages 12–15 years. | **Scheuermann disease** |
| Generally familial. | **Idiopathic hyperkyphosis** |
| **Abnormal hollow back** | **Hyperlordosis** |
| The hyperlordosis disappears during the forward bending test and when sinking through the knees. | **Postural hyperlordosis** |
| The hyperlordosis disappears entirely or partially during the forward bending test. The hyperlordosis does not disappear when sinking through the knees. | **Spondylolisthesis** |
| There are flexion contractures of the hip. The hyperlordosis disappears during the forward bending test and when sinking through the knees. | **Flexion contractures of the hip(s)** |
| The hyperlordosis persists during the forward bending test and when sinking through the knees. | **Congenital hyperlordosis** |
| **Combined curves** | **Scoliosis and hyperkyphosis** |
| Lateral curve and hyperkyphosis. | Diastrophic dwarfism<br>Morquio disease<br>Neurofibromatosis<br>Osteogenesis imperfecta<br>Spondyloephiphyseal dysplasia<br>Scheuermann disease |
| Lateral curve and hyperlordosis. | **Scoliosis and hyperlordosis**<br>Marfan disease<br>Ehlers-Danlos syndrome |

more extensive operation is required. An anterior as well as a posterior spondylodesis must be performed to correct the hyperlordosis.

## Combined Curvatures

There is also a frequent scoliotic component next to the thoracic hyperkyphosis in skeletal dysplasia, such as spondyloepiphyseal dysplasia,[2] and a mucopolysaccharidosis such as in Morquio disease[2].

Scheuermann disease has a scoliotic component in 30 % of cases. Connective tissue diseases such as Marfan syndrome[2], osteogenesis imperfecta[2] and Ehlers-Danlos syndrome[2] frequently present with a scoliosis with a hyperlordotic component (Table 3.4). Curvatures that are part of more general anomalies are almost always progressive.

# Back Pain

## Back Pain

Complaint: the patient complains of back pain.

Assessment: the mobility of the thoracolumbar spine may be normal or limited. Sometimes a scoliosis or painful scoliosis may be present.

Differential diagnosis:

**spondylolysis**
**spondylolisthesis**
congenital or dysplastic
spondylolisthesis
isthmic extension spondylolisthesis
isthmic spondylolytic
spondylolisthesis
traumatic spondylolisthesis
pathological spondylolisthesis
degenerative spondylolisthesis

[2]See Appendix.

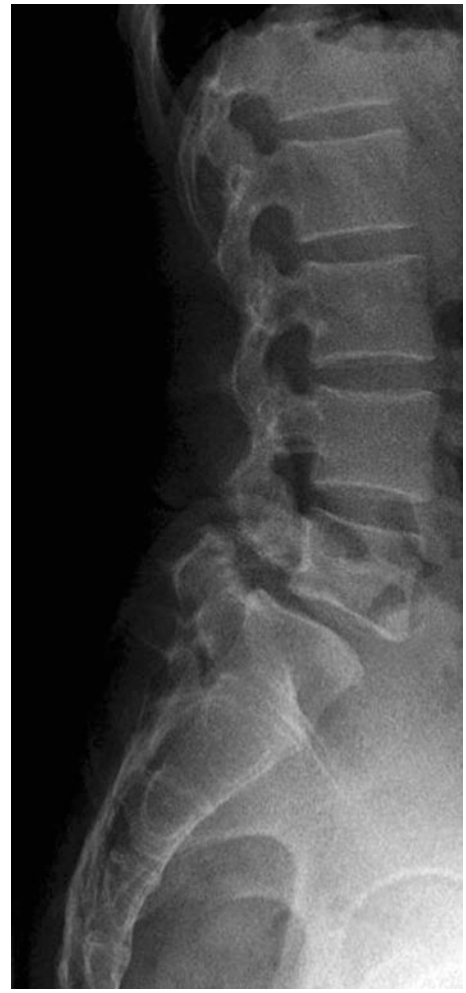


**Fig. 3.36** Spondylolysis of the arches of L5 with a grade-1 spondylolisthesis

**Scheuermann disease**
**disc calcification**
**spondylodiscitis (discitis)**
**tumor**
**aspecific back pain**

Explanatory note: **spondylolysis.** Spondylolysis (spondylos is Greek for vertebra and lysis for release) is a discontinuity of the pars interarticularis of the spinal arch (Fig. 3.36). The pars interarticularis is the connection between the upper and the lower articular processes. This anomaly does not appear

before the age of 18 months. Prevalence at the age 6 is 4.5 % and amongst young adults it is 6 %. Prevalence does not increase in adulthood. The anomaly is more frequent in people who do certain sports, such as gymnastics. The most common cause of spondylolysis is probably a stress fracture of the pars interarticularis of the spinal arch (also called isthmus) in a non-optimally developed vertebral arch (dysplastic). The latter is probably hereditarily determined. This could also explain the 50 % spondylolysis rate among some Eskimo tribes. 85 % of spondylolyses are localized in the arch of L5 and at this level it is usually bilateral. The other cases are localized at the levels L3 and L4, and are usually unilateral. Spondylolysis does not cause back problems in most of the cases. In the period of development of a spondylolysis 15 % of cases have back problems. It occurs twice as often in boys than girls, although girls tend to have more symptoms. There is a painful scoliosis as an avoidence reaction to pain in 15 % of cases. On a bone scan this shows increased activity at the level of the spondylolysis. Overloading the back when there is a spondylolysis can also cause back problems. These problems are probably the consequence of stretching of the fibrous tissue that has formed at the level of the stress fracture in the arch.

**Spondylolisthesis** Spondylolisthesis (olisthanein is Greek for sliding down) involves the anterior displacement of a vertebral body including the entire cranial part of the spinal column with respect to the vertebra below. This generally involves level L5-S1. The position of the spinous process in the vertebral body that has been displaced anteriorly remains unchanged with respect to the underlying vertebral body. The spinous process of the vertebral body above it has been displaced anteriorly with respect to the rest of the spinal column so that a clinically palpable step appears at that level. Hence in a spondylolisthesis at the L5-S1 level and a step is palpable at the L4-L5 level (Fig. 3.37). The prevalence of this anomaly is 2–4 %. The degree of spondylolisthesis is graded 1–5 (Fig. 3.38).

A total anterior displacement is called spondyloptosis (Fig. 3.39). There is scoliosis in 20–50 %

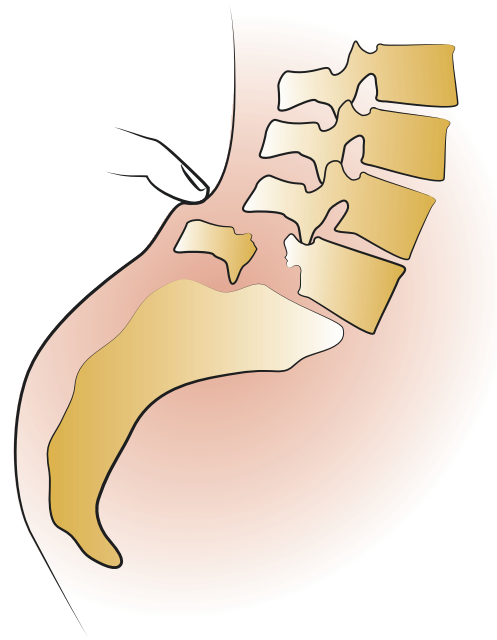


**Fig. 3.37** A step at the L4-L5 level is palpable in a spondylolisthesis at the L5-S1 level

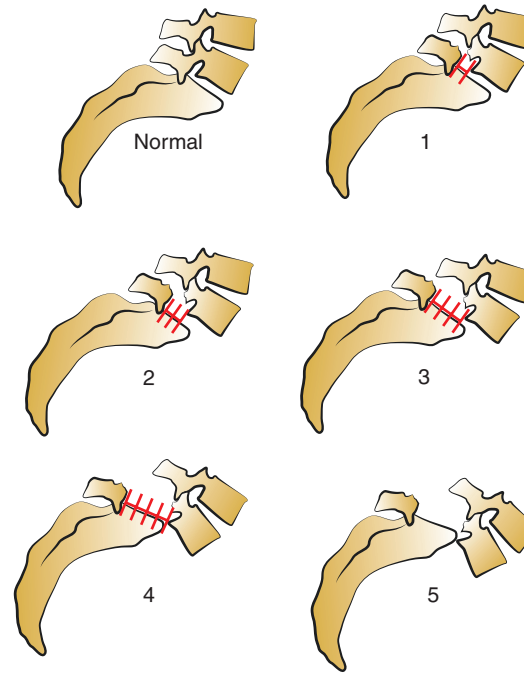



**Fig. 3.38** Meyerding classification. The degree of spondylolisthesis is indicated by a gradation from 1 to 5. *Grade 1*: displacement of 0–25 %; *grade 2*: 25–50 %; *grade 3*: 50–75 %; *grade 4*: 75–100 %; *grade 5*: total displacement (spondyloptosis) (Redrawn from: Meyerding H. Spondylolisthesis. Surg Gynecol Obstet. 1932;54:371–7)

of cases in a spondylolisthesis. If kyphosis develops at the level of the lumbosacral junction with an enhanced compensatory lumbar lordosis, with flat buttocks resulting from a backward-tilting pelvis and bent knees, then we are dealing with a spondylolisthesis that has an anterior displacement of more than 50%. In some cases a spondylolisthesis will remain symptom-free. Lower back problems may occur, especially after contact sports. The complaints manifest themselves mostly during puberty. Sometimes there radiating pain to the posterior side of one or both legs. There are six types of spondylolisthesis:

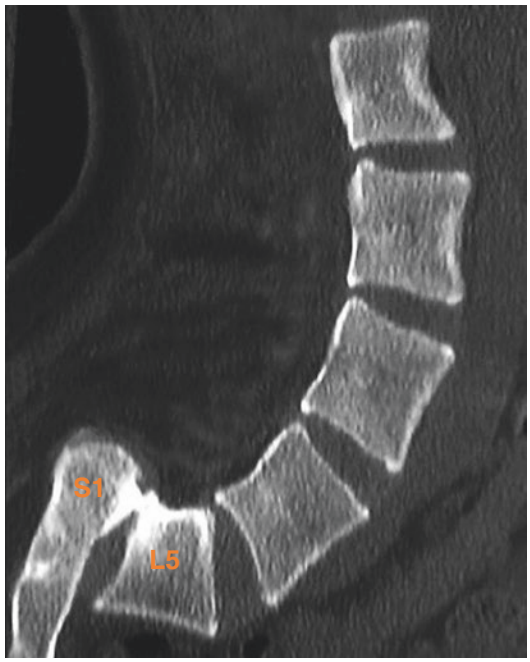



**Fig. 3.39** A spondylolisthesis grade 5 (spondyloptosis) at the L5-S1 level with an enhanced compensatory lumbar lordosis

*Congenital or dysplastic spondylolisthesis* This type is extremely rare and only occurs at the L5-S1 level. The joint protuberances are hardly developed. The joint protuberances of L5 no longer remain hooked behind those of S1, so that L5 is displaced anteriorly along with the rest of the upper spinal column with respect to S1. The part of the arch of L5 between the joint protuberances (pars interarticularis) is stretched and thin on both the right and the left side and there is potentially a risk of a secondary fracture here (Fig. 3.40). The degree of anterior displacement is usually quite severe. Neurological symptoms are not rare because the posterior side of the arch (lamina) of L5 is displaced towards the dural sac. Compression of the cauda equina can already occur with a displacement of 30%.

*Isthmic extension spondylolisthesis* This abnormality is based on the stretching of the pars interarticularis from the L5 arch (Fig. 3.40). This process occurs during childhood. The stretching and the ensuing anterior displacement can increase gradually. Full anterior displacement is even possible. Extension of the pars interarticularis occurs

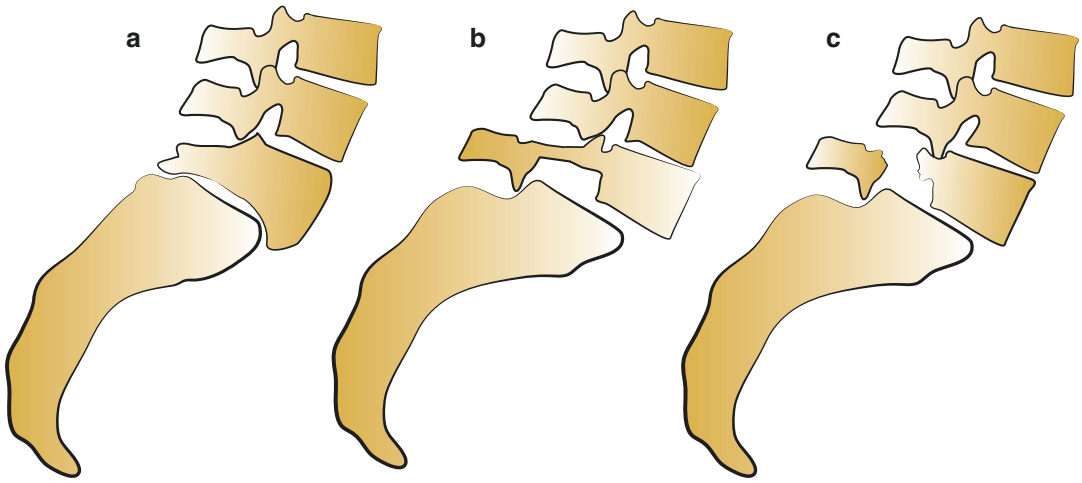



**Fig. 3.40** Spondylolisthesis. (**a**) Congenital or dysplastic spondylolisthesis. (**b**) Isthmic extension spondylolisthesis. (**c**) Isthmic spondylolytic spondylolisthesis

because of repeated stress fractures in this section, causing a small forward displacement of the vertebral body with respect to the posterior part of the spinal arch (lamina), after which the fracture heals. This type is also infamous because of neurological complications. The anomaly continues to develop further after 20 years of age.

*Isthmic spondylolytic spondylolisthesis* This is the most common type. The anterior displacement is a consequence of a bilateral spondylolysis (Fig. 3.40). This process occurs during childhood and can increase gradually but stops progressing after the age of 20. It appears most often at the L5-S1 level. In some cases the root of S1 is compressed. Compression of the S1 spinal root causes radiating pain. This is caused by hypertrophy of the fibrous tissue at the level of the spondylolysis or as a result of a disc herniation at the level of the displacement.

*Traumatic spondylolisthesis* Great force is required in order to cause an acute fracture in the pars interarticularis and this is always accompanied by other fractures of the same vertebra. An acute isolated pars interarticularis fracture has never been seen.

*Pathological spondylolisthesis* This is very rare and is caused by a pathological process in bone as in osteogenesis imperfecta, an infection or a tumor in which destruction of the arch or intervertebral joints has occurred after which an anterior displacement occurs.

*Degenerative spondylolisthesis* This type is based on degeneration of the intervertebral disc in combination with arthritic changes in the intervertebral joints. This can cause abnormal mobility at the level of the intervertebral joints, resulting in a forward displacement of the vertebra including the upper spinal column (anterolisthesis) or a posterior displacement (retrolisthesis). A degenerative spondylolisthesis is located mostly at the level of L4-L5. It is three times more common in women than in men, and is almost never seen in individuals under the age of 40. In a large number of cases the root of L5 is compressed, which causes radiating pain in the lower extremities.

The isthmic variant accounts for 20 % of cases, the spondylolytic for 50 %, the degenerative for more than 25 %, and the others less than 5 %. Children of course do not get degenerative spondylolisthesis, it only affects adults. The most common anterior displacement in isthmic and spondylolitic spondylolisthesis in children ceases developing after the age of 20. If there were no back problems before 20 years of age it is improbable that this will play a role in the development of back problems at an older age.

**Scheuermann disease** In this disease there is an enhanced non-redressable thoracic kyphosis (hyperkyphosis) (see pp. 35–41). In general, patients go to the doctor because of an abnormally rounded back. The patient sometimes complains of back pain, mostly around the apex of the curvature located at the level of T9. The pain usually disappears when adulthood is reached. Only in a limited number of cases does the pain persist during adulthood.

**Disc calcification** Disc calcification occurs in 70 % of cases at the cervical level, in 20 % at the thoracic level and in 10 % at both the cervical and thoracic levels. It is rare in children at the lumbar level. In a thoracic localization there is generally pain and reduced mobility of the thoracic spine. There may be a subfebrile temperature, and the WBC count as well as the CRP and ESR can also be elevated. X-rays will not show any abnormality at first, but after the symptoms begin the calcification becomes visible on X-ray images within two weeks. The abnormality recovers spontaneously.

**Spondylodiscitis** An inflammation of the vertebra is called spondylolytis, an inflammation of an intervertebral disc discitis and that of an intervertebral disc with adjoining vertebral bodies spondylodiscitis. The infection probably occurs in one of the endplates of a vertebral body, and the intervertebral disc is secondarily infected. This problem occurs at all ages, but especially in

the first 3 years of life. Manifestation at the lumbar level occurs three times more often than at the thoracic level. Spondylodiscitis in children is usually caused by a Staphylococcus aureus. The cause of the infection is not clear. The onset of the disease can be acute or chronic and therefore vague. The child may complain about leg pain and is sometimes unable to walk or will complain about back pain and in some cases even about having abdominal pain. The young patient refuses to bend his or her back. The Lasègue test is often positive. Especially in young children there may be no fever; in more than half of the cases the CRP is not abnormal, and the WBC count is also mostly abnormal. The ESR is usually elevated. Blood cultures are positive in 30 % of cases. A biopsy of the intervertebral space is generally not necessary and is only performed if there are diagnostic problems or if the therapy is not effective. A biopsy is positive in only 25 % of cases. Initial radiological assessment shows no abnormalities. After 10–14 days a narrowing of the intervertebral space is observed on X-rays, as well as irregular endplates of the adjoining vertebral bodies (Fig. 3.41). MRI shows an abnormal intervertebral disc at an early stage, even before anomalies can be seen on the bone scan. The intervertebral disc is often destroyed by the infection, and disappears at a later stage. This causes a fusion (block vertebra formation) in the adjoining vertebral bodies in 20 % of cases. The block vertebra formation causes few or no symptoms.

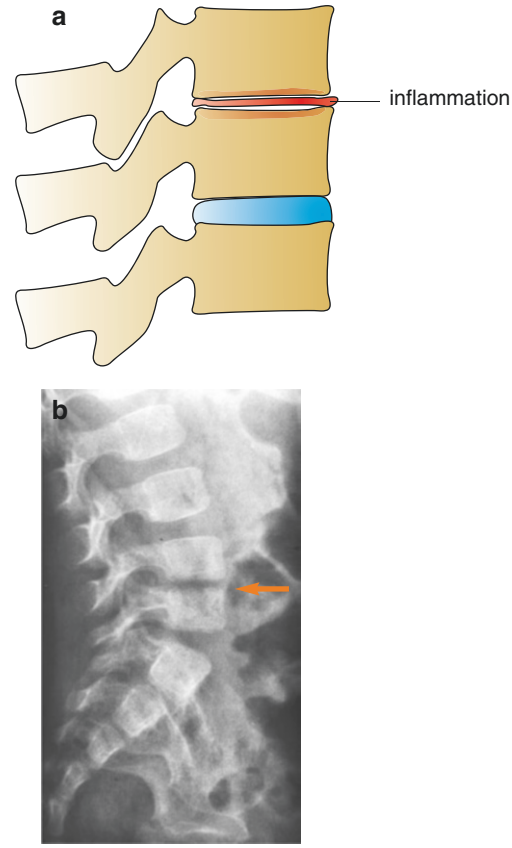



**Fig. 3.41** (**a**) Inflammation of the intervertebral disc (discitis), reducing the space between the vertebral bodies. (**b**) Lateral X-ray of the lumbar spine and spondylodiscitis at the L4-L5 level (*arrow*)

**Tumor** Tumors of the back are rare in children. A thoracic localization is the most frequent. A subclassification can be made into metastases (Table 2.2), primary bone tumors (Table 2.3) and spinal cord tumors (Table 2.4).

Patients complain of localized pain, often during the night. The pain does not decrease with activities or with rest, which is the case with other back abnormalities. There is usually a painful scoliosis. The tumor is located on the concave side of the apex of the scoliosis.

About half of spinal cord tumors occur in the first 4 years of life. As mentioned previously the pain is often vague, mostly constant, and does not disappear with rest. Physical assessment can produce local pressure pain. Neurological symptoms only occur later (Table 2.5) (see p. 17).

**Aspecific back pain** Back pain in children is not that rare. In a great number of cases no specific cause is identifiable; this is called aspecific back pain. In back pain lasting longer than 3 months we speak of a chronic aspecific back pain. Thus, this is a diagnosis per exclusionem. There is often a trivial trauma in the medical history of aspecific back problems that would not give rise to a long period of back pain.

In adults there is often a somatized low back pain due to stress or possible financial

gains from being ill. This type of problem is increasingly seen in children in our achievement-orientated society. It is especially seen in girls at the age of puberty. The parents or a close relative also often have back pain. Physical assessment shows normal mobility of the thoracolumbar spine, and movement assessment of the back generally shows no evidence of increased pain. Radiological assessment shows no abnormalities.

Supplementary assessment: in order to rule out a specific underlying cause, radiological assessment of the thoracic and lumbar spine is performed in children with back pain that has lasted more than 3 months. A fault that is often made is that only anteroposterior and lateral X-rays of the lumbar spine have been requested. One should realize that in 20 % of cases a spondylolysis is not visible on anteroposterior and/or lateral X-rays of the spinal column. However on oblique X-rays of the lumbar spine a spondylolysis may be displayed. A spondylolisthesis is visible on lateral X-rays of the lumbar spine.

A diagnosis of Scheuermann disease can be established on the basis of irregular apophyses or wedge-forming on the anterior side of the thoracic or thoracolumbar vertebral bodies as seen on lateral X-rays of the thoracic and lumbar spine. Disc calcifications are also clearly visible on lateral X-rays of the thoracic spine. If there is fever, the CRP, the ESR and WBC count should also be determined to rule out spondylodiscitis. However: CRP and WBC counts may often be normal.

X-rays show no abnormalities in the first 10 days. If conventional radiological assessment of the thoracic and lumbar spine and laboratory assessment show no anomalies and one does suspect a problem, a bone scan and an MRI are indicated. An osteoid osteoma may especially be missed on conventional X-ray images because of its small size. A bone scan will show a hotspot. A spondylodiscitis and an intraspinal tumor can be seen on a MRI.

Primary care treatment: there are no symptoms in most cases of a spondylolysis. If a spondylolysis does give rise to complaints the symptoms may be present for several months. Children with an asymptomatic spondylolisthesis do not need treatment if the displacement is less than 25 %. In order to prevent back pain contact sports and back-loading activities are not recommended if the displacement is more than 25 %. In Scheuermann disease children sometimes complain of backpain, this usually disappears in adulthood.

Disc calcification recovers spontaneously. The initial symptoms disappear after 7–10 days. Analgesics should be given during the symptomatic period.

Children with aspecific low back pain must remain as active as possible and avoid bed rest. Stimulate the children to move as much as possible and expand their activities despite the pain. Pain medication, exercise therapy and manual therapy are generally not necessary. Traction, TENS (Transcutaneous Electrical Nerve Stimulation) treatment, massage and a lumbar orthosis are not necessary. There should be a follow-up assessment to rule out a specific cause in cases where the symptoms last longer than 3 months.

When to refer: children with a painful scoliosis, a spondylolisthesis, a hyperkyphosis and/or limited flexion of the thoracolumbar spine and/or fever with abnormal laboratory results that indicate an inflammation, should be sent in for orthopedic assessment. This is even the case if the X-rays of the spinal column requested by the general practitioner show no abnormalities.

Secondary care treatment: **spondylolysis.** A trunk orthosis can be prescribed to immobilize the back in cases where the low back problems are present for more than 3 months. Children and young adults react well to this treatment and in general the treatment can be stopped after 6 months.

**Spondylolisthesis** In case of a prolonged painful displacement of less than 25 % orthosis treatment

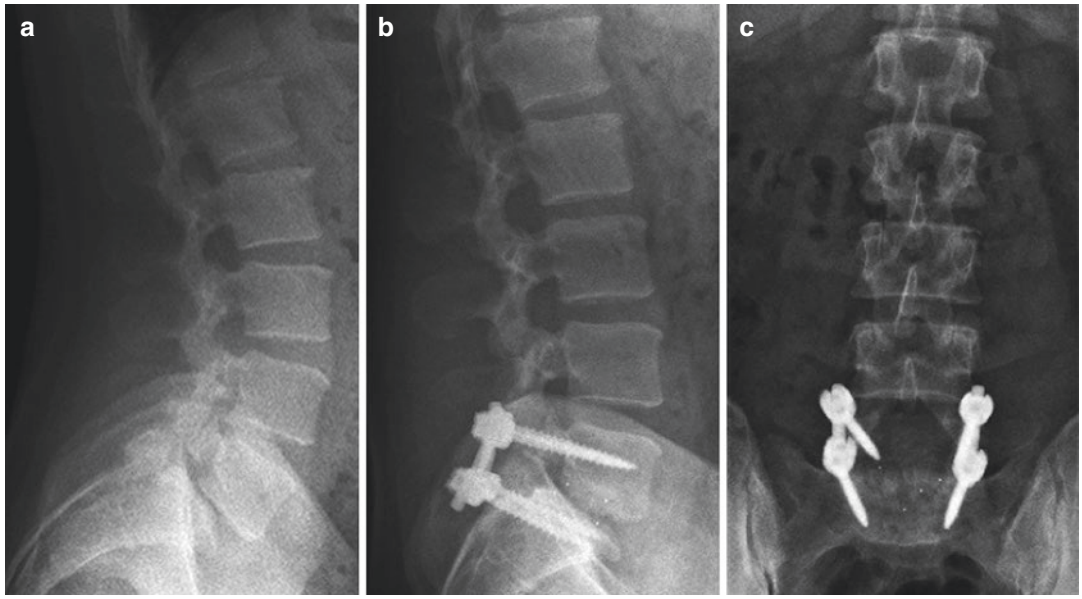



**Fig. 3.42** (**a**) Spondylolisthesis L5-S1 grade 1–2. (**b**, **c**) posterior lumbar interbody fusion L5-S1 (PLIF). The intervertebral disc is removed, the anterior displacement (spondylolisthesis) at the level of the intervertebral space is corrected as much as possible. The intervertebral space is filled with a carbon fiber cage containing bone transplants obtained from the iliac crest. Next, the vertebrae are fixed with screws on both sides and these are connected with rods

may be prescribed to control symptoms. None operative management or "watchful waiting" of the minimally symptomatic or asymptomatic child in a high grade spondylolisthesis is safe and does not lead to significant problems. In cases with more than minimal back pain with a displacement of more than 25 % there is an indication for spondylodesis of the displaced vertebra with the vertebra located distally (Fig. 3.42). In the case of neurological symptoms, decompression must take place at the same time.

**Scheuermann disease** (see p. 40, 41).

**Spondylodiscitis** Immobilization of the back using a lumbar orthosis and antibiotic therapy for 2 weeks intravenously and afterwards oral antibiotics until the symptoms disappear and the parameters are normalized. This usually takes 6–8 weeks.

**Tumor** Treatment of benign as well as malignant tumors should take place in a specialized medical center. Treatment of tumors is usually surgical, if necessary supplemented with radiotherapy and/or cytostatic drugs, depending on the nature of the tumor.

## Back Pain Accompanied by Noctural Sweating

- Complaint: a painful back at the level of the thoracolumbar spine. Mobility in the thoracolumbar spine is limited. The patient is sick, has subfebrile temperature, has nocturnal sweating and is losing weight. Pulmonary, intestinal or renal tuberculosis in the medical history may be known.
- Assessment: the affected vertebrae are pressure-sensitive. There is a considerable movement limitation of the back.
- Diagnosis: **tuberculous spondylitis**
- Explanatory note: Spinal tuberculosis is very rare in developed countries.

Tuberculous spondylitis should be considered when the following symptoms are present:

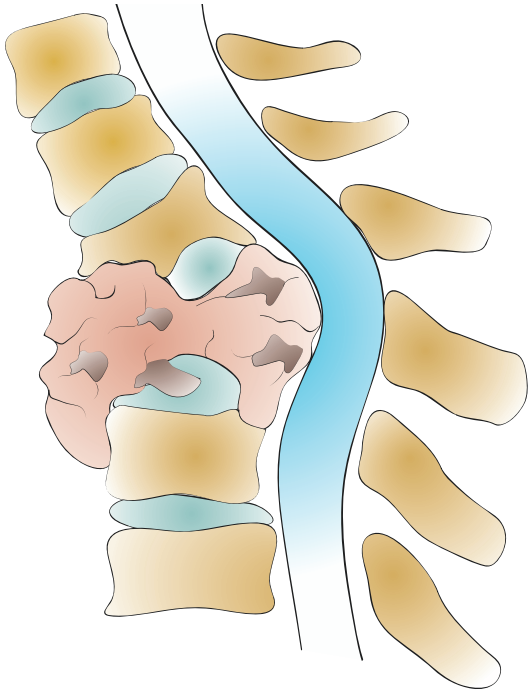


**Fig. 3.43** Tuberculous spondylitis. It begins in the vertebral body. Adjoining vertebral bodies can be affected through the intervertebral discs (Redrawn from: Johnston JO. A syllabus on orthopedic infections, dysplasias and joint disease. Oakland: Department of Orthopedics, Kaiser Foundation Hospital; 1984)

- The medical history reveals a pulmonary, intestinal or renal tuberculosis. The tuberculous infection of the vertebrae occurs due to hematogenic transfer of the tuberculum bacilli from the primary source.
- A tuberculous infection of the vertebral body may occur about 3 years after the primary infection.
- The patient is sick, has a subfebrile temperature, complains of nocturnal sweating, and there is weight loss.
- The affected vertebrae are pressure-sensitive.

In contrast to spondylodiscitis, tuberculous spondylitis begins in the vertebral body and can subsequently affect another vertebral body via the intervertebral discs (Fig. 3.43). The infection is mainly located in the low thoracic spine or the high lumbar spine, which may possibly cause an abscess on the anterior side of the spine, which via the psoas muscle may be displaced into the insertion of this muscle on the femoral trochanter minor. This is called a cold abscess because general symptoms of infection tend to be absent. X-rays reveal a destruction of the anterior side of the vertebral bodies, with displacement of one or more vertebral bodies on the anterior side and expansion into the psoas shadow. Anteriorly an angular kyphosis may appear (Pott gibbus) due to the displacement of the vertebral bodies. Paraplegia can occur as a complication (Pott paraplegia). This paraplegia can be caused by spinal cord pressure due to pus, pressure of a displaced vertebrae on the spinal cord, or destruction of the local blood vessels.

Supplementary assessment: X-rays and MRI of the thoracolumbar spine, CRP, ESR and a Mantoux test.

Primary care treatment: none.

When to refer: as quickly as possible.

Secondary care treatment: a biopsy and a bacterial culture will generally be taken to confirm the diagnosis. However the results can take several weeks. Treatment can be started even before the results are known in cases with an obvious suspicion of tuberculosis. Treatment consists of a combination try-out of different tuberculosis medications. These drugs are generally administered for 1 year, and in severe cases for 18 months. In cases when there is compression of the spinal canal prompt intervention is required. In a kyphosis a spondylodesis should be performed after excision of the infected focus.

## Back Pain with Neurological Symptoms

Complaint: in addition to back pain there is severe limitation of movement in the thoracolumbar spine and there is pain and/or paresis in one or both legs.

Assessment: there are clear neurological abnormalities: radicular syndrome, monoparesis or paraparesis, monoplegia or paraplegia.

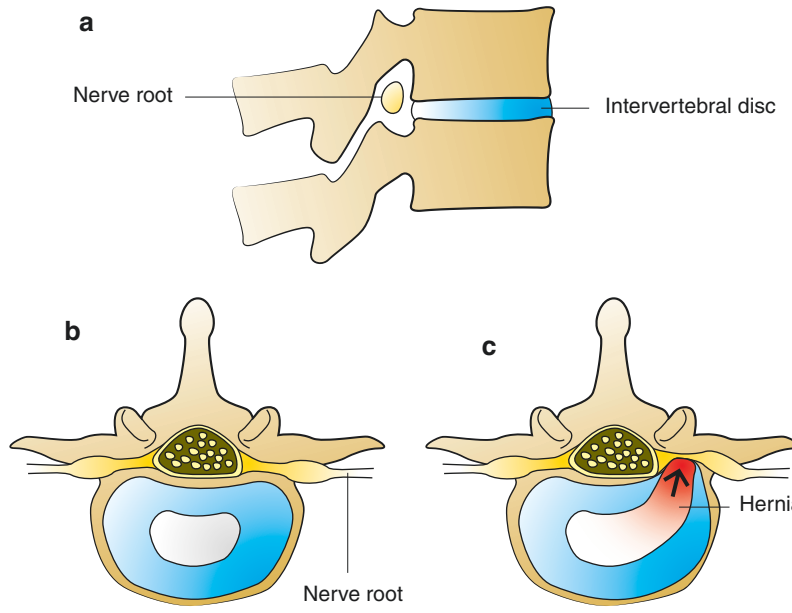



**Fig. 3.44** (**a**) Lateral view of the intervertebral disc between two vertebral bodies. The nerve root runs posterior to the inter-vertebral disc. (**b**) Intervertebral disc seen from below, the nerve root is not compressed. (**c**) Protrusion of the inner part of the intervertebral disc. The protrusion puts pressure on the nerve root (disc herniation)

Differential diagnosis:

**disc herniation**
**tumor**

Explanatory note: **disc herniation.** In a disc herniation there is a rupture of the fibers of the annulus fibrosus with subsequent shifting of the central core of the disc (nucleus pulposus) and the annular fibers, mostly posterolaterally or posteromedially (Fig. 3.44). If the nucleus shifts posteriorly but remains within the boundaries of the annulus fibrosus, one speaks of a protrusion. If the central core breaks through the fibers of the annulus fibrosus, then there is an extrusion or even a sequestration when the material from the central core ends up in the spinal canal (Fig. 3.45). A disc herniation in children is relatively rare. Only 2% of all hernias occur in children; by far most appear during puberty, and never in children younger than 7 years of age. About two-thirds of children have only back pain as the main symptom. One-third also have pain radiating into the leg. The abnormality occurs at the same levels as in adults, in 90–98% of cases at the L4-L5 or L5-S1 level. In nearly all cases there is a scoliosis as an avoidance reaction to the pain (painful scoliosis). Flexion of the thoracolumbar spine is severely limited in most cases (Fig. 3.46). Flexion of the hip with an extended knee while lying in a supine position is limited (Lasègue test) (Fig. 3.47). In cases of disc herniation, radicular symptoms (radiating pain into one or both legs) and loss of normal reflexes as well as loss of strength may be lacking during a prolonged period. Thus, the clinical picture is different than in adults.

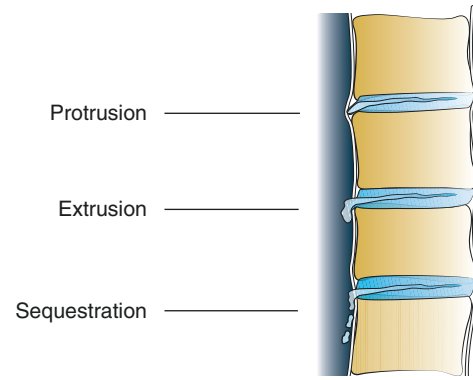



**Fig. 3.45** There is a protrusion if the nucleus is shifted dorsally but remains within the boundaries of the annulus fibrosus. There is an extrusion, and with several fragments a sequestration if the central core breaks through the fibers of the annulus fibrosus

*Disc herniation level L3-L4* A protrusion of the disc at L3-L4 level can put pressure on the spinal nerve root of L4 leading to pain radiating down

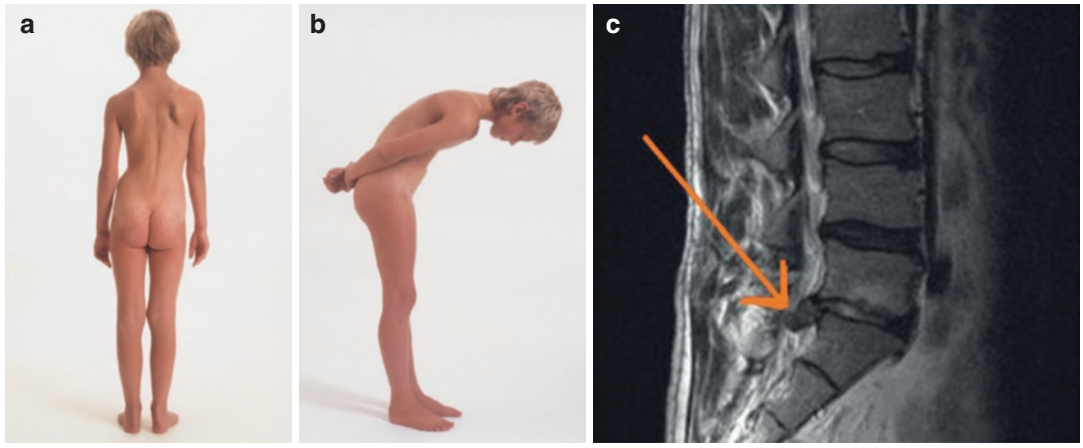



**Fig. 3.46** (**a**) Nine-year-old girl with painful scoliosis due to a hernia nucleus pulposus and (**b**) severely limited flexion of the thora-columbar spine. (**c**) MRI: disc herniation L5-S1

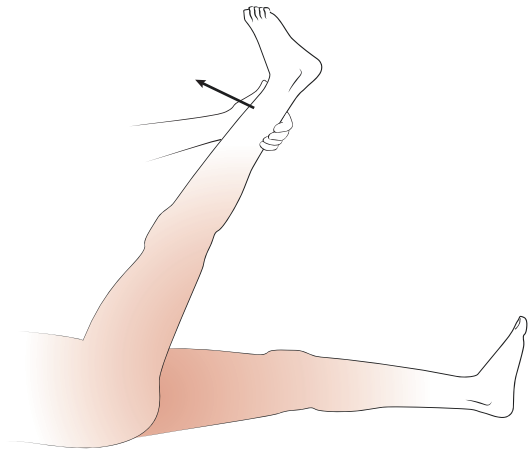


**Fig. 3.47** Lasègue test. Flexion of the hip with an extended knee lying supine is severely limited in a disc herniation

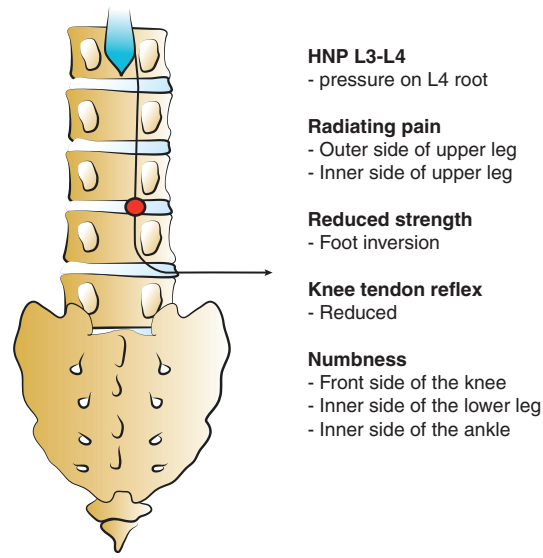



**Fig. 3.48** Disc herniation at the L3-L4 with pressure on the L4 root

the lateral side of the thigh and the medial side of the lower leg and ankle with a numbness on the anterior aspect of the knee and the medial side of the lower leg and the ankle, with reduced inversion strength of the foot and a reduced patella tendon reflex (Fig. 3.48).

*Disc herniation L4-L5* In this case the spinal nerve root of L5 may be compressed, causing radiating pain along the posterior side of the thigh, the lateral side of the lower leg and the medial side of the foot. This may be accompanied by a numb feeling on the lateral side of the lower leg and on the medial side of the foot, with reduced dorsal extension strength of the toes. There is no available reflex for the spinal nerve root of L5 (Fig. 3.49).

*Disc herniation L5-S1* In this case the spinal nerve root of S1 is compressed by the prolapsed disc, which leads to radiating pain to the back of the thigh, lower leg and the lateral side of the foot and ankle. There is a numb feeling on the lateral side of the foot and ankle. Eversion and plantar flexion strength of the foot may be reduced and the Achilles tendon reflex may be reduced or absent (Fig. 3.50).

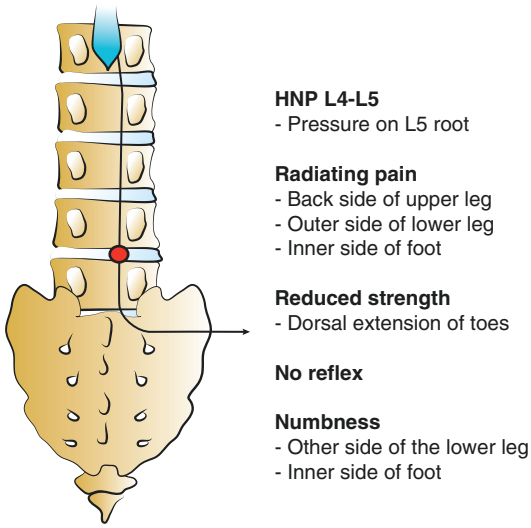



**Fig. 3.49** Disc herniation at the L4-L5 level with compression of the L5 root

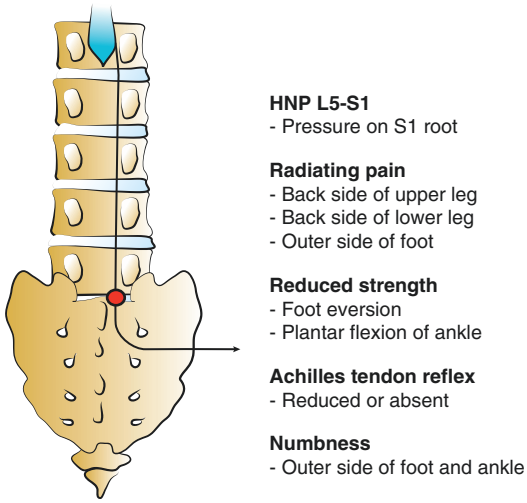



**Fig. 3.50** Disc herniation L5-S1 with compression of the S1 root

The classical findings at different levels such as those described above are more the exception than the rule. Reduced sensation is only present in 50 % of cases. There is a paresis in only 5–10 % of cases, and an absent Achilles or patella tendon reflex in 40–50 %. There is usually only a slight paresis because most muscles are innervated by two or more nerve roots and the disc herniation tends to only compress one root. Only with a sudden rupture and a large hernia will more roots be compressed, causing for instance a drop foot (L4 and L5) and paresis of the calf (S1 and S2). In the first case the patient cannot stand on his heel and in the second case he cannot walk on the toes of the affected foot. Such sudden motor deterioration is sometimes preceded by the disappearance or a change in the ischias, which indicates a displacement of a loose part of the disc. A painful scoliosis occurs in 65 % of cases and reduced flexion of the thoracolumbar spine in 96 %. A positive Lasègue test is found in 90 % of cases of disc herniation and a crossed Lasègue test in 30 %. Although a crossed Lasègue test is not very sensitive, it is more specific than the normal Lasègue test. The level of the hernia cannot be determined on physical assessment. Absence of the Achilles tendon reflex occurs in one third of hernias proximal to the L5-S1 level. Although the absent patella tendon reflex is six to seven times more common in hernias at the L3-L4 level, only a quarter of patients with absent patella tendon reflex have a hernia at that level.

Spasm of the lower lumbar paravertebral musculature is often present. This muscular spasm can cause a scoliosis. Limited lateral flexion of the lumbar spine located on the side the hernia is most noticeable with a bulging of the disc lateral to the spinal nerve, whereas limitation in lateral flexion towards the contra-lateral side is most noticeable with a bulging medial to the spinal nerve root (Kemp test).

A diagnosis of disc herniation can be established using clinical assessment. It is important to do flexion assessment of the lumbar spine, the Lasègue test and the crossed Lasègue test. The level of the hernia cannot be determined on physical examiniation.

**Tumor** In addition to histological classification metastasis (Table 2.2), primary bone tumor (Table 2.3), primary spinal cord tumor (Table 2.4), another classification can be made according to localization: extramedullary and intramedullary tumors. Intramedullary tumors can be subdivided into extradural and intradural tumors.

Bone tumors are extramedullary tumors of the spinal column. Pain and limitation of movement are most obvious. Neurological symptoms may

appear only after several months. This usually involves a radicular syndrome (Table 2.5).

In extradural tumors, especially neuroblastomas, radicular pain can for a long time precede paresis or paralysis. Intradural tumors, especially astrocytomas, develop slowly over many years. The symptoms are caused primarily by infiltrations, secondarily by pressure on the surrounding intact tissue.

Supplementary assessment: X-rays, CT-scans and MRIs of the thoracic and lumbar spine.

Primary care treatment: none.

When to refer: refer as quickly as possible in the case of deteriorating symptoms.

Secondary care treatment: **disc herniation.** Initially, 6 weeks prescribed rest, if necessary supported by analgesics and an orthosis. The symptoms disappear in 90% of cases. A discectomy can be considered if there is no improvement. This gives good results in 90% of cases. A paresis is not an indication for surgery.

**Tumor** Treatment of benign as well as malignant intraspinal tumors must take place in a specialized medical center. Treatment of spinal tumors is usually surgical, if necessary supplemented with radiotherapy and/or cytostatic drugs, depending on the nature of the tumor.

**Differential Diagnosis: Back Pain**

| **Back pain** | |
|---|---|
| Radiological assessment: oblique X-rays of lumbar spine images show an absent pars interarticularis. | **Spondylolysis** |
| Radiological assessment: anterior displacement of L4 with respect to L5. | **Spondylolisthesis** |
| The joint protuberances are hardly developed. | Congenital or dysplastic spondylolisthesis |
| Radiological assessment: anterior displacement of L5 with respect to S1, stretched pars interarticularis. | Isthmic extension spondylolisthesis |
| Radiological assessment: anterior displacement of L5 with respect to S1, spondylolysis of the arches of L5. | Isthmic spondylolytic spondylolisthesis |
| Acute fracture of the pars interarticularis, always accompanied by other fractures of the same vertebrae. | Traumatic spondylolisthesis |
| Radiological assessment: pathological process. | Pathological spondylolisthesis |
| Degenerative changes of the vertebrae joints, it only affects adults. | Degenerative spondylolisthesis |
| Radiological assessment: irregular end plates on the anterior side of the vertebral bodies. At a later stage, wedge-formation of the vertebral bodies. | **Scheuermann disease** |
| Radiological assessment: calcification at the level of the intervertebral disc. | **Disc calcification** |
| Radiological assessment initially not abnormal, but abnormal MRI. | |
| ESR usually elevated. | **Spondylodiscitis (discitis)** |
| Severe movement limitation. Painful scoliosis. MRI abnormal. | **Tumor** |
| Normal mobility of the back. Radiological assessment not abnormal. | **Aspecific back pain** |
| **Back pain accompanied by nocturnal sweating** | |
| Medical history: pulmonary or renal tuberculosis. | **Tuberculous spondylitis** |
| **Back pain with neurological symptoms** | |
| Radicular syndrome. | **Disc herniation** |
| Radicular syndrome, monoparesis, paraparesis, monoplegia or paraplegia. | **Tumor** |

# Pelvis

4

## Pelvic Pain

Complaint: the patient complains about pain at the pelvic level.

Assessment: if there is an anomaly at the level of one or both sacroiliac joints, then local pressure pain can be induced and Patrick test (Faber test) is positive (Fig. 4.1).

Differential diagnosis:
**ankylosing spondylitis (Bechterew disease)**
**septic sacroiliitis**
**tumor**

Explanatory note: **ankylosing spondylitis.** This is a disease with inflammatory symptoms of the joints and in which the sacroiliac and intervertebral joints become involved before the other joints. The disease begins between the ages of 4 and 16. Patients may complain about the heels, the back, and pain at the level of the sacroiliac joints. Ankylosing spondylitis is characterized by the following symptoms:

- Heel symptoms, low back symptoms or pain at the level of the sacroiliac joints that last longer than 3 months and does not clearly improve
- stiffness of the thoracolumbar spine, especially in the mornings
- thoracic excursions of less than 5 cm
- iritis or iridocyclitis
- elevated CRP and ESR
- radiological erosions and narrowing of the sacroiliac joints
- fast and good reaction to NSAID's
- in 90% of cases there is an elevated HLA-B27
- the rheumatoid factors are generally negative.

If one of the first four symptoms occurs in combination with a radiologically demonstrable bilateral sacroiliitis, this is sufficient for establishing a diagnosis of ankylosing spondylitis. About 0.1% of the population older than 15 has ankylosing spondylitis. The disease is 45 times more common in boys than girls, and tends to become manifest at the end of puberty and early adulthood. The first symptoms are limited lateral mobility of the lumbar spine. The sacroiliac joints, which have already been affected at a very early stage, are painful on pressure. This disappears at a later stage once the joints are ankylosed.

The cervical spine is generally affected at a later stage. The hip joints are also affected in addition to the sacroiliac joints and the intervertebral joints. In 20% of cases the more peripheral joints are also involved in the process. The limited respiratory excursion is caused by inflammation of the costovertebral joints. In 40% of cases ocular anomalies occur. In the initial stage ESR levels are not or only minimally elevated, but as the disease process progresses they quickly rise to relatively high levels. In ankylosing spondylitis rheumatoid factors are not positive more often than in healthy persons.


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_4

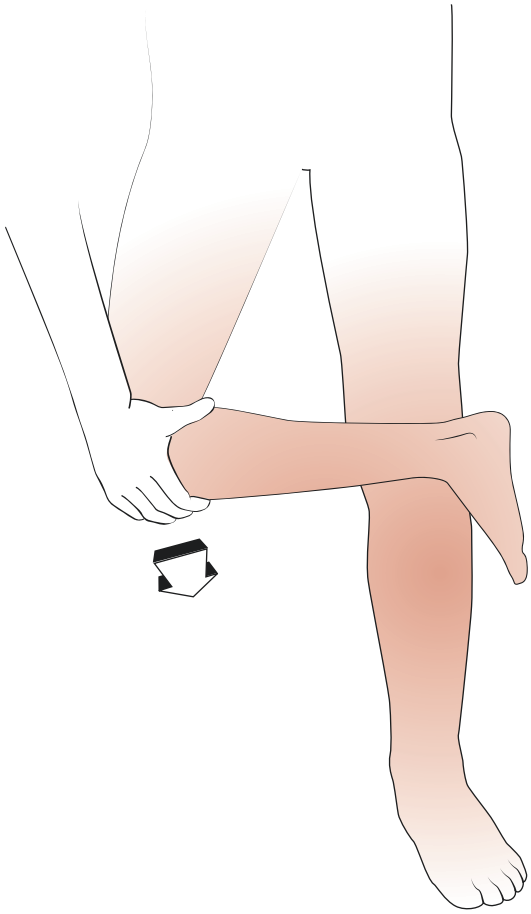


**Fig. 4.1** Patrick test (Faber test): the child lies supine. The leg is placed in a figure of "four". Next, the knee is pushed downwards. The test is positive if the patient indicates pain at the level of the ipsilateral sacroiliac joint

**Septic sacroiliitis** The pain is usually localized on one side and is often diffuse, which makes diagnosis more difficult. All signs of an infection may be present to a slight degree. Assessment reveals local pressure pain of the level of the sacroiliac joint. The pain in the sacroiliac joint increases when the hip is flexed and simultaneously adducted. Another provocation test is the Patrick or Faber test (Fig. 4.1). A needle puncture of the joint is necessary to confirm the diagnosis.

**Tumor** Pain at the pelvic level is of a variable nature and occurs primarily after sports. There may also be night pain (Table 4.1).

**Table 4.1** Bone tumors at the level of the sacrum and pelvis. The tumors identified with the § sign are rare

| Location | Benign | Malignant |
|---|---|---|
| Sacrum | Aneurysmal bone cyst | Ewing sarcoma |
| | Chondroblastoma | Lymphoma § |
| | Eosinophilic granuloma | Osteosarcoma § |
| | Osteoblastoma | |
| | Osteoid osteoma | |
| | Giant cell tumor | |
| Pelvis | Aneurysmal bone cyst | Ewing sarcoma |
| | Chondroblastoma § | Osteosarcoma § |
| | Endochondroma § | |
| | Eosinophilic granuloma | |
| | Fibrous dysplasia § | |
| | Osteochondroma (exostosis) | |
| | Osteoid osteoma § | |
| | Primary bone lymphoma | |
| | Solitary (juvenile, unicameral) bone cyst § | |

Based on Adler and Kozlowski (1993)

Supplementary assessment: an anteroposterior X-ray of the pelvis and detailed images of the sacroiliac joints. Bone scan of the spinal column and the pelvis, CT scan and MRI of the pelvis. Laboratory tests: CRP, ESR and HLA-B27.

Primary care treatment: **ankylosing spondylitis.** The treatment consists of combating pain and dosaged rest. Patients get physiotherapy to prevent crookedness in the spinal column. Back-straining activities should be avoided. The purpose of treatment is to allow the process to settle down as much as possible and to counteract deformities.

When to refer: if there are ocular anomalies, referral to an ophthalmologist should be made. Start by referring the patient to a pediatric rheumatologist if the sacroiliac joints are affected. Refer to an orthopedic surgeon if a pyogenic infection of the sacroiliac joint or a tumor of the pelvis are suspected.

Secondary care treatment: **septic sacroiliitis.** A needle puncture of the joint should be carried

out in order to make the diagnosis if there is arthritis in the sacroiliac joint. Treatment consists of rest and antibiotic therapy for 4 weeks.

**Tumor** Treatment of benign and malignant bone tumors take place in a center specialized in bone tumors.

**Differential Diagnosis: Pelvic Pain**

| | |
|---|---|
| Heel, low back, pelvic pain and/or stiff back and/or reduced respiratory excursions and/or iritis or iridocyclitis in combination with bilateral sacroiliitis. | **Ankylosing spondylitis (Bechterew disease)** |
| Inflammatory symptoms. | **Septic sacroiliitis** |
| Varying pain, especially after sports. | **Tumor** |

# Shoulder

# 5

## Shoulder Anomalies

### Absent Collarbone

Complaint: the parents discover, usually within their child's first 2 years of life, that one or both collarbone(s) are entirely or partially absent.

Assessment: absence of the clavicle can be felt on palpation. In addition, there tends to be a noticeably large forehead with widely placed eyes and a relatively small face. The chest is narrow and the shoulders hang. The anomaly may be unilateral or bilateral. A part of the clavicle may be absent, or the medial as well as the lateral part, but sometimes the entire clavicle. When the abnormality is bilateral, protraction of the shoulders is sometimes such that the shoulders can be approximated on the front side of the body (Fig. 5.1). Disability is minimal.

Diagnosis: **cleidocranial dysostosis**

Explanatory note: **cleidocranial dysostosis.** Heredity plays an important role here. In two thirds of cases the anomaly is familial (autosomal dominant hereditary). The anomaly is as common in boys as in girls. In addition to the absent clavicle other abnormalities may be present, such as a large forehead with a small face, a widened nose, hypertelorism, a small chest, and sometimes repeated voluntary shoulder and elbow dislocations. X-rays can show short and pointed distal phalanges of hands and feet. An extra epiphysis may be present in the proximal part of the metacarpal and metatarsal bones II to V. In a large number of cases hip abnormalities such as coxa vara may be present (Fig. 5.2). Children with this condition usually don't become tall. Boys reach an average height of 156 cm as adults, and girls 144 cm.

Supplementary assessment: X-ray of the clavicle to confirm the diagnosis and an anteroposterior X-ray of the pelvis to either show or rule out a coxa vara.

Primary care treatment: do not forget to request an anteroposterior X-ray of the pelvis.

When to refer: only when there is irritation of the brachial plexus. This may occur even if the clavicle is partially present. Also refer if there is a coxa vara.

Secondary care treatment: **cleidocranial dysostosis.** Operative removal may be necessary if there is irritation of the brachial plexus due to a partially present clavicle.

### Elevated Shoulder Blade

Complaint: one shoulder blade is higher than the other. There is a more or less noticeable swelling above the collarbone.


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_5

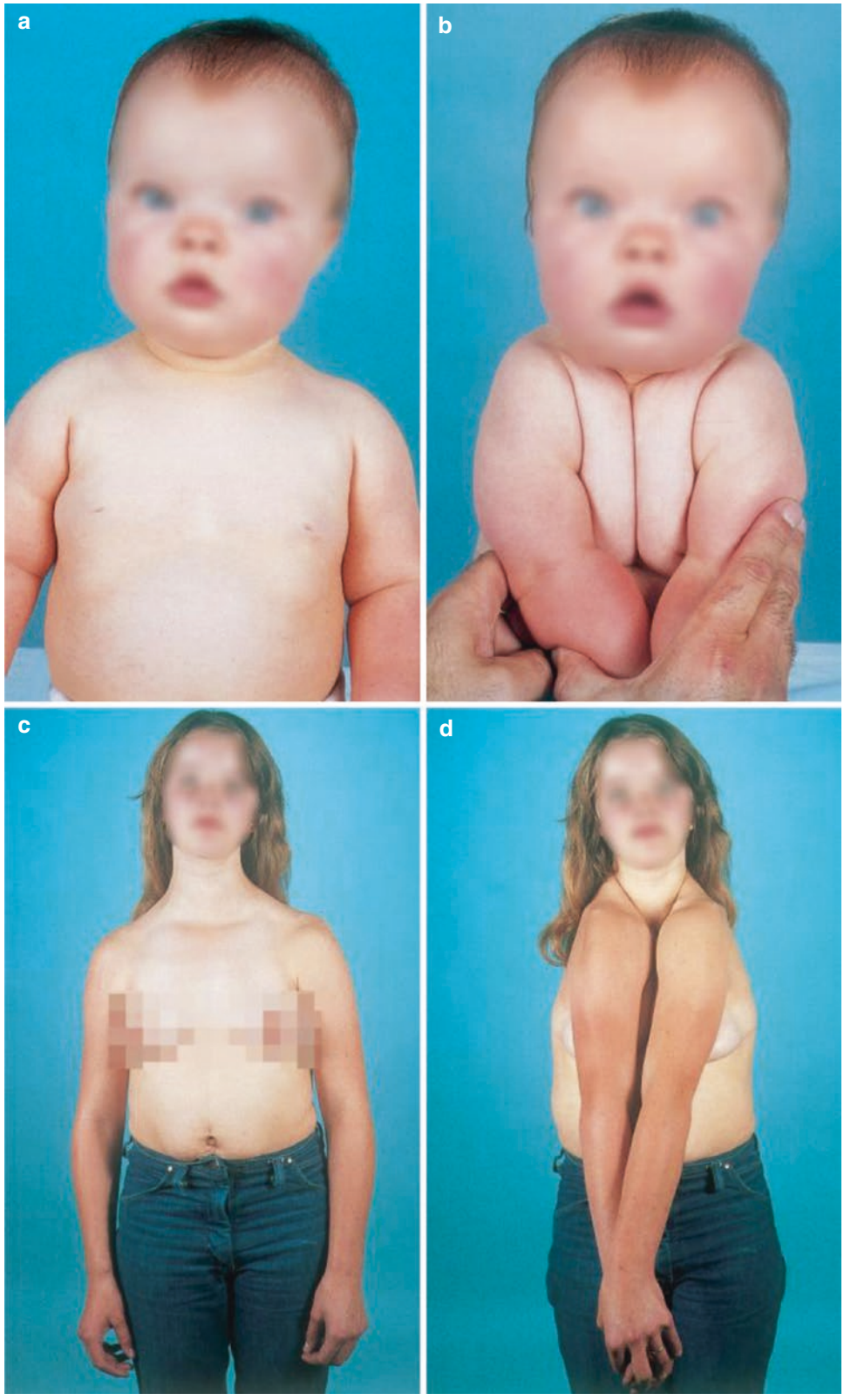



**Fig. 5.1** (**a**) Absent clavicles (cleidocranial dysostosis). (**b**) The protraction of the shoulders is very enhanced. (**c**, **d**) situation in adulthood

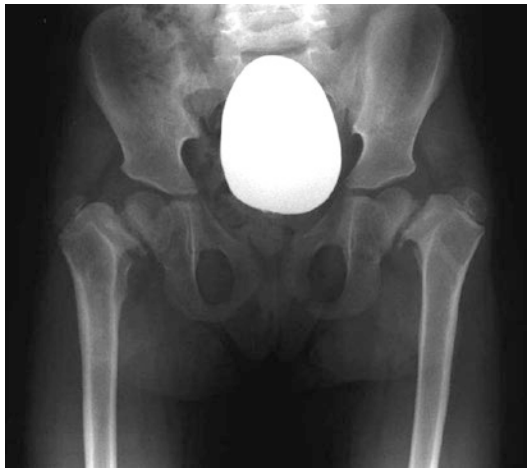


**Fig. 5.2** Coxa vara, characterized by a small angle between the femoral neck and shaft

Assessment: one shoulder blade is higher than the other and is rotated, and the upper edge of the shoulder blade projects above the collarbone. The shoulder blade is smaller than normal. Abduction of the involved shoulder tends to be limited (Fig. 5.3).

Diagnosis: **Sprengel deformity**

Explanatory note: **Sprengel deformity.** This anomaly is usually unilateral. In 70% of cases there is another congenital abnormality present such as scoliosis, kyphosis, torticollis, Klippel-Feil syndrome[1] or Poland syndrome[1]. The deformity develops because

[1]See Appendix.

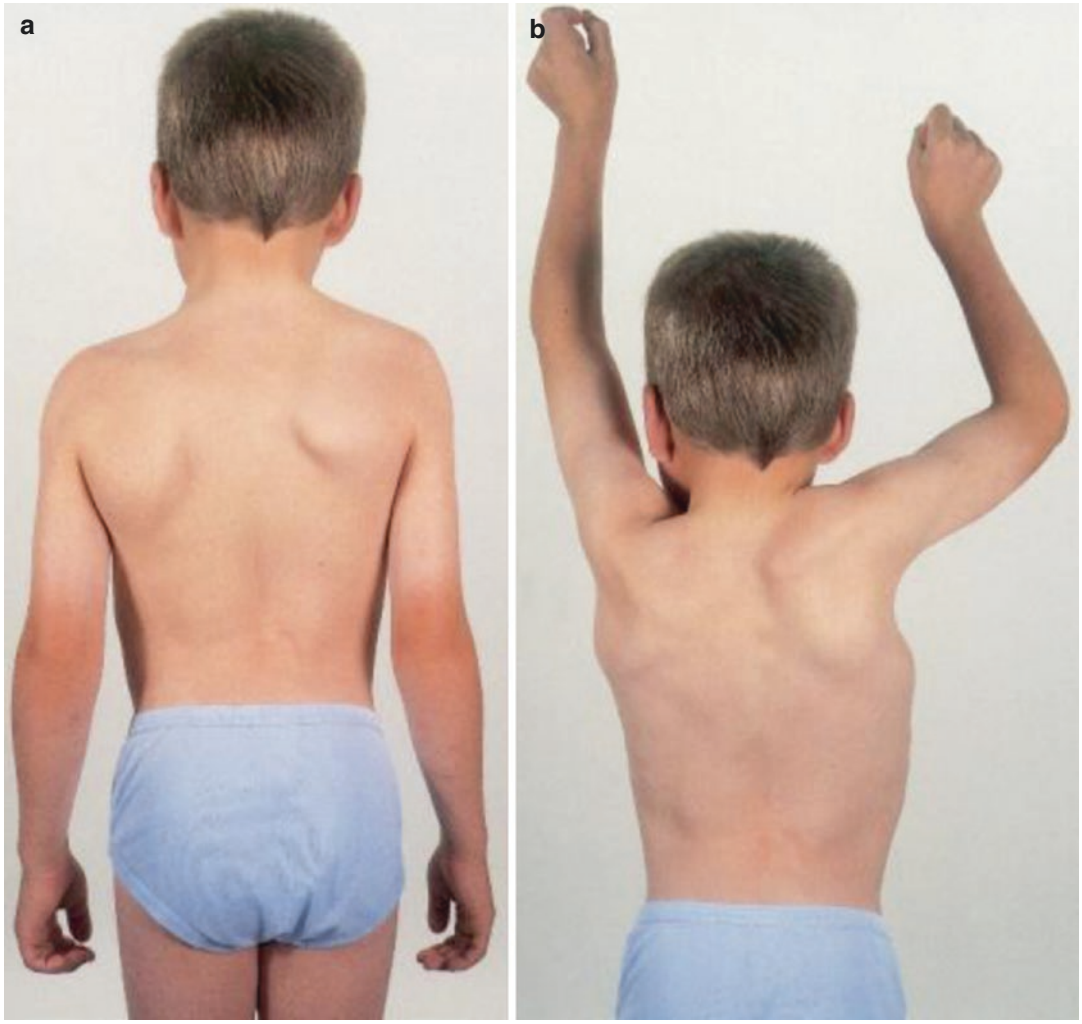



**Fig. 5.3** (**a**) Right-sided Sprengel deformity. There is an elevated right shoulder blade. (**b**) This is a mild case in which the abduction in the involved shoulder is limited to 120°

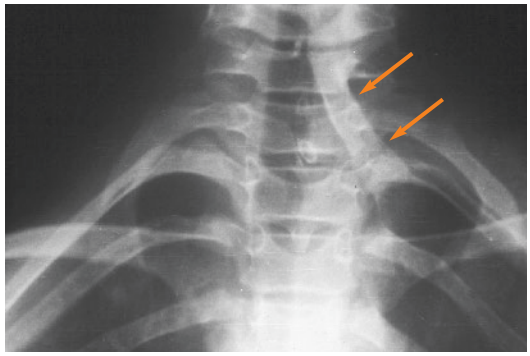


**Fig. 5.4** X-rays of an omovertebral bone (*arrows*)

in the second fetal month no distal migration of the shoulder blade occurs. Normally the scapula falls to equal levels between the second and seventh thoracic vertrebrae. The deformity varies in severity. The affected shoulder blade can be 1–12 cm higher than the normal contralateral side, with an average of 3–5 cm. Abduction is limited because of the rotation of the shoulder blade. However in about one-third of cases there is also a bony, connection between the shoulder blade and the fourth to seventh cervical vertebrae known as an omovertebral bone (Fig. 5.4). The severity of the limitation movement of varies considerably.

Supplementary assessment: X-rays of the shoulders and additionally a CT-scan or MRI to visualize an omovertebral connection (often difficult to see on a standard X-ray).

Primary care treatment: none.

When to refer: when abduction is less than 120° or if there are cosmetic issues.

Secondary care treatment: **Sprengel deformity.** No treatment should be carried out in mild cases with an abduction of 120° or more and if there are no cosmetic objections. Only if the abduction is less than 120° operative reposition of the shoulder blade will be considered. If the upper edge of the shoulder blade will project far above the collarbone but there is still a good abduction, resection of the supraspinous portion of the scapula may be considered. In severe cases, where the elevation is more than 5 cm, it is better to perform the operation in the first year of life. The operation can be performed between the ages of 3 and 8 years in less severe cases, with an elevation between 3 and 5 cm.

### Green Procedure (1957)

The muscles connecting the scapula to the trunk are divided at the scapula insertion and if present the omovertebral bone or fibrous tissue is excised. The supraspinous part of the scapula is resected. The scapula is moved distally to the level of the opposite normal side and the malrotation is corrected. The scapula is held in the new position using wire traction between the scapula and ilium. The muscles are attached more proximally to the scapula in its new position. The traction wire is removed after 3 weeks.

### Woodward Procedure (1961)

The Woodward procedure is general preferred. The trapezius and rhomboid muscles are detached from their origins on the trunk. The omovertebral bone or fibrous tissue is excised if present. The supraspinous part of the scapula is resected. The scapula is moved distally and the malrotation is corrected. The origins of the trapezius and rhomboid muscles are relocated onto the spinous processes at a more distal level (Fig. 5.5). In severe deformities several authors have recommended a double osteotomy or morcellation of the clavicle on the ipsilateral side as a first step to prevent compression of the brachial plexus between the clavicle and first rib.

## Repeated Non Traumatic Shoulder Subluxation/Dislocation

Complaint: the patient can subluxate or dislocate one or both shoulders spontaneously or on command. There has been no appreciable trauma.

Assessment: on command, the patient can dislocate the glenohumeral joint, in one or several directions (multidirectionally). In the latter case the examiner can shift the humeral head with respect to the glenoid

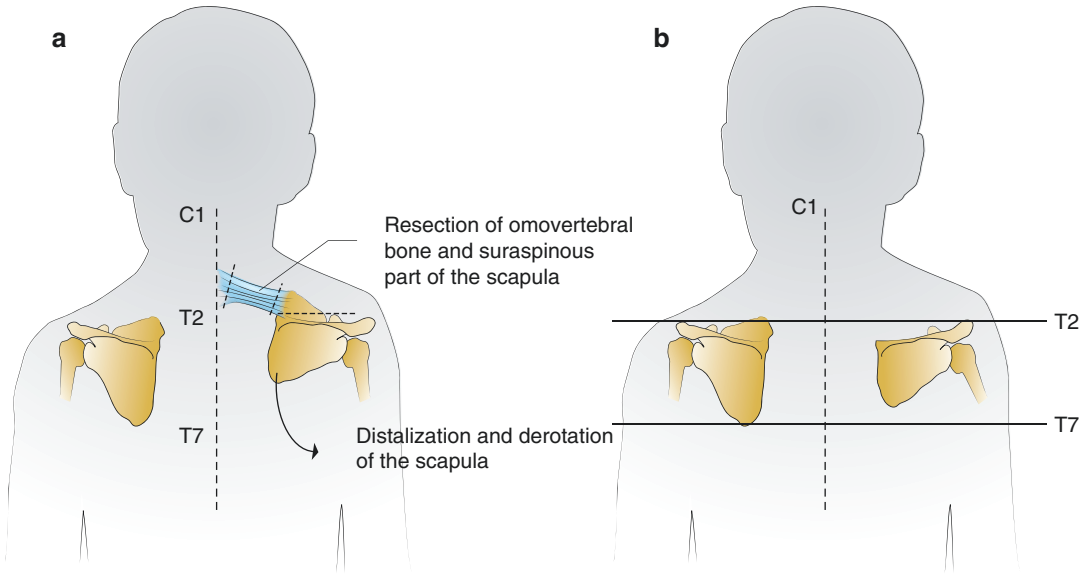



**Fig. 5.5** (**a**) Woodward procedure: the trapezius and rhomboid muscles are detached from their origins on the trunk. The omovertebral bone and the supraspinous part of the scapula are excised. (**b**) Next the scapula is moved distally, the malrotation is corrected and held in it's new position, by reattaching the muscles onto the trunk at a more distal level (Redrawn from: Woodward JW. Congenital elevation of the scapula by release and transplantation of muscle origins. J Bone Joint Surg Am. 1961;43-A:219–28)

cavity forwards and backwards (drawer test) (Fig. 5.6) and pull it downwards.

By pulling on the hanging arm one sees an indentation, the sulcus sign, which appears between the acromion and the humeral head. The sulcus sign is a consequence of a distal subluxation of the humeral head (Fig. 5.7).

Differential diagnosis:

**recurrent non traumatic glenohumeral joint subluxation/dislocation**
voluntary glenohumeral joint subluxation/dislocation
habitual glenohumeral joint subluxation/dislocation

Explanatory note: **recurrent non traumatic glenohumeral joint subluxation/dislocation.** There are two conditions in which painless repeated dislocation of the shoulder may occure: voluntary glenohumeral joint subluxation/dislocation and habitual glenohumeral joint subluxation/dislocation. One condition occurs after trauma and is painful: recurrent posttraumatic glenohumeral joint subluxation/dislocation (see p. 67).

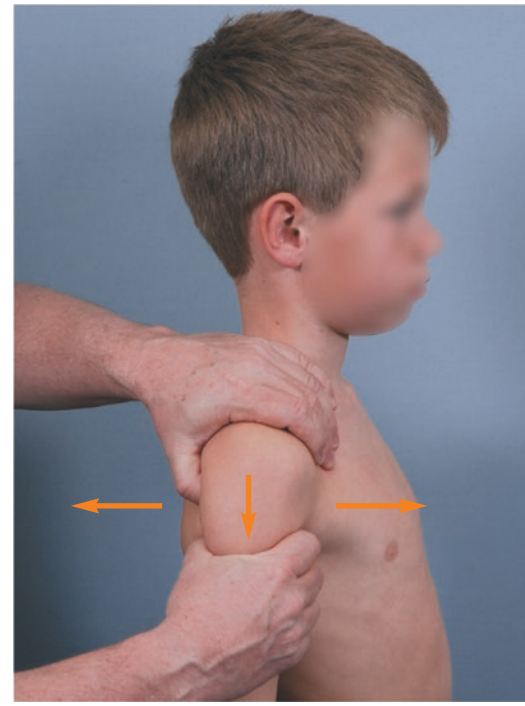


**Fig. 5.6** Drawer test for the shoulder: the examiner fixes the scapula with one hand while fixing the humeral head with the other. The humeral head can now be moved anteriorly or posteriorly or distally with respect to the glenoid (*arrows*)

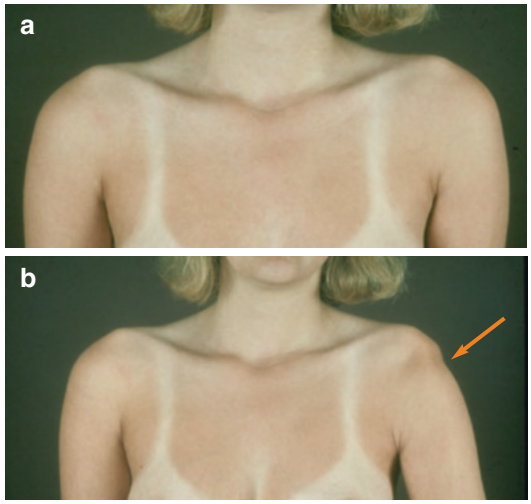



**Fig. 5.7** Habitual dislocation of the left shoulder. (**a**) Normal situation. (**b**) Situation after the left glenohumeral joint has been dislocated distally on command. As a consequence of the subluxation/dislocation an indentation between the acromion and the humeral head can be seen (*arrow*). This is known as a sulcus sign

*Voluntary glenohumeral joint subluxation/dislocation* This type is associated with severe ligamentous laxity due to hypermobility disorders such as in Down[2] and Ehlers-Danlos syndromes[2].

*Habitual glenohumeral joint subluxation/dislocation* This anomaly refers to repeated subluxation/dislocation in normal children with no associated ligamentous laxity.

Supplementary assessment: X-rays and CT-scan of the shoulder.

Primary care treatment: voluntary and habitual glenohumeral joint subluxation/dislocation are painless and need no manipulated reduction. These children can do it by themselves by contraction of certain shoulder muscles depending on the direction of the subluxation/dislocation. The patient is strongly discouraged from inciting dislocations. Physiotherapy will be prescribed with musclestrengthening exercises. The voluntary and habitual dislocations often disappear as the children get older.

When to refer: when lifestyle rules and muscle strengthening exercises produce insufficient results. There is no need to refer a habitual glenohumeral joint subluxation/dislocation. Spontaneous cure occurs in most of the cases after about 2 years.

Secondary care treatment: **voluntary glenohumeral joint subluxation/dislocation.** Operative treatment is seldom indicated and should be avoided as much as possible. Especially multidirectional dislocations often give disappointing results. Only in those cases where the dislocation occurs in a single direction should capsulorraphy be considered. In problem cases it may be necessary to place a bone block to limit motions and prevent dislocations. An anterior bone block for anterior and a posterior bone block for posterior subluxations/dislocations. Arthrodesis may be required as a last resort in those cases that remain symptomatic despite other treatments.

## Repeated Traumatic Shoulder Subluxation/Dislocation

Complaint: a recurrent subluxation/dislocation may occur after trivial traumas after a significant initial trauma that has resulted in a dislocation.

Assessment: the mobility of the shoulder joint is normal. A stability test for the shoulder is the drawer test in which the patient sits on a chair and lets his arms hang, relaxed. The examiner fixes the scapula with one hand while fixing the humeral head with the other. The humeral head can now be moved forwards, backwards and downwards with respect to the glenoid cavity (Fig. 5.6). The examiner will feel that something has shifted or "jumped up" in the case of a lesion of the glenoid labrum. About the same feeling can be experienced when you rub the knuckles of your fists over each other. This test serves to assess anterior as well as posterior and inferior instability. Posterior instability can be assessed with what is known as the posterior stress test. In this test the

[2]See Appendix.

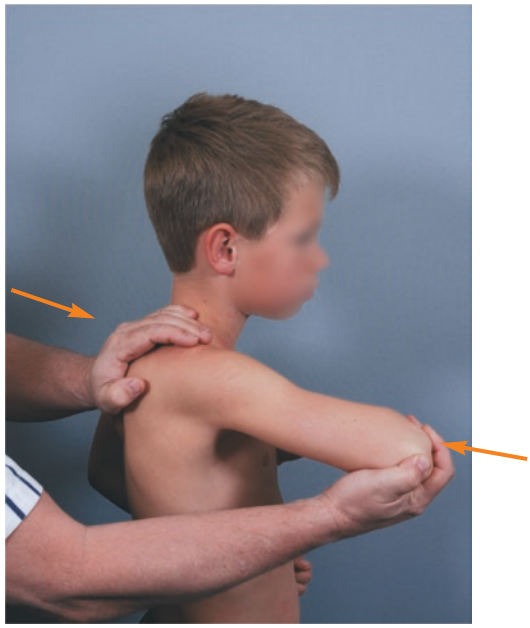


**Fig. 5.8** Posterior stress test: in this test the shoulder is brought into 90° anteflexion and internal rotation. The elbow is flexed. The examiner holds back the shoulder blade with one hand and presses the elbow posteriorly with the other hand. The test is positive if there is pain and/or posterior displacement of the humeral head (*arrows*)

shoulder is brought into 90° anteflexion and internal rotation, and the elbow is flexed. The examiner holds back the shoulder blade with one hand and presses the elbow posteriorly with the other hand. The test is positive when there is pain and/or posterior displacement of the humeral head (Fig. 5.8). Anterior instability can be determined using the apprehension test. The patient sits in a chair, lies down or stands. The arm is abducted to 90° and maximally externally rotated. The test is positive if this is painful or if the humeral head (sub)luxates anteriorly (Fig. 5.9).

Differential diagnosis:

**Recurrent posttraumatic glenohumeral joint subluxation/dislocation**

Anterior glenohumeral joint subluxation/dislocation.

Posterior glenohumeral joint subluxation/dislocation.

Multidirectional sublglenohumeral joint subluxation/dislocation

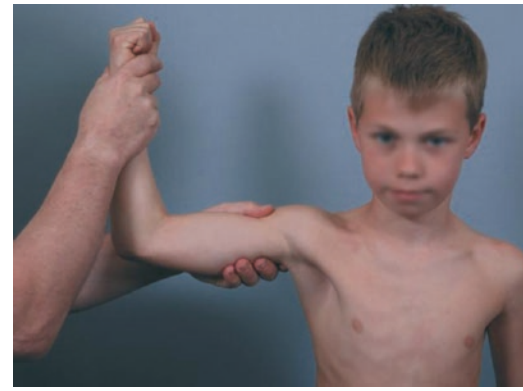


**Fig. 5.9** Apprehension test: the arm is abducted 90° and maximally external rotated. The test is positive if this is painful or if the humeral head (sub)luxates anteriorly

Explanatory note: **recurrent posttraumatic glenohumeral joint subluxation/dislocation.** In 99 % of cases these are anterior subluxations/dislocations and in less than 1 % posterior. In rare cases there is multidirectional instability.

*Anterior glenohumeral joint subluxation/dislocation* Recurrent anterior subluxations/dislocations occur in more than 60 % of cases after an initial dislocation in patients younger than age 20. In patients older than 40 this is only 6 %. A recurrent anterior subluxation/dislocation occurs when the shoulder is abducted for more than 90° and fully externally rotated. This position occurs, for example, when swimming a back stroke, or when hitting a volleyball or even when putting on a coat. A recurrent anterior dislocation can generally be reduced easily and most patients are capable of doing this themselves. Some of them have learned to prevent such recurrent dislocations and do not wish to have any further treatment. Others cannot accept this and they will come to you for advice.

After the initial dislocation permanent anomalies have occurred such as a detached anteroinferior part of the glenoid labrum, known as a Bankart lesion (Fig. 5.10). In 3–30 % of adults this is accompanied by an avulsion fracture on the anteroinferior side of the glenoid cavity or a fracture through this socket. The major difference in the percentages is explained by the fact that in the lower percentage group only the

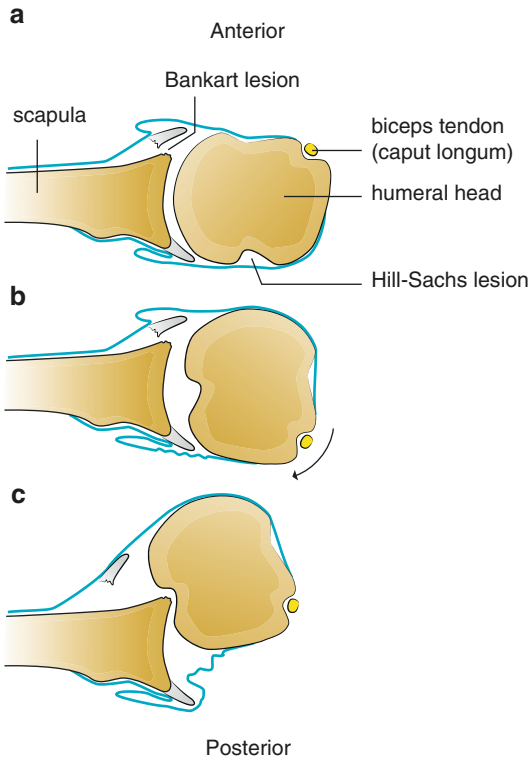



**Fig. 5.10** Cross-section of the right shoulder joint. (**a**) A Bankart lesion can occur on the anteroinferior side of the glenoid cavity as a result of the initial anterior dislocation. In addition to the Bankart lesion there may be an indentation fracture on the posterior side of the humeral head, known as a Hill-Sachs lesion. (**b**) The Hill-Sachs lesion turns forward with external rotation of the shoulder. (**c**) In a recurrent dislocation the Hill-Sachs lesion can lock on the anterior side of the glenoid cavity

fractures of the glenoid cavity are counted, and in the high percentage group avulsion fractures are also included. These fractures do not occur in young patients, but the glenoid labrum on the anteroinferior side of the glenoid cavity is torn.

In addition to a Bankart lesion, in 10–50 % of cases there is an indentation fracture on the posterior side of the humeral head, this is a Hill-Sachs lesion, caused when the hard edge of the glenoid cavity produces an indentation on the spongeous humeral head during the subluxation/dislocation (Figs. 5.10 and 5.11). This indentation fracture may be so small that it almost goes unrecognized, but can also be as large as one-third of the size of the humeral head. This also

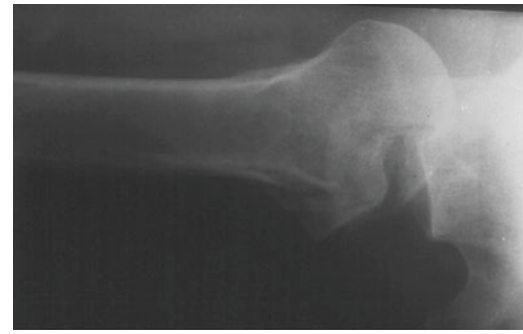


**Fig. 5.11** Anterior dislocation of the humeral head

explains its widespread publication in the literature. There is a greater tubercle fracture in 40 % of the primarily dislocations in 12–13-year-olds. Strangely enough, after an initial dislocation patients who also have a greater tubercle fracture experience almost no recurrent dislocations.

*Posterior glenohumeral joint subluxation/dislocation* In recurrent posterior dislocations, there is a torn glenoid labrum or bony Bankart lesion but it is now on the posterior side of the glenoid cavity. Just as with an anterior subluxation/dislocation there can be an indentation (Hill-Sachs lesion), but it is now on the anterior side of the humeral head.

*Multidirectional glenohumeral joint subluxation/dislocation* This is a rare manifestation. The head luxates forwards, backwards and downwards.

Supplementary assessment: X-rays of the shoulder joint in two planes (Fig. 5.11), supplemented with an MRI-arthrogram.

Primary care treatment: if there have been one or two recurrences, refer to a physiotherapist for shoulder musclestrengthening exercises. If there is a recurrent anterior subluxation/dislocation, muscle-strengthening exercises for the internal rotators should be prescribed and in a recurrent posterior subluxation/dislocation focus on the external rotators.

When to refer: when the shoulder has dislocated three times or more.

Secondary care treatment: **recurrent post-traumatic glenohumeral joint subluxation/**

**dislocation.** The most common techniques used for recurrent subluxation/dislocation of the shoulder are:

*Anterior glenohumeral subluxation/dislocation* Fixation of the torn glenoid labrum (Bankart lesion). This is being increasingly carried out arthroscopically with tightening of the stretched capsule at the same time.

In an open procedure the joint is opened up with a much larger incision, the Bankart lesion is corrected and the subscapular tendon is reefed. This is known as the Putti Platt procedure. The technique does cause a loss of external rotation, which can hinder carrying out certain sports like tennis, volleyball, basketball, handball, baseball, javelin throwing, etc. This problem may eventually be corrected by doing a proximal humeral external rotation osteotomy.

There is a recurrence of 5 % after the open procedure and 10 % after arthroscopic surgery.

*Posterior glenohumeral subluxation/dislocation* Treatment is mostly conservative, with muscle-strengthening exercises, especially for the external rotators. An operation should be considered only in exceptional cases if the patient has extreme discomfort. In such cases a choice can be made to restore the torn glenoid labrum or bony Bankart lesion on the posterior side of the glenoid cavity and/or reef the joint capsule on the posterior side. In a large Hill-Sachs lesion the insertion of the subscapular muscle is fixed into the defect. An alternative is an osteotomy on the posterior side of the scapular neck, aimed at anterior repositioning of the glenoid cavity. One can also try to prevent posterior subluxation/dislocation by inserting a block of bone on the posterior side of the glenohumeral joint, attaching this to the posterior side of the scapular neck. The result of operations for a recurrent posterior subluxation/dislocation is often unpredictable.

*Multidirectional glenohumeral subluxation/dislocation* Operative treatment involves pulling up the lowest part of the capsule of the glenohumeral joint by reefing and reattaching it in what is known as the "inferior capsular shift procedure".

## Shoulder Swelling

### Collarbone Swelling in Newborns

Complaint: immediately after birth the parents or carers notice that their child has a non-painful swelling at the level of the collarbone. There are no indications of a birthrelated or other trauma.

Assessment: there is a swelling at the level of the collarbone. The swelling is not painful. The movements of the shoulder joint are normal.

Diagnosis: **congenital pseudarthrosis of the clavicle**

Explanatory note: **congenital pseudarthrosis of the clavicle** (Fig. 5.12). This rare congenital abnormality appears on the right side in 90 % of cases. The long medial (sternal) part is pulled upwards with respect to the shorter lateral part.

Supplementary assessment: anteroposterior X-ray of the clavicle.

Primary care treatment: request an X-ray of the clavicle to make the diagnosis with certainty.

When to refer: it is usually only a cosmetic problem and treatment can be avoided. Refer only when the parents want surgical correction.

Secondary care treatment: **congenital pseudarthrosis of the clavicle.** The sclerotic bone ends at the level of the pseudarthrosis may be

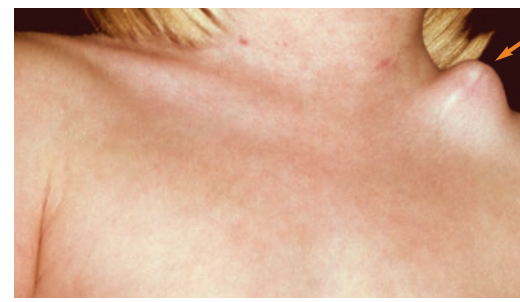


**Fig. 5.12** Congenital pseudarthrosis of the left clavicle

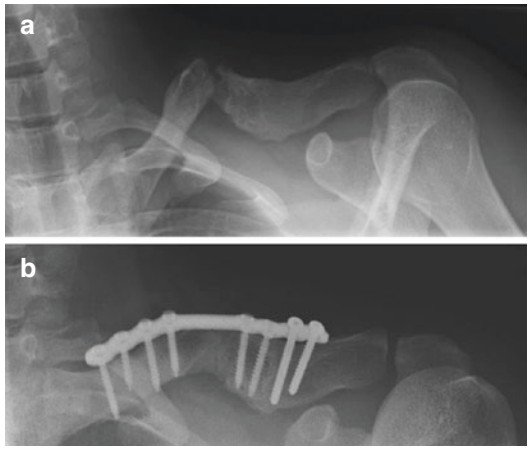



**Fig. 5.13** (**a**) Congenital pseudarthrosis of the left clavicle. (**b**) Fixed with a plate and screws

resected in young children until about age 4. It is important to make sure that the periosteum is divided lengthwise and afterwards carefully closed with absorble sutures. The continuity of the clavicle usually recovers after this.

After the age of 4 treatment consists of an operation in which the pseudarthrosis is removed. The two fragments of the clavicle are fixed using a plate and screws (Fig. 5.13). A spongious bone graft is inserted at the level of the excised pseudarthrosis.

## Swelling of the Shoulder Blade or Collarbone or Upper Arm

Complaint: swelling at the level of the shoulder.

Assessment: a hard bony swelling.

Differential diagnosis:

**benign bone tumor**
**malignant bone tumor**

Explanatory note: primary malignant bone tumors are estimated at 8 per million inhabitants per year. The frequency of benign bone tumors is not known. It is estimated to be about 10 times more frequent than malignant bone tumors.

**Table 5.1** Tumors at the shoulder girdle level

| | Benign bone tumors | Malignant bone tumors |
|---|---|---|
| **Humerus** | Unicameral bone cyst | Osteosarcoma |
| | Osteochondroma | Ewing sarcoma |
| | Chondroblastoma | |
| **Clavicle** | Eosinophilic granuloma | Osteosarcoma |
| | | Ewing sarcoma |
| **Scapula** | Osteochondroma | Osteosarcoma |
| | Eosinophilic granuloma | Ewing sarcoma |
| | Aneurysmal bone cyst | |
| | Fibrous dysplasia | |

Based on Adler and Kozlowski (1993)

**Benign bone tumor** (Table 5.1) The most common bony tumor is at the level of the shoulder joint, which is accompanied by a swelling. This is an osteochondroma, also known as osteocartilaginous exostosis or exostosis (Fig. 5.14). It involves a developmental disorder in the periphery of the epiphyseal growth plate. This can produce a bony outgrowth that usually has a cartilaginous top. This is usually a hard painless swelling that can give rise to symptoms when muscles and tendons rub against it. The anomaly increases in size until growth is completed. Such an abnormality can be solitary but can also occur in several other places. In this case there is a 60 % chance of a hereditary condition. Hereditary multiple osteochondromata occur in about 10–25 in one million.

Osteochondromata are present in the proximal part of the humerus in more than 70 % of cases.

**Malignant bone tumor** (Table 5.1) There are few symptoms in the early stages. Generally the first symptoms are of a vague, variable pain, especially after sports activities. The pain is gradually experienced throughout the whole day and there may be nocturnal pain. ! Usually the tumor is only palpable at a later stage, as it increases in size. Pathological fractures only occur as the last symptoms in these tumors.

Supplementary assessment: standard X-rays. Tumors may generally be seen on them.

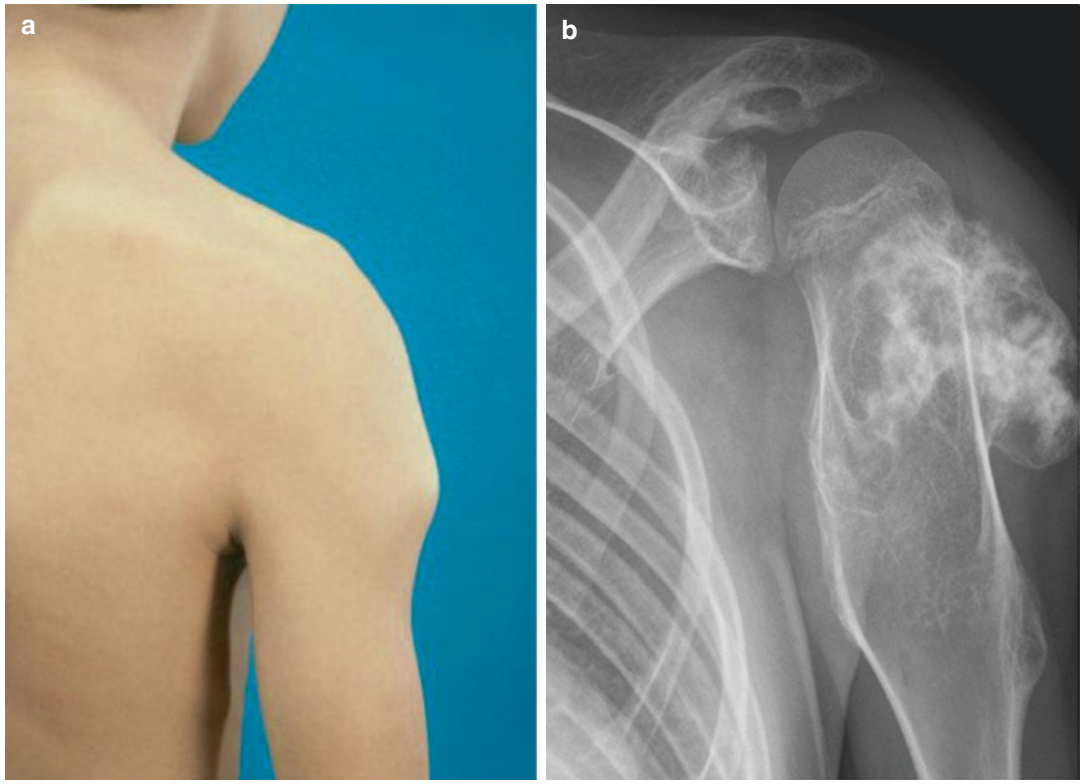



**Fig. 5.14** (**a**) Osteochondroma (exostosis) at the level of the proximal part of the humerus. (**b**) X-ray of this osteochondroma

However if there is any doubt, a bone scintigraphy and MRI should be requested.

Primary care treatment: in the case of bone pain the possibility of a malignant tumor should always be considered.

When to refer: when the X-rays or MRI show a bone lesion or when increased activity is observed on bone scintigraphy.

Secondary care treatment: **benign bone tumor.** An osteochondroma will only be removed if it is causing mechanical symptoms or pain.

**Malignant bone tumor** Treatment of primary malignant tumors must take place in a medical center specialized in bone tumors. A combination of surgery and chemotherapy is preferable unless the tumor is sensitive to chemotherapy. Occasionally radiotherapy may be necessary. If possible, bony defects are reconstructed with a prosthesis. Amputation or exarticulation will only be carried out in cases where reconstruction is no longer possible.

## Shoulder Movement Limitations in Newborns

### Paralysis of the Arm at Birth

Complaint: there is a limp arm immediately after birth.

Assessment: there is a limp arm.

Differential diagnosis:

**neonatal brachial plexus palsy**
Erb palsy (Erb-Duchenne palsy)
Erb-Duchenne-Klumpke palsy
Klumpke palsy

Explanatory note: **neonatal brachial plexus palsy.** A paralysis of the arm at birth occurs due to an abnormal traction on the arm during delivery, often with a high birth weight of over 3500 g with an abnormal position during the delivery. It occurs primarily in occipital presentations with the shoulder as the part lying anteriorly (shoulder dystocia) and in breech presentations. The incidence is 1 in 1000 births. Additional lesions may be a clavicular or humeral fracture (10 %) and a facial nerve paresis (10 %). Brain damage may also occur in the case of a traumatic birth. There is also a torticollis in a large number of cases.

There is a classical classification as well as a classification according to severity of the anomaly. The classical classification is as follows:

*Erb palsy (type I)* This is the most common (90 %). It involves an injury to the nerve roots from C5 and C6 with the corresponding motor and to a lesser degree sensory deficit. In this type of paralysis there is weakness of the abductors and external rotators of the shoulder, flexors of the elbow and supinators of the lower arm. The abnormality is easy to recognize. The arm is adducted and internally rotated at the level of the shoulder, the elbow is extended and the forearm lies in pronation. The wrist and fingers can be moved.

*Erb-Duchenne-Klumpke palsy (type II)* There is failure of the entire brachial plexus (C5 to T1). The arm is generally abducted at the level of the shoulder; the arm is weak, the wrist is flexed and there is a claw hand. There tends to be considerable loss of sensation. A Horner syndrome may also be present, manifested by a small pupil and an upper eyelid that hangs low (ptosis). In 5 % of cases the nerve root of C4 is also affected, resulting in a unilateral paralysis of the diaphragm with possible respiratory problems.

*Klumpke palsy (type III)* In this palsy there is damage to C7, C8 and T1. There is paralysis of motor functions in the thumb and fingers. A Horner syndrome may also be present.

Another classification groups the paralyses according to the severity of the abnormality. Here a distinction is made between a mild paralysis corresponding to the Erb palsy (C5, C6), moderate paralysis (C5, C6 and C7) and severe paralysis (C5 to T1), which corresponds to the Erb-Duchenne-Klumpke palsy (Fig. 5.15).

The moderate paralysis (C5, C6 and C7) is different from Erb palsy in that, besides adduction, internal rotation of the shoulder and pronation of the forearm there is also slight flexion of the elbow resulting from paralysis of the triceps muscle as well as flexion of the wrist resulting from paralysis of the wrist extensors.

At a later age manifestations of a birth paralysis of the arm may still be seen to a lesser or greater degree depending on the level of recovery. In severe cases the growth in length of the arm is reduced by 20 % compared to the healthy side. Due to the disturbed muscular balance a posterior subluxation/dislocation or an adduction and internal rotation contracture of the shoulder may occur. Especially in Erb palsy limited supination of the forearm may be present usually with little functional problems.

Supplementary assessment: X-rays of the clavicle and humerus. In 10 % of cases there is a fracture of the clavicle or humerus. In the case of an adduction and internal rotation contracture of the shoulder, ultrasonography is recommended to see if there is any posterior dislocation in the glenohumeral joint.

Primary care treatment: wait-and-see. Spontaneous recovery occurs in 90 % of cases. The greatest improvement occurs in the first 3 months; further recovery is possible until 2 years of age. The prognosis is favourable if flexion in the elbow is restored within 3 months. ! It is prudent to instruct the parents to keep moving the arm joints at each nappy change. External rotation of the shoulder should especially be exercised. There may be a posterior shoulder dislocation if shoulder external rotation diminishes in the course of time. In this case ultrasonography assessment of the shoulder is indicated.

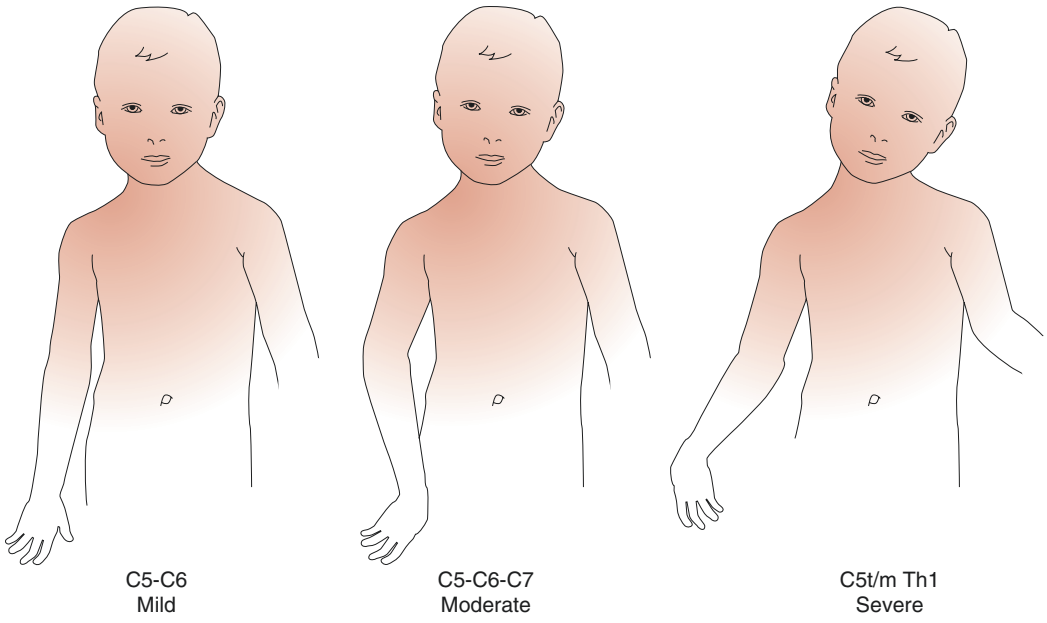



**Fig. 5.15** Classification of a neonatal brachial plexus palsy according to severity of the anomaly

When to refer: if there is no active flexion of the elbow after 3 months and if a posterior shoulder subluxation/dislocation is suspected.

Secondary care treatment: **neonatal brachial plexus palsy.** Surgical treatment of the plexus is controversial. Avulsions of the nerve roots cannot be repaired. A neurolysis and nerve transplants with microsurgery may be considered in more peripheral lesions. Operative treatment is carried out between 4 months and 1 year of age. The results are unpredictable.

A posterior shoulder subluxation/dislocation should be avoided as much as possible by doing muscle-strengthening exercises for the external rotators. However, if a posterior shoulder subluxation/dislocation has occurred it can to be reduced noninvasively or if necessary operatively. Operative treatment comprises detachment of the insertions of the pectoralis major and teres major muscles and a transposition of the latissimus dorsi and teres major muscles onto the rotator cuff.

A very common residual anomaly is lost abduction and external rotation in the shoulder. In an adduction/internal rotation contracture of the shoulder the most applied operative procedure is the transposition according to Sever L'Episcopo. In this operation the attachments of the sternal part of the pectoralis major and subscapularis muscles and the joint capsule on the anterior side of the glenohumeral joint are detached. The insertion of the tendons of the teres major and latissimus dorsi muscles are transferred from the anteromedial side to the posterolateral side of the humerus. This operation is usually performed in young children.

A transposition of the latissimus dorsi and teres major muscles to the rotator cuff is performed if there is only weakness in the abductors (Fig. 5.16). In older children an external rotation osteotomy of the upper arm may be performed, aiming for a situation with as much internal rotation as external rotation if the muscles are too weak to allow the Severe L'Episcopo transposition.

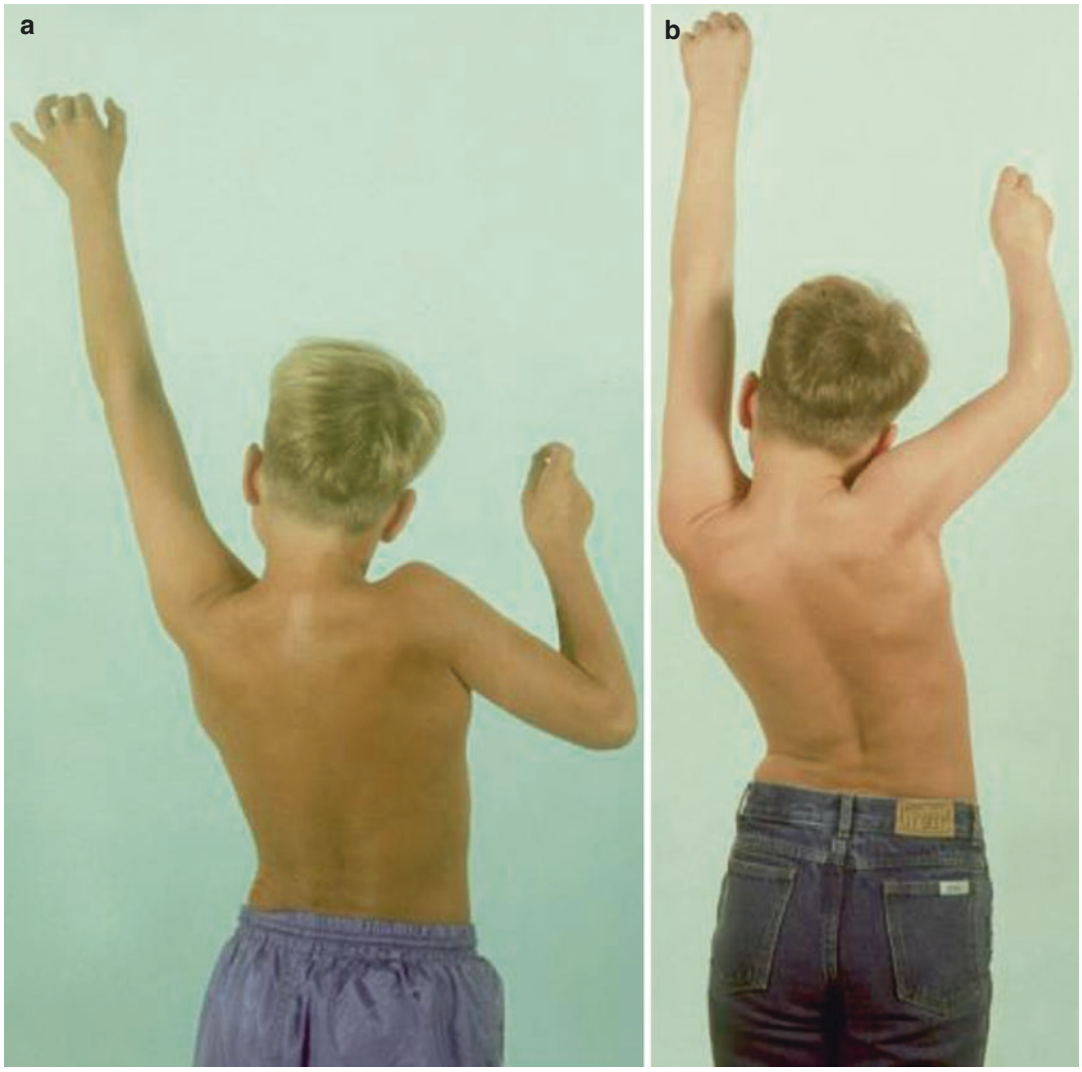



**Fig. 5.16** (**a**) Neonatal brachial plexus palsy with a residual anomaly of the right arm and an abduction weakness of the right shoulder. (**b**) Situation after transposition of the insertion of the teres major and the latissimus dorsi tendons from the anteromedial side to the posterolateral side of the rotator cuff of the right shoulder

## Shoulder Pain

### Vague Shoulder Pain

Complaint: the child complains about pain in the proximal part of the upper arm. The nature of the pain keeps changing and occurs primarily after sport activities. ! The child is not sick and has full movement of the shoulder and there may be nocturnal pain.

Assessment: at first shoulder assessment shows no irregularities. Sometimes there is slight pressure pain in the proximal part of the humerus.

Diagnosis: **malignant bone tumor** (see Table 5.1)

For explanatory note, supplementary assessment, primary care treatment, when to refer and secondary care treatment, see pp. 70, 71.

## Shoulder Pain After a Trivial Trauma

Complaint: the child complains about shoulder pain after a trivial trauma.

Assessment: mobility of the shoulder is mostly limited by the pain. There is pressure pain in the proximal part of the upper arm.

Diagnosis: **fractured unicameral bone cyst (juvenile-, solitary-, simple bone cyst)**

Explanatory note: **fractured unicameral bone cyst.** An unicameral bone cyst is asymptomatic unless fractured. The cyst develops in the first or second decade of life. It is filled with yellow fluid and lined with a fibrous capsule. 70 % are males. In 60 % of cases it is localized in the proximal part of the humerus and in 30 % in the proximal part of the femur. An unicameral bone cyst is generally localized in a metaphysis, but due to growth it ends up being localized in the diaphysis. In more than 80 % of cases a spontaneous fracture is the first symptom in this so far symptom-free anomaly.

Supplementary assessment: X-rays of the shoulder in two planes (Fig. 5.17).

Primary care treatment: if the cyst is discovered by accident one can wait, perhaps with radiological checkups once a year.

When to refer: when there are complaints and if there is doubt about the diagnosis.

Secondary care treatment: **fractured unicameral bone cyst.** The treatment of a fractured unicameral bone cyst is no different to the treatment of other fractures. Sometimes the bone cyst disappears after the fracture. However such chances are less than 25 % (Fig. 5.17).

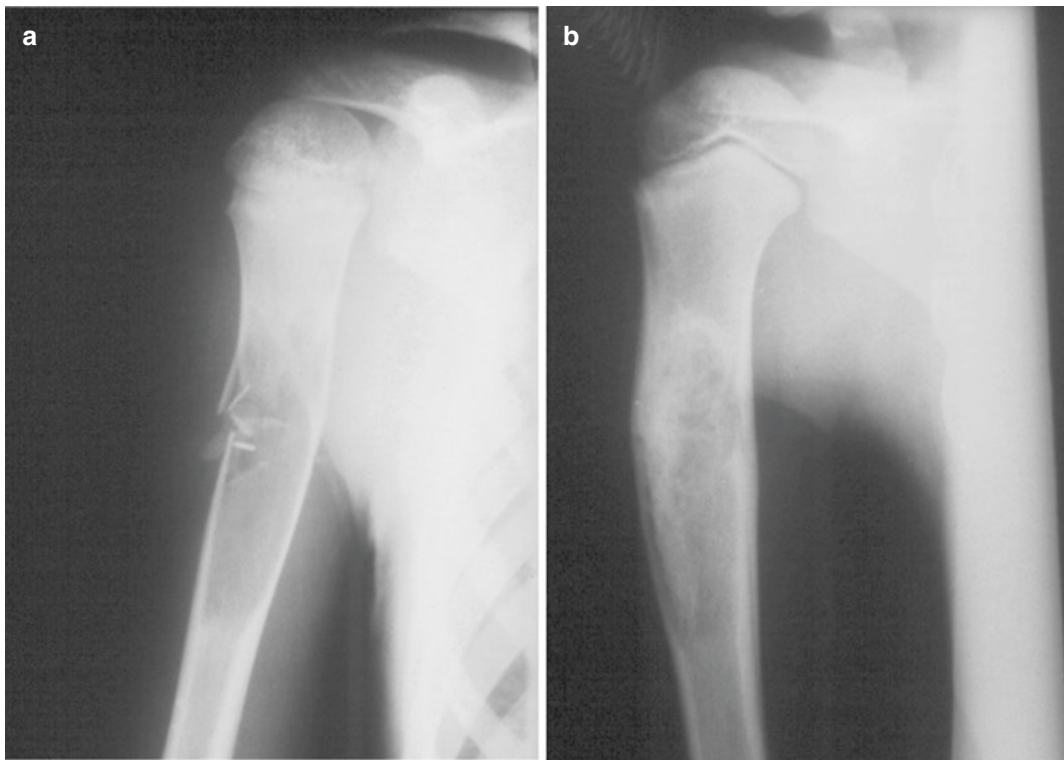



**Fig. 5.17** (**a**) Fracture caused by a unicameral bone cyst of the right humerus. (**b**) Sometimes a bone cyst disappears after the fracture, however, such chances are less than 1 %

Treatment of such bone cysts is postponed if possible until the end of puberty. By then the bone cyst has migrated from the metaphysis to the diaphysis. The further the solitary bone cyst lies from a growth plate, the more inactive it is and the smaller chances are of recurrence after treatment. The chances of recurrence are greatest in children younger than 10 years of age. The first treatment choice is local injection with corticosteroids. This treatment gives full recovery in 40% of cases, incomplete recovery in 50% and has no effect in 10%. The local injection with corticosteroids may be repeated three to four times as needed, with periods of several months in-between. Irresponsive cases can be treated with curettage and grafting with autogenous or bank bone.

## Shoulder Pain in Overhead Athletes

Complaint: pain in the shoulder after intensive activity in baseball, javelin throwing and tennis.

Assessment: the movements of the shoulders tend to be painful. There is pressure pain at the level of the humeral head.

Diagnosis: **Little League shoulder (proximal humeral epiphysiolysis, osteochondrosis of the proximal humerus, traction apophysis of the proximal humerus)**

Explanatory note: **Little League shoulder.** Repeated overhead throwing such as in baseball, javelin throwing or in tennis may lead to a stress fracture of the proximal humeral growth plate, mostly between 3 and 4 years of age. In the initial stage X-rays show a widening of the growth plate, followed by metaphysial periosteal bone formation (Fig. 5.18). The term Little League shoulder originates in the USA and refers to young baseball pitchers.

Supplementary assessment: X-rays of the shoulder in two planes.

Primary care treatment: a sling for 6 weeks.

When to refer: referral is not necessary.

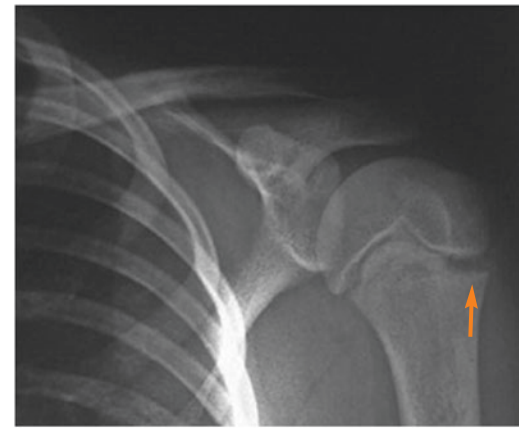


**Fig. 5.18** Widening of the proximal humeral growth plate, especially on the lateral side (apophysis)

## Painful Shoulder Movement Trajectory

Complaint: pain during overhead activities.

Assessment: in shoulder abduction and anteflexion there is a painful arc.

Differential diagnosis:

**insufficient rotator cuff**
**Bankart lesion**

Explanatory note: **insufficient rotator cuff.** In adolescent mountaineers, tennis players and swimmers chronic overload of the rotator cuff can cause a relative insufficiency in it, leading to internal impingement in abduction.

**Bankart lesion** Sometimes a torn glenoid labrum on the anteroinferior side of the glenoid cavity (Bankart lesion), is responsible for an internal impingement of the shoulder without subluxations. Assessment reveals a positive anterior drawer test and/or a positive apprehension test (see pp. 65 and 67).

Supplementary assessment: X-rays of the shoulder in two planes and a MRA should be carried out to find out if there is a Bankart lesion or not.

Primary care treatment: referral to a sports physiotherapist is necessary in the case of insufficiency of the rotator cuff. In addition to exercise therapy a training scheme to improve technique should be prescribed to prevent a recurrence.

When to refer: if there is Bankart lesion.

Secondary care treatment: **rotator cuff insufficiency.** None.

**Bankart lesion** Arthroscopic fixation on the anterior side of the glenoid cavity.

## Extremely Painful Immobile Shoulder

Complaint: the shoulder is extremely painful and is immobile.

Assessment: the shoulder is extremely painful.

Differential diagnosis:

**acute osteomyelitis**
**septic arthritis**

For explanatory note, supplementary assessment, primary care treatment, when to refer and secondary care treatment, see Chap. 18.

**Differential Diagnosis: Shoulder Anomalies**

| | |
|---|---|
| **Absent collarbone** | |
| Narrow chest and hanging shoulders, clavicle missing entirely or partially. | **Cleidocranialis dysostosis** |
| **Elevated shoulder blade** | |
| The shoulder blade is elevated and rotated, and there is a more or less noticeable swelling above the clavicle. | **Sprengel deformity** |
| **Repeated non traumatic shoulder subluxation/dislocation** | |
| | **Recurrent non traumatic glenohumeral joint subluxation/dislocation** |
| Associated with hypermobility disorders such as a Down and Ehlers-Danloss syndrome. | |
| | Voluntary glenohumeral joint subluxation/dislocation |
| Not associated with hypermobility disorders and non traumatic. | Habitual glenohumeral joint subluxation/dislocation |
| **Repeated traumatic shoulder subluxation/dislocation** | |
| Repeated shoulder subluxation/dislocation after a clear trauma causing the initial subluxation/dislocation. | **Recurrent posttraumatic glenohumeral joint subluxation/dislocation** |
| Forward subluxation/dislocation. | Anterior glenohumeral joint subluxation/dislocation |
| Backward subluxation/dislocation. | Posterior glenohumeral joint subluxation/dislocation |
| Foreward, backward and downward subluxation/dislocation. | Multidirectional glenohumeral joint subluxation/dislocation |

**Differential Diagnosis: Shoulder Swelling**

| **Collarbone swelling in newborns** | |
|---|---|
| A swelling in the clavicle level immediately after birth. | **Congenital pseudarthrosis of the clavicle** |
| **Swelling of the shoulder blade or collarbone or upper arm** | |
| Swelling at shoulder level. | **Benign tumor** |
| | **Malignant tumor** |

**Differential Diagnosis: Shoulder Movement Limitations in Newborns**

| **Paralysis of the arm at birth** | **Neonatal brachial plexus palsy** |
|---|---|
| Weakness of the arm. | Erb palsy (Erb-Duchenne palsy) (type I) |
| The arm is adducted and internally rotated at the level of the shoulder, the elbow is extended and the forearm lies in pronation. | |
| The wrist and fingers can be moved. | |
| The arm is abducted at the level of the shoulder, the arm is weak, the wrist is flexed and there is a claw hand. | Erb-Duchenne-Klumpke palsy (type II) |
| There is only a paralysis of the thumb and fingers. | Klumpke palsy (type III) |

**Differential Diagnosis: Shoulder Pain**

| **Vague shoulder pain** | |
|---|---|
| The nature of the pain changes, and there is mainly pain after sports. There is full mobility of the shoulder. | **Malignant bone tumor** |
| **Shoulder pain after a trivial trauma** | |
| After a trivial trauma the child complains about pain in the shoulder. Shoulder mobility is limited. | **Fractured unicameral bone cyst (juvenile-, solitary-, simple bone cyst)** |
| **Shoulder pain in overhead athletes** | |
| Pain in the shoulder after intensive use in javelin throwing, tennis or baseball. | **Little League shoulder (proximal humeral epiphysiolysis, osteochondrosis of the proximal humerus, traction apophysitis of the proximal humerus)** |
| **Painful shoulder movement trajectory** | |
| In abduction there is a painful movement trajectory. | **Insufficient rotator cuff** |
| Positive drawer and/or apprehension test. | **Bankart lesion** |
| **Extremely painful immobile shoulder** | |
| The shoulder is severely painful and is no longer mobile. | **Acute osteomyelitis** |
| | **Septic arthritis** |

# Elbow and Forearm

# 6

## Elbow Misalignment

### Crooked Elbow After a Fracture

Complaint: the parents come with the complaint that their child's elbow has become crooked after a fracture. The forearm points outwards or inwards with respect to the upper arm.

Assessment: there is a valgus or varus deformity at the elbow.

Diagnosis: **posttraumatic cubitus valgus or varus**

Explanatory note: **posttraumatic cubitus valgus or varus.** This usually occurs after a supracondylar humeral fracture. If it has insufficiently been reduced a valgus or varus deformity may occur (Fig. 6.1). In 90 % of cases of misalignment there is a varus deformity and in 10 % a valgus deformity. The valgus deformity in particular gives problems, in for instance eating. The child may have to abduct the shoulder 90° to get its hand up to its mouth.

After an inadequate reduction or redislocation of a lateral humeral condyle fracture a valgus deformity or a non-union resulting in a consequent valgus deformity and a tardy ulnar nerve palsy will develop (Fig. 6.2).

Supplementary assessment: an anteroposterior and lateral X-ray of the elbow.

Primary care treatment: a general practitioner can assess whether the deformity gives rise to functional problems.

When to refer: only in cases with functional problems.

Secondary care treatment: **posttraumatic cubitus valgus or varus.** If there are in case of a supracondylar humeral fracture severe functional problems a supracondylar correction osteotomy may be considered in order to correct the deformity.

A lateral humeral condyle fracture with a malunion less than 2 months and/or a non-union less than 6 months old may be revised for anatomic reduction. The complication

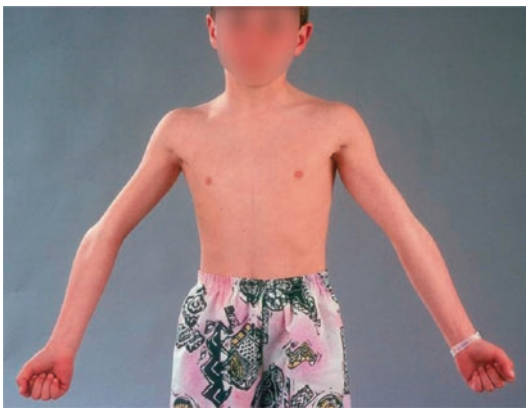


**Fig. 6.1** Varus deformity of the right elbow after a supracondylar humeral fracture


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_6

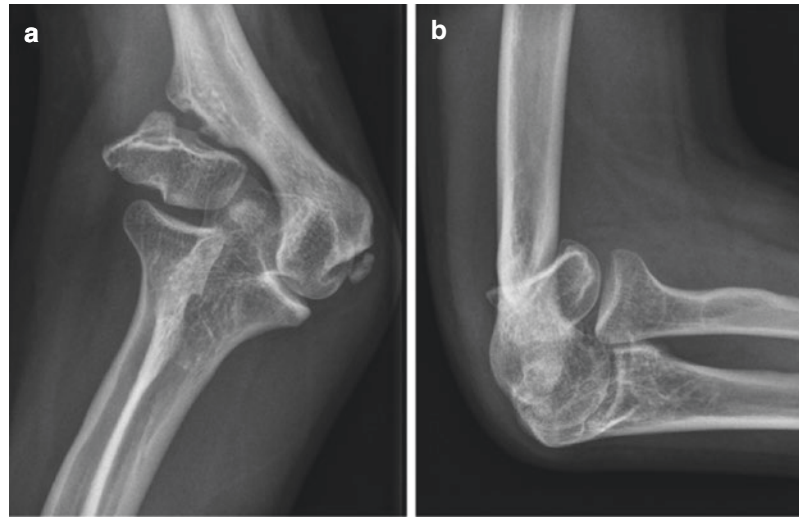



**Fig. 6.2** Anteroposterior (**a**) and lateral X-ray (**b**) of the right elbow. Non-union of the lateral humeral condyle with a valgus deformity of 40° and recurvation deformity of 50°. Situation 17 years after the injury. At time of injury the child was 4 years old

rate of these procedures of joint stiffness, ischemic necrosis and premature physeal closure is very high. In the literature the consensus is a "hands-off" policy for a fracture 3 weeks old or more. The valgus deformity may be treated by a supracondylar closing wedge osteotomy. When the cubitus valgus is due to a non-union of the lateral condyle fracture both conditions require treatment. Besides the supracondylar closing wedge osteotomy an attempt should be made to fuse the metaphyseal fragment of the lateral condyle to the metaphysis.

At the earliest signs of an ulnar nerve entrapment the ulnar head is transferred anteriorly to the medial epicondyle.

## Repeated Elbow Subluxation/ Dislocation

Complaint: the child complains that the elbow keeps shifting out of its socket with or without a preceding trauma.

Assessment: the elbow subluxation/dislocation is usually reduced when the child goes to the general practitioner. In this case, no anomalies are found.

Differential diagnosis:

**recurrent elbow subluxation/dislocation**
voluntary elbow subluxation/dislocation
habitual elbow subluxation/dislocation
recurrent posttraumatic elbow subluxation/dislocation

Explanatory note: **recurrent elbow subluxation/dislocation.** There are three types of elbow subluxation/dislocation.

*Voluntary elbow subluxation/dislocation* This type is associated with ligamentous laxity such as in Down[1] and Ehlers-Danlos syndromes[1].

*Habitual elbow subluxation/dislocation* A habitual elbow subluxation/dislocation refers to repeated subluxation/dislocation in a normal child with no ligamentous laxity or preceding significant elbow trauma.

This is a rare anomaly. In more than 90 % of cases it is a posterior or posterolateral dislocation. In 10 % of cases the anomaly is bilateral. This anomaly is more common among boys.

[1] See Appendix.

Sometimes defects develop on the lateral side of the capitellum and radial head.

*Recurrent posttraumatic elbow subluxation/dislocation* This type is a sequela from a nonunion of a medial epicondular fracture or due to residual instability from a previous dislocation. In contrast to a voluntary or habitual elbow subluxation/dislocation, the subluxations/dislocations are painful in this type.

Supplementary assessment: anteroposterior and lateral X-rays of the elbow.

Primary care treatment: in a voluntary or habitual dislocation there should be a wait-and-see policy at first, most of the time spontaneous cure occurs after 2 years after the initial subluxation/dislocation. There is also no indication for further treatment if a subluxation/dislocation occurs only once a year or even less frequently. The parents are encouraged to persuade the child to avoid dislocations.

When to refer: referral to an orthopedic surgeon after a recurrent posttraumatic subluxation/dislocation and 2 years after the initial voluntary subluxation/dislocation if this still leads to subluxation or dislocation.
An habitual elbow subluxation/dislocation does not need to be referred.

Secondary care treatment: **recurrent posttraumatic and voluntary elbow subluxation/dislocation.** Before operation one should try an elbow brace for a period of 6 months and wait-and-see if that's enough to prevent subluxation/dislocation. In case of residual instability from a previous traumatic dislocation and in a voluntary subluxation/dislocation reefing and reatachment of the capsular ligament apparatus on the posterior side for a posterior or posterolateral dislocation or on the anterior side for an anterior dislocation. Operative correction is indicated in the case of a pseudarthrosis after a medial epicondylar fracture. In exceptional cases it may be necessary to place a bone block to limit motion and prevent subluxation/dislocation.

# Elbow Swelling

## Elbow Swelling on the Back or Front or Outer Side

Complaint: there are usually no complaints. Sometimes the child complains about a "click" or stiffness. The anomaly, which is present at birth, usually goes unrecognized until the child is 3–5 years old, when the parents see a swelling around the elbow. In other cases there has been an injury.

Assessment: a swelling is visible on the posterior (Fig. 6.3), anterior (Fig. 6.4) or lateral side depending on the direction of the subluxation/dislocation. In anterior dislocations there is a limitation in flexion, whereas in a posterior subluxation/dislocation there is a limitation in extension.

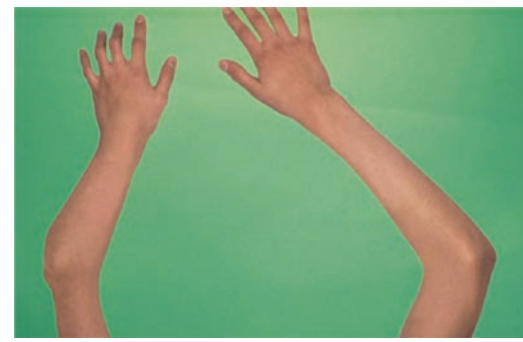


**Fig. 6.3** Bilateral congenital posterolateral radial head dislocation

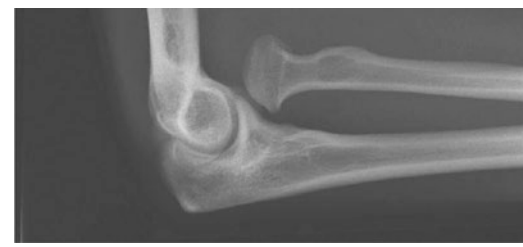


**Fig. 6.4** Congenital anterior dislocation of the radial head

Differential diagnosis:

**congenital radial head subluxation/dislocation**
**missed posttraumatic radial head subluxation/dislocation**

Explanatory note: **congenital radial head subluxation/dislocation.** The subluxation/dislocation is frequently posterior but sometimes anterior or lateral. An anterior congenital radial head subluxation/dislocation is usually associated with a syndrome such as acrocephalopolysyndactyly, acrocephalosyndactily (Apert syndrome[2]) or Cornelia de Lange syndrome[2]. Through the years the swelling increases, so the problem is easier to recognize. Sometimes movement of the forearm will not be possible as a consequence of a bony connection between the proximal part of the radius and the ulna, called a radioulnar synostosis (see p. 84).

**Missed posttraumatic radial head subluxation/dislocation** A congenital radial head subluxation/dislocation should be distinguished from a missed traumatic subluxation/dislocation. A radial head subluxation/dislocation occurs especially after a fractured ulna (Monteggia fracture) which is missed in 20–50% of cases.

Supplementary assessment: an anteroposterior and lateral X-rays of the elbow. X-rays of the entire forearm are required to determine whether there is a posttraumatic dislocation with or without an old fracture of the ulna or a radioulnar synostosis.

Primary care treatment: the general practitioner can explain to the parents that there is no point in treating a congenital radial head dislocation until the child is around 10 years of age. Operative removal of the radial head may be considered. Physiotherapy will not help this problem.

When to refer: after the age of 10 if pain, functional and/or cosmetic problems are present, or if there has been an old Monteggia fracture.

Secondary care treatment: **congenital radial head subluxation/dislocation.** Operative reduction of a congenital radial head subluxation/dislocation and reconstruction of the annular ligament is not advisable for this anomaly. The nature of the deformation of the joint and the soft parts make this nearly impossible. Only in the case of pain and/or functional and/or cosmetic problems may one consider removal of the radial head after the age of 10. However, even after such an operation it is possible that the mobility of the elbow will not increase and the function will not improve. It is preferable not to remove the radial head before 10 years of age because that causes a progressive valgus deformation in the elbow.

**Missed posttraumatic radial head subluxation/dislocation** The radial head can be reduced up to six years after the accident by means of a correction osteotomy of the ulna (Fig. 6.5), if there has been a Monteggia fracture, in which the dislocated radial head was not reduced immediately after the accident. If the radial head is still unstable, the annular ligament has to been reconstructed.

## Elbow and Forearm Movement Limitations

### Limited Extension and/or Flexion in the Elbow After a Fracture

Complaint: the elbow cannot be fully extended and/or flexed. There has been a fracture at the elbow.

Assessment: there is a limitation in extension and/or flexion at the elbow.

Diagnosis: **posttraumatic flexion and/or extension contracture of the elbow**

Explanatory note: **posttraumatic flexion and/or extension contracture of the elbow.** Most posttraumatic movement limitations

[2]See Appendix.

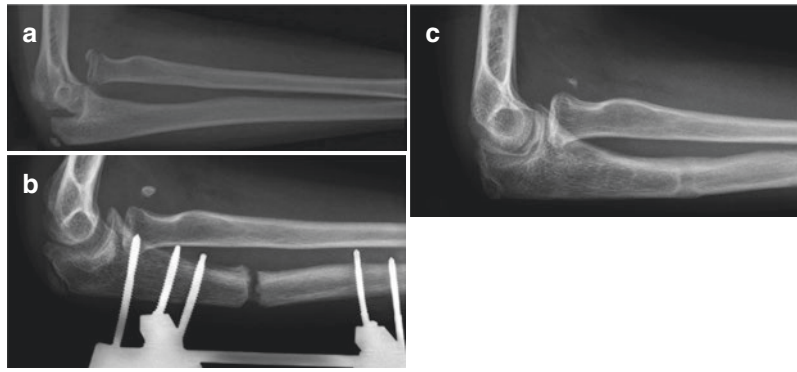



**Fig. 6.5** A radial head dislocation with a fractured ulna (Monteggia fracture) is frequently missed. (**a**) Situation after a Monteggia fracture with a healed fracture of the ulna and a missed radial head dislocation. (**b**) After a correction osteotomy of the ulna the radial head can be reduced. (**c**) Situation after the correction osteotomy is consolidated

**Fig. 6.6** Elbow swivel contracture orthosis. By using this orthosis the elbow can be passively slowly extended or flexed

are localized at the elbow. The movement limitation is mostly caused by a cast immobilization for more than five weeks which can cause the joint capsule adherence.

Supplementary assessment: anteroposterior and lateral X-rays of the elbow to rule out a loose body or other pathology.

Primary care treatment: referral to a physiotherapist can be a first step if there is joint capsule adherence.

When to refer: if physiotherapy does not result in an improvement in the extension and/or flexion limitation, or if there is an extension limitation of more than 20°.

Secondary care treatment: **posttraumatic flexion and/or extension contracture of the elbow.** An elbow swivel contracture orthosis can be prescribed (Fig. 6.6). The orthosis is worn at night and extension or flexion may slowly improve. The joint capsule can be released operatively anteriorly or posteriorly if there is no improvement.

## Forearm Movement Limitation

Complaint: the parents notice that their child cannot rotate the forearm. This tends to be noticed between the ages of 3 and 5. There has not been a trauma.

Assessment: pronation and supination of the forearm are not possible (Fig. 6.7).

Diagnosis: **congenital radioulnar synostosis**

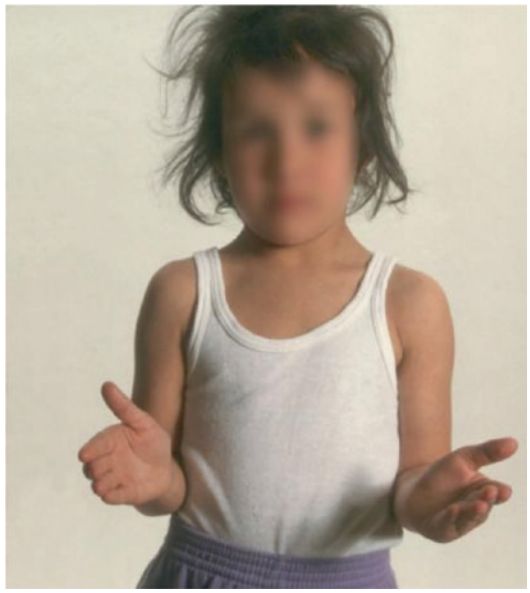


**Fig. 6.7** A radioulnar synostosis of the right forearm. Pronation and supination is not possible

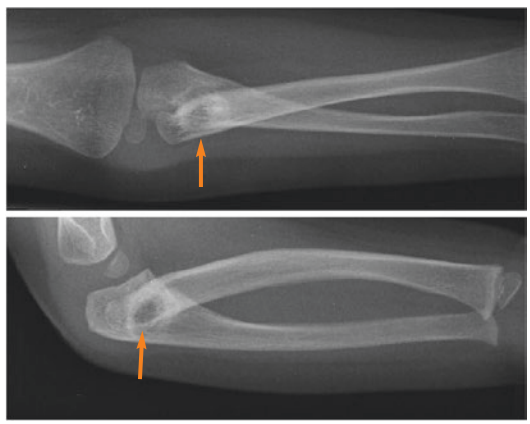


**Fig. 6.8** Radioulnar synostosis in the proximal part of the forearm with posterior dislocation of the radial head. (**a**) Anteroposterior and (**b**) lateral X-ray of the elbow and forearm (*arrows*)

Explanatory note: **congenital radioulnar synostosis.** A congenital radioulnar synostosis is a bony connection between the radius and the ulna, mostly localized in the proximal part of the forearm. It is sometimes accompanied by a dislocation of the radial head. In 60 % of cases the condition is bilateral and as common in boys as in girls. Sometimes the radioulnar synostosis is part of a syndrome such as acrocephalopolysyndactily[3] or acrocephalosyndactily (Apert syndrome)[3].

Supplementary assessment: X-rays of the elbow and forearm (Fig. 6.8).

Primary care treatment: in most cases no treatment is required. There is no point in physiotherapy.

When to refer: when functional problems occur as a result of a hyperpronation or hypersupination contracture of more than 45°.

Secondary care treatment: **congenital radioulnar synostosis.** A correction osteotomy of the forearm is performed in contractures that cause functional problems. In an unilateral synostosis the forearm is fixed in 20° pronation and in bilateral cases the dominant side is fixed in 20° pronation and the non-dominant side in 20° supination. The latter is carried out because with a hand in pronation wiping off the anus at the toilet becomes difficult. A rotation correction of the forearm can be carried out maximally over a trajectory of 45°. Further correction leads to traction on nerves and blood vessels, which can lead to severe complications. In a congenital radioulnar synostosis there is no point in operatively removing the bony connection between the radius and the ulna because there will be no improvement in mobility of the forearm.

## Elbow Pain

### Pain in the Outer Side of the Elbow After Arm Traction

Complaint: after jerking or pulling on the arm, for example to prevent a child from falling over or when lifting a child from a sitting or squatting position, a "click" may sometimes be heard or the person involved has the feeling that something is displaced. Immediately after this pain is experienced

[3] See Appendix.

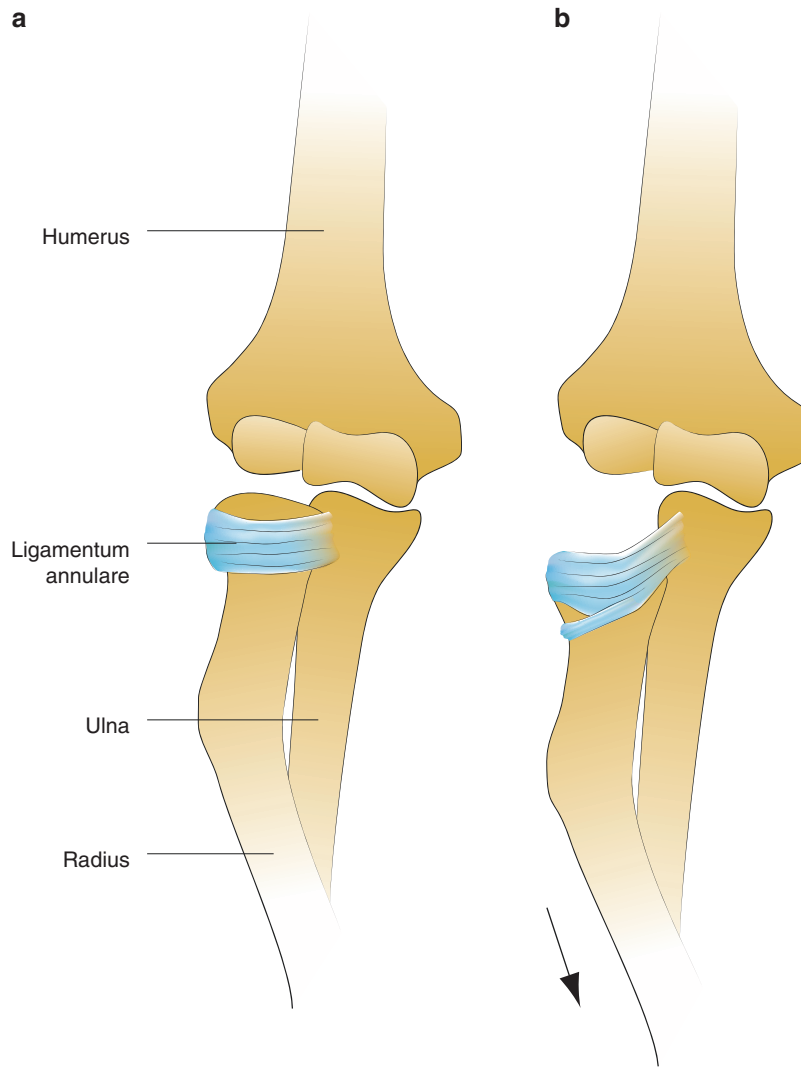



**Fig. 6.9** Mechanism of a pulled elbow. (**a**) Normal situation. (**b**) The radial head is pulled through the annular ligament due to lengthwise traction on the arm with the elbow in extension (Redrawn from: Rang M. Children's fractures. 2nd ed. Philadelphia: JB Lippencott Company; 1983. p. 193)

on the outer side of the elbow and the child refuses to use the arm.

Assessment: the forearm is always in pronation and the elbow is slightly flexed. The child does not use the arm. Flexion and extension are limited, and supination is severely limited and very painful.

Diagnosis: **pulled elbow (nursemaid elbow)**

Explanatory note: **pulled elbow.** A subluxation of the radial head that occurs suddenly after jerking or pulling on the arm is known as pulled elbow or nursemaid elbow. The annular ligament is pulled by lengthwise traction on the arm with the elbow extended (Fig. 6.9) and the elbow becomes blocked in a position of pronation. It is one of the most common injuries in children younger than 4 years of age. The problem is most common between ages of 1 and 3 years, and is almost never seen after the age of 5. A pulled elbow is more common in girls than in boys, and more common on the left side than the right side.

Supplementary assessment: not necessary.

Primary care treatment: the treatment consists of reduction the radial head by hyper-

pronating the forearm or bringing the elbow carefully into 90° of flexion, supinating the forearm and pushing the head of the radius back into position with the thumb. This is accompanied by an audible click and a feeling that something has recoiled.

When to refer: when the subluxation of the radial head has been present for more than 12 h.

Secondary care treatment: **pulled elbow.** After reduction of the radial head has been carried out, the elbow is immobilized in a cast for a week with the arm in 90° of flexion and the forearm in full supination. After a recurrent subluxation, which occurs in 5 % of cases, immobilization in a cast for two or 3 weeks after reduction is necessary.

## Pain in the Outer Side of the Elbow

Complaint: after throwing a ball the child complains about pain on the lateral side of the elbow. Sometimes the child complains that the elbow can no longer be fully stretched.

Assessment: there is pressure pain around the lateral side of the elbow. In some cases there is a limitation in extension of 10–20° and a slight limitation in flexion.

Differential diagnosis:

**Panner disease**
**osteochondritis dissecans of the capitulum**

Explanatory note: both of these osteochondroses are caused by an aseptic bone necrosis of the capitulum of the humerus as a result of a locally diminished blood supply.

**Panner disease** In children up to about age 10 the bone fragment with reduced blood supply is not demarcated; this is identified as Panner disease.

**Osteochondritis dissecans of the capitulum** This is the case with demarcation of the fragment. Osteochondritis dissecans is usually seen between the ages of 13 and 17.

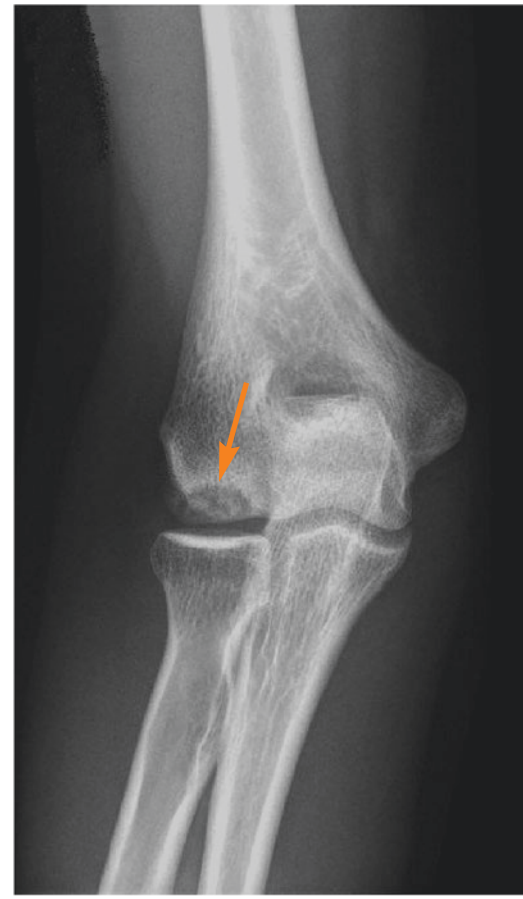


**Fig. 6.10** Osteochondritis dissecans of the capitulum (*arrow*)

Supplementary assessment: anteroposterior and lateral X-rays of the elbow (Fig. 6.10).

Primary care treatment: spontaneous healing usually occurs in children younger than 10. A ban on throwing sport activities is usually sufficient. For older children rest is usually prescribed for a period of 6–8 weeks using a collar and cuff. There is no point in giving physiotherapy.

When to refer: if radiological assessment indicates a loose body (corpus liberum).

Secondary care treatment: **Panner disease.** No treatment.

**Osteochondritis dissecans of the capitulum** Remove the corpus liberum if necessary.

## Pain in the Inner Side of the Elbow

Complaint: pain on the medial side of the elbow when exerting force. This often involves young athletes, in the USA mostly baseball players, but also volleyball and handball players, and gymnasts.

Assessment: there is pressure pain on the medial side of the elbow.

Diagnosis: **Little League elbow**

Explanatory note: **Little League elbow.** "Little League" refers to junior baseball matches in the USA. The complaints are caused by a valgus stress on the elbow during sports. As a result, distraction occurs in the structures on the medial side of the elbow such as ligaments and muscles that originate from the medial epicondyle or an epiphysiolysis of the epicondyle. There may also be pain on the lateral side of the elbow as a result of compression forces between the radial head and the capitulum, which can cause osteochondral damage.

Supplementary assessment: anteroposterior and lateral X-rays of the elbow. As a result of the valgus stress, fragmentation of the medial epicondyle or epiphysiolysis of the medial condyle can occur (Fig. 6.11). The X-rays may also show osteochondral damage of the capitulum and radial head (Fig. 6.12). It is useful to do an MRI in order to show osteochondral damage if no anomalies are found on conventional X-rays.

Primary care treatment: dosaged rest. Sports activities should be avoided for a long period of time.

When to refer: if there is an epiphysiolysis of the medial epicondyle or in the case of osteochondral damage.

Secondary care treatment: **Little League elbow.** Three weeks cast immobilization is sufficient in an epiphysiolysis of the medial epicondyle with limited distraction. Operative reduction and fixation is indicated with a dislocation of more than 2 mm.

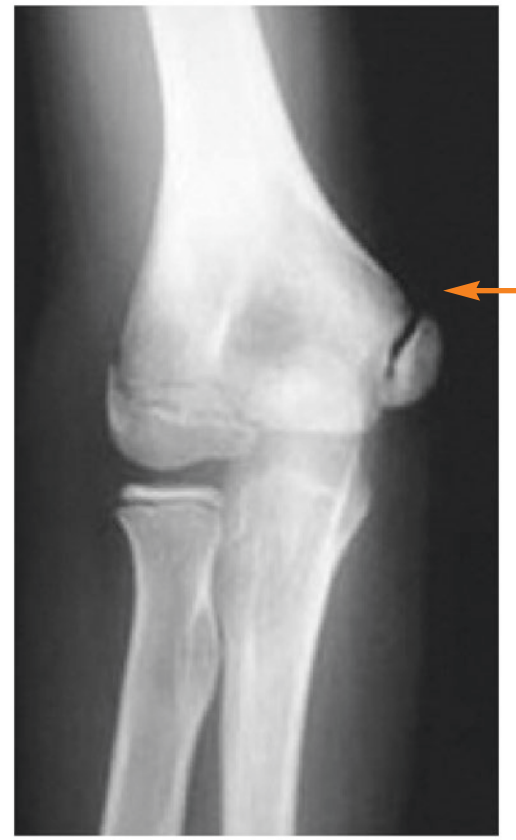


**Fig. 6.11** Epiphysiolysis of the medial epicondyle (*arrow*)

An arthroscopic removing of loose bodies damaged cartilage may be considered in the case of an osteochondral defect of the capitulum.

## Extremely Painful Immobile Elbow

Complaint: the elbow is extremely painful and is immobile.

Assessment: the elbow is extremely painful.

Differential diagnosis:

**acute osteomyelitis**
**septic arthritis**

For explanatory note, supplementary assessment, primary care treatment, when to refer and secondary care treatment, see Chap. 18.

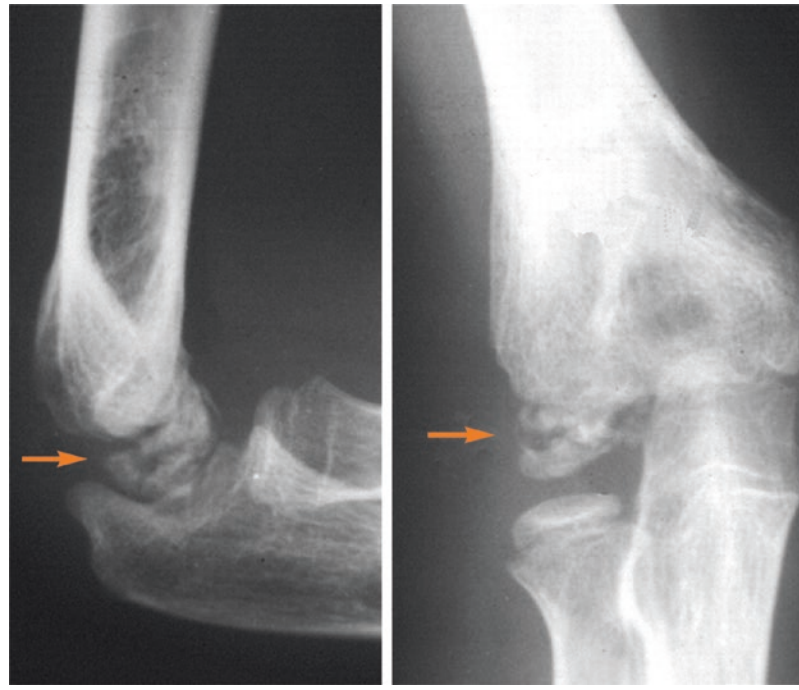


**Fig. 6.12** Osteochondral damage to the capitulum (*arrows*)

**Differential Diagnosis: Elbow Misalignment**

| | |
|---|---|
| **Crooked elbow after a fracture** | |
| There is a valgus or varus deformity of the elbow after a supracondylar humerus or lateral humeral condyle fracture. | **Posttraumatic cubitus valgus or varus** |
| **Repeated elbow subluxation/dislocation** | **Recurrent elbow subluxation/dislocation** |
| Repeated elbow subluxation/dislocation after a clear trauma causing the initial subluxation/dislocation. | Recurrent posttraumatic elbow ubluxation/ dislocation |
| Associated with hypermobility disorders. | Voluntary elbow subluxation/dislocation |
| Not associated with hypermobility disorders and non traumatic. | **Habitual elbow subluxation/dislocation** |

**Differential Diagnosis: Elbow Swelling**

| | |
|---|---|
| **Elbow swelling on the back, front or outer side** | |
| There is a swelling on the outer side of the elbow. | |
| Sometimes there is a "click" or stiffness of the elbow. | **Congenital radial head subluxation/dislocation** |
| Situation after a Monteggia fracture. | **Missed posttraumatic radial head subluxation/ dislocation** |

**Differential Diagnosis: Elbow Pain**

| | |
|---|---|
| **Pain in the outer side of the elbow after arm traction** | |
| The arm is no longer used, the forearm is held in fixed pronation. | **Pulled elbow (nursemaid elbow)** |
| **Pain in the outer side of the elbow** | |
| Pain with throwing activities, sometimes movement limitations in children of 10 years of age or younger. | **Panner disease** |
| Older than 10 years of age. | **Osteochondritis dissecans of the capitellum** |
| **Pain in the inner side of the elbow** | |
| Pain on the inner side of the elbow in athletes who throw. | **Little League elbow** |
| **Extremely painful immobile elbow** | |
| The elbow is severely painful and is no longer mobile. | **Acute osteomyelitis** |
| | **Septic arthritis** |

**Differential Diagnosis: Elbow and Forearm Movement Limitations**

| | |
|---|---|
| **Limited extension and/or flexion in the elbow after a fracture** | |
| The elbow cannot be fully extended and/or moved. | **Posttraumatic flexion contracture and/or extension contracture of the elbow** |
| **Forearm movement limitation** | |
| Pronation and supination of the forearm is not possible. | **Congenital radioulnar synostosis** |

# Wrist

# 7

## Wrist Misalignment

### Crooked Wrist

Complaint: the wrist has an abnormal position.

Assessment: there is ulnar deviation of the hand and a curvature in the radius. There are also multiple swellings of the skeleton elsewhere.

Diagnosis: **osteochondroma (exostosis).**

Explanatory note: **osteochondroma.** In 60 % of patients with hereditary multiple osteochondromata there are osteo-chondromata around the wrist (Fig. 7.1). Osteochondromata localized in the distal part of the ulna cause a shortening of the ulna and a curvature in the radius with an ulna orientated distal radial growth plate, resulting in ulnar deviation of the hand and ulnar shifting of the carpus.

Supplementary assessment: anteroposterior and lateral X-rays of the wrist.

Primary care treatment: none.

When to refer: if there are functional problems.

Secondary care treatment: **osteochondroma.** Treating this is controversial. As a rule, patients aren't bothered much by this deformation. Some orthopedic surgeons advise excision of the osteochondromata and operative lengthening of the ulna. An objection to this is that recurrences are frequent so that ulnar lengthening has to be redone. In addition, mobility of the wrist is not improved.

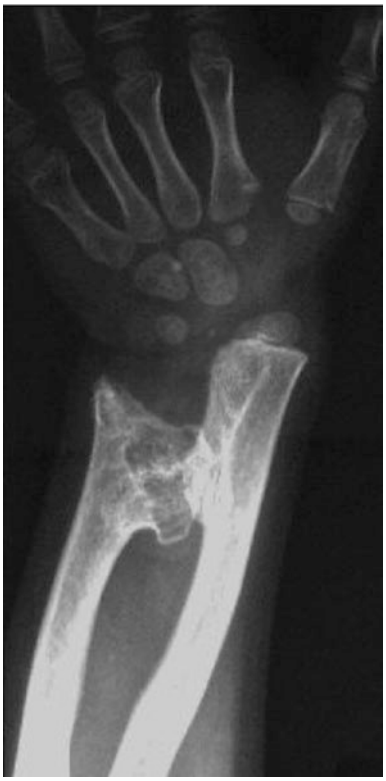


**Fig. 7.1** An osteochondroma localized in the distal part of the ulna may cause shortening of the ulna and ulnar deviation of the hand


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_7

## Wrist Swelling

### Hard Swelling on the Back of the Wrist

Complaint: there is a slow onset hard swelling on the back of the wrist.

Assessment: there is a swelling on the dorsoulnar side of the wrist.

Differential diagnosis:

**Madelung deformity**
congenital type
acquired type

Explanatory note: **Madelung deformity.** Madelung deformity is a growth disorder of the distal radial growth plate (Fig. 7.2). This growth disorder is usually congenital, but can also be caused by a trauma or infection.

*Congenital type* The congenital anomaly is twice as common bilaterally as unilaterally, and is four times more common in boys than in girls. This deformity is also seen in certain types of dwarfism (Léri-Weill disease[1]). The anomaly is seldom noticed before the age of 10, and it increases in severity from this age until growth stops. A congenital Madelung deformity presents an abnormal 5–7 mm thick fibrous structure (known as the Vicker ligament) (Fig. 7.3), that runs from the antero-ulnar part of the distal radial metaphysis to the lunatum and the triangular fibrocartilaginous complex (TFCC). The TFCC consists of an articular disc between the ulna and carpus and the ligaments between the ulnar styloid process and the ulnar carpal bones (lunate, triquetrum, hamate and the basis of the fifth metacarpal). There is disturbed growth around the antero-ulnar part of the distal radial growth plate. This causes continued growth of the posteroradial part of the distal part of the radius, resulting in palmar flexion and ulnar deviation. The ulna becomes relatively too long and this causes a posterior subluxation of the distal part, which explains the swelling on the postero-ulnar

[1] See Appendix.

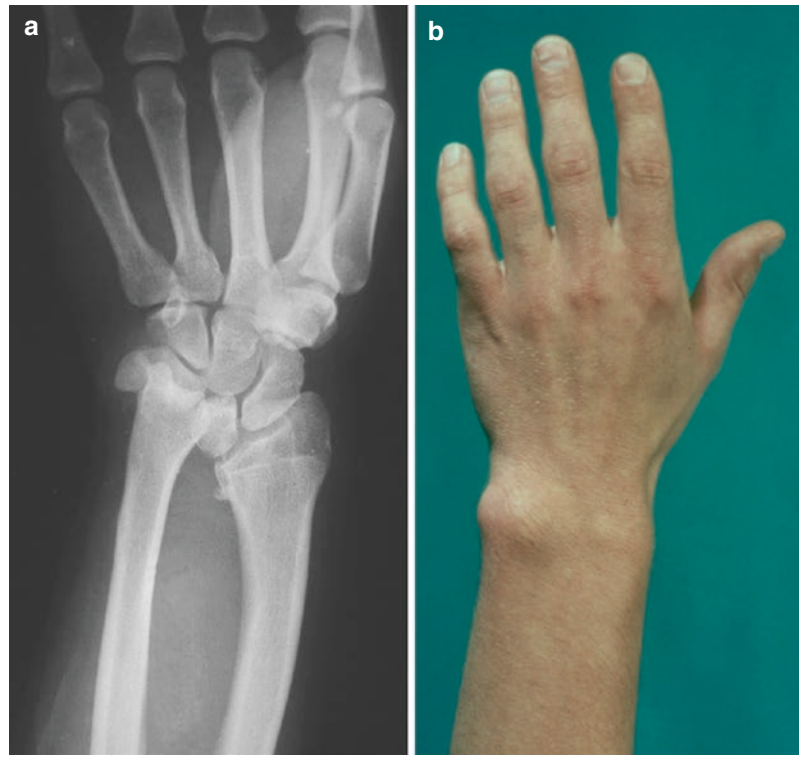



**Fig. 7.2** (**a**) Madelung deformity. In a congenital Madelung deformity there is disturbed growth around the anteroulnar part of the distal radial growth plate. This causes continued growth in the posteroradial part of the distal part of the radius, resulting in palmar flexion and ulnar deviation. (**b**) The ulna is relatively too long and the distal part is posteriorly subluxated

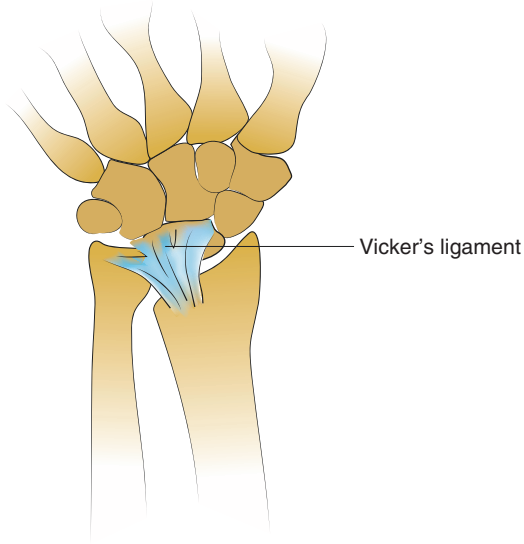



**Fig. 7.3** Vicker ligament in Madelung deformity. Vicker ligament is an abnormal 5–7 mm-thick fibrous structure that runs from the antero-ulnar part of the distal radial metaphysis to the lunate and the triangular fibrocartilaginous complex (TFCC)

side of the wrist. Assessment shows that the wrist is much broader than normal. Patients consult a physician primarily for cosmetic reasons but later on because of pain and reduced dorsal extension.

*Acquired type* The deformity resulting from a trauma or infection is caused by an osseous bridge around the ulnar part of the epiphysis in the distal part of the radius.

Supplementary assessment: anteroposterior and lateral X-rays of the wrist joint.

Primary care treatment: there tend to be few or no problems and treatment isn't usually necessary.

When to refer: when there are functional problems or if there is pain.

Secondary care treatment: **Madelung deformity.** Operative removal of the osseous bridge on the ulnar side in the distal part of the distal radial epiphyseal growth plate caused by a trauma or infection tends to produce disappointing results. Firstly, this is because removal of the peripheral bony bridge is not very successful. Secondly, children are almost grown up when the anomaly is recognized, so that after removal of the bony bridge correction due to growth can no longer be expected. It is better to do a correction osteotomy in the distal part of the radius and an osteotomy to shorten the ulna. The Vicker ligament must also be removed in the congenital type.

## Soft Swelling on the Back or Front of the Wrist

Complaint: there is an acute swelling on the back or front of the wrist.

Assessment: there is a soft swelling on the mid-posterior or anteroradial side of the wrist that can often be dispersed with pressure.

Diagnosis: **ganglion**

Explanatory note: **ganglion.** A ganglion is a cyst filled with mucus in which the microscopic image of the lining is not in continuity with that of the joint capsule or a tendon sheath. It does however make close contact with the adjacent joint or tendon sheaths. About 60 % of all the ganglia lie on the posterior side of the wrist, at the level of the scapholunate ligament next to the tendons of the extensor digitorum communis muscle. In 20 % of cases ganglia on the anteroradial side of the wrist are located between the tendons of the flexor carpi radialis and abductor pollicis longus muscles (Fig. 7.4). Ganglia are usually seen between 20 and 35 years of age, but some children can also have ganglia. At first it is a painless swelling, which can become painful later, especially during sporting activities such as tennis.

Supplementary assessment: none.

Primary care treatment: most ganglia disappear spontaneously within 1–2 years. A ganglion can be dispersed by pressure on the outside of it. In 90 % of cases this results in a

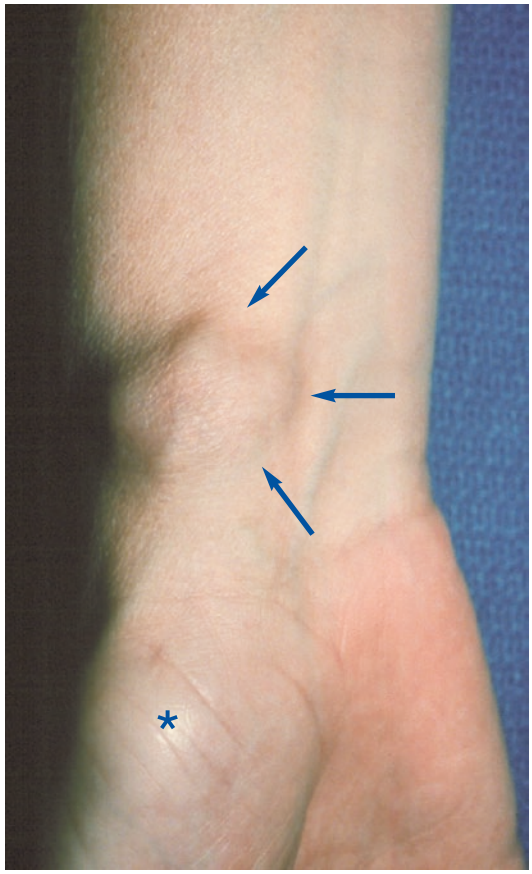


**Fig. 7.4** Ganglion on the anteroradial side of the wrist (*arrows*). * thenar

recurrence. An injection with corticosteroids results in recurrence in 60 % of cases.

When to refer: when there are functional problems.

Secondary care treatment: **ganglion.** Operative removal has a recurrence rate of 10 %. This is usually not a recurrence of the ganglion itself, but from a number of satellite ganglia which also have to be removed. If this is not done, recurrences easily occur.

## Wrist Pain

### Pain on the Back of the Wrist

Complaint: the patient complains about pain on the back of the wrist.

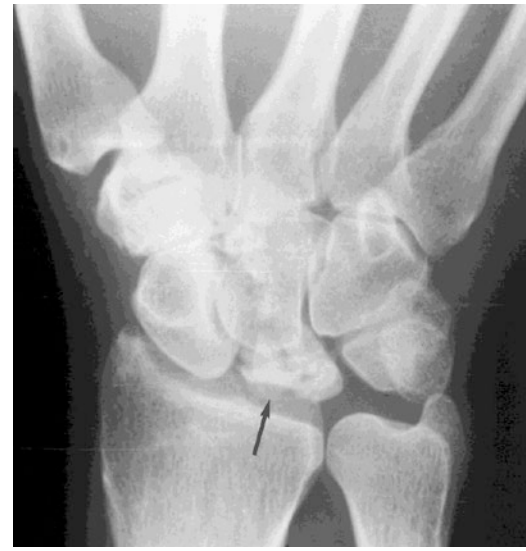


**Fig. 7.5** Kienböck disease in a young adult (*arrow*). The radius is relatively too short in this case

Assessment: there is pressure pain in the wrist mid-posteriorly.

Diagnosis: **Kienböck disease (lunatomalacia)**

Explanatory note: **Kienböck disease.** Kienböck disease is an avascular necrosis of the lunatum. This anomaly is rare in children. It usually manifests itself between the ages of 20 and 40, and is twice as common among men than among women. The probable cause is repeated trauma, often in combination with an ulna that is relatively too short. Patients complain about pain on the mid-posterior side of the wrist at rest and during movement. There is clearly decreased grip strength. X-rays show no irregularities at an early stage. Only later is a sclerosis of the lunate seen and the entire lunate may collapse much later (Fig. 7.5). Kienböck disease can be recognized at an early stage on a MRI.

Supplementary assessment: an anteroposterior and lateral X-rays of the wrist and a MRI if no abnormalities are seen on standard X-rays.

Primary care treatment: Kienböck disease can be treated with rest and analgesics at first. Symptoms may rarely persist.

When to refer: if the symptoms of Kienböck disease do not disappear with rest and analgesics.

Secondary care treatment: **Kienböck disease.** Good results can be obtained with a shortening osteotomy of the radius or a lengthening procedure for the ulna if the radius is relatively too long.

## Wrist Pain After a Trauma

Complaint: the patient complains about wrist pain after a fall.

Assessment: there may be pain on pressure around the anatomical snuffbox or in the entire wrist.

Differential diagnosis:

**missed scaphoid fracture**
**missed buckle fracture (torus fracture)**

Explanatory note: **missed scaphoid fracture.** A fracture of the scaphoid bone is the most common carpal fracture. It is seen mainly in adolescents and young men between the ages of 14 and 30, but can also occur during puberty; it's a very rare anomaly in young children. The fracture is initially missed in about 40 % of cases because the patient pays little attention to it or because the fracture was not recognised on the X-rays in the accident of emergency department. The fracture only becomes commonly visible on the X-rays 10 days after the accident (Fig. 7.6). Assessment reveals clear pressure pain around the anatomical snuffbox (Fig. 7.7).

**Missed buckle fracture** A buckle fracture is localized in the distal part of the radius, just proximal to the distal growth plate. Buckle fractures in particular tend to be missed because as a result of this fracture in which an elevation appears in the cortex no misalignment occurs (Fig. 7.8). This fracture is also called a torus fracture (torus is Latin for elevation).

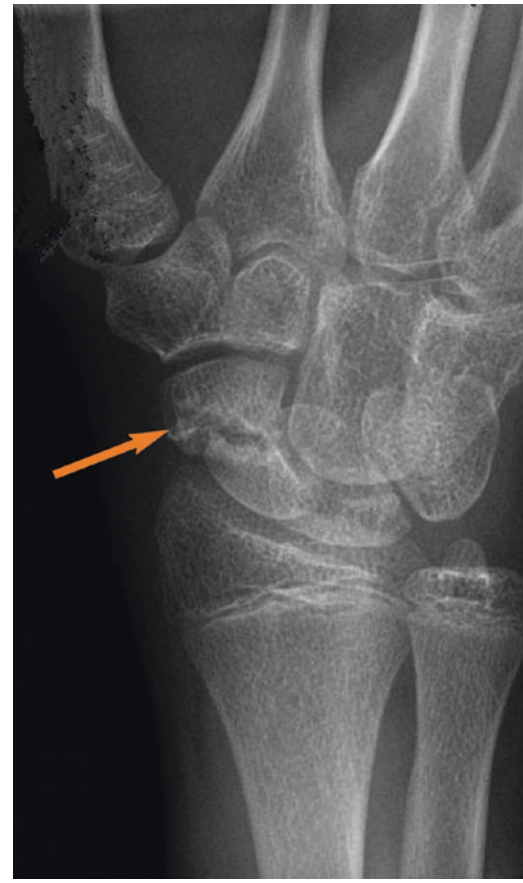


**Fig. 7.6** A fracture of the scaphoid bone can also occur during puberty. If the fracture remains untreated it can even lead to a pseudarthrosis (*arrow*)

Supplementary assessment: anteroposterior and lateral X-rays of the wrist. Scaphoid bone X-rays should be taken if a fracture of the scaphoid bone is suspected (3/4 X-rays of the wrist).

Primary care treatment: none.

When to refer: if a fracture is suspected.

Secondary care treatment: **missed scaphoid fracture.** Fractures of the scaphoid bone in children tend to heal well. A choice can be made for cast immobilization from 6 to 12 weeks even if the fracture is already several months old.

**Missed buckle fracture** It will already be consolidated if the fracture is 3 weeks old or more. Forearm cast immobilization can be given for 3 weeks if symptoms persist.

**Fig. 7.7** Anatomical snuffbox. This is a cavity on the radial side of the wrist, between the tendons of the extensor pollicis longus and extensor pollicis brevis muscles. The cavity is clearly seen when extending and abducting the thumb

## Severely Painful Immobile Wrist

Complaint: the wrist is severely painful and is immovable.

Assessment: the wrist is severely painful.

Differential diagnosis:

**acute osteomyelitis**
**septic arthritis**

For explanatory note, supplementary assessment, primary care treatment, when to refer and secondary care treatment, see Chap. 18.

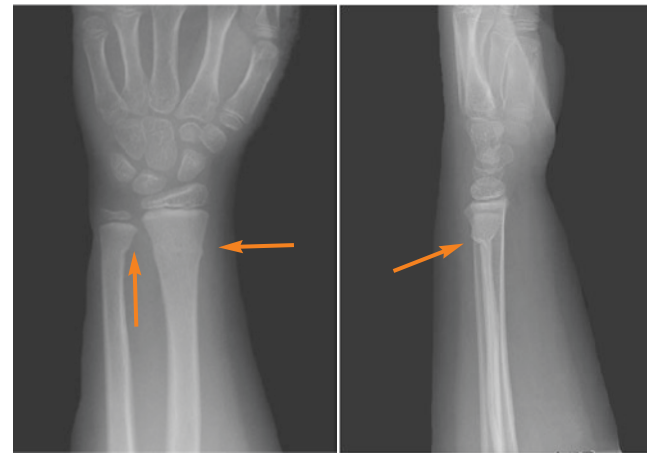


**Fig. 7.8** (**a**) Anteroposterior and (**b**) lateral X-rays of the wrist with a buckle fracture in the distal part of the radius and ulna (*arrows*)

**Differential Diagnosis: Wrist Misalignment**

| **Crooked wrist** | |
|---|---|
| Ulnar deviation of the wrist and a curvature in the radius. | **Osteochondroma (exostosis)** |

**Differential Diagnosis: Wrist Swelling**

| **Hard swelling on the back of the wrist** | |
|---|---|
| A hard swelling on the posteroulnar side. | **Madelung deformity** |
| Present from birth. | Congenital type |
| From a trauma or infection. | Acquired type |
| **Soft swelling on the back or front of the wrist** | |
| A swelling on the mid-posterior or anteroradial side that can be dispersed with pressure. | **Ganglion** |

**Differential Diagnosis: Wrist Pain**

| **Pain on the back of the wrist** | |
|---|---|
| Pressure pain in the wrist mid-posteriorly. | **Kienböck disease (lunatomalacia)** |
| **Wrist pain after a trauma** | |
| Pressure pain around the anatomical snuffbox. | **Missed scaphoid fracture** |
| The entire wrist is painful. | **Missed buckle fracture (torus fracture)** |
| **Extremely painful immobile wrist** | |
| The wrist is severely painful and is no longer mobile. | **Acute osteomyelitis** |
| | **Septic arthritis** |

# Hand

# 8

## Thumb and Finger Abnormalities

### Extra Thumb or Finger(s)

Complaint: there is an extra thumb or there are too many fingers.

Assessment: there is an extra thumb or there are more than four fingers.

Diagnosis:

**polydactyly**
- radial polydactyly (preaxial polydactyly, extra thumb)
- ulnar polydactyly (postaxial polydactyly, extra little finger)
- central polydactyly (extra finger)

**ulnar dimelia (mirror hand)**

Explanatory note: **polydactyly.** There is an extra thumb, little finger or other finger.

*Radial polydactyly* An extra thumb occurs in eight out of every 10,000 births. What is often called a double thumb is in fact a split thumb and is generally unilateral. The most accepted classification is that of Wassel (Fig. 8.1). This classification is based on the level of the split, starting at the distal phalanx. Splitting of the thumb tends to be sporadic. Only with a triphalangeal thumb may there be an autosomal dominant anomaly that may be hereditary. Other abnormalities that can be present are congenital anomalies of the spinal column, tibial hypoplasia or aplasia, cleft palate or anal atresia. A split thumb is also present in several syndromes (Table 8.1).

*Ulnar polydactyly* An extra little finger is the most common type of polydactyly (Fig. 8.2). According to the classification of Stelling and Turek there are three types: type A presents with a rudimentary little finger without phalanges. In type B there is a complete duplication of the little finger. In type C not only the little finger but also the fifth metacarpal is duplicated (Fig. 8.3). Type C is very rare.

An extra little finger is often associated with other hand anomalies such as syndactyly, triphalangeal thumb, absence of a thumb, nail anomalies and a coalition of carpal bones. These types of hand anomalies are also seen combined with radioulnar synostosis or anomalies of the lower extremity such as polydactyly of the toes, curvature of the femur, hypoplasia or aplasia of the tibia or fibula, and other irregularities of the postural and locomotor apparatus such as hemivertebrae or dwarfism. Additional anomalies may also be seen such as a hydrocephalus, harelip, dental abnormalities, ocular abnormalities, deafness and mental retardation. There may also be other anomalies such as hypogonadism, horseshoe kidney, bladder obstruction, a micro penis, anal atresia and Hirschsprung disease (large intestine). The extra little finger can also be part of a syndrome (see Table 8.2).


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_8

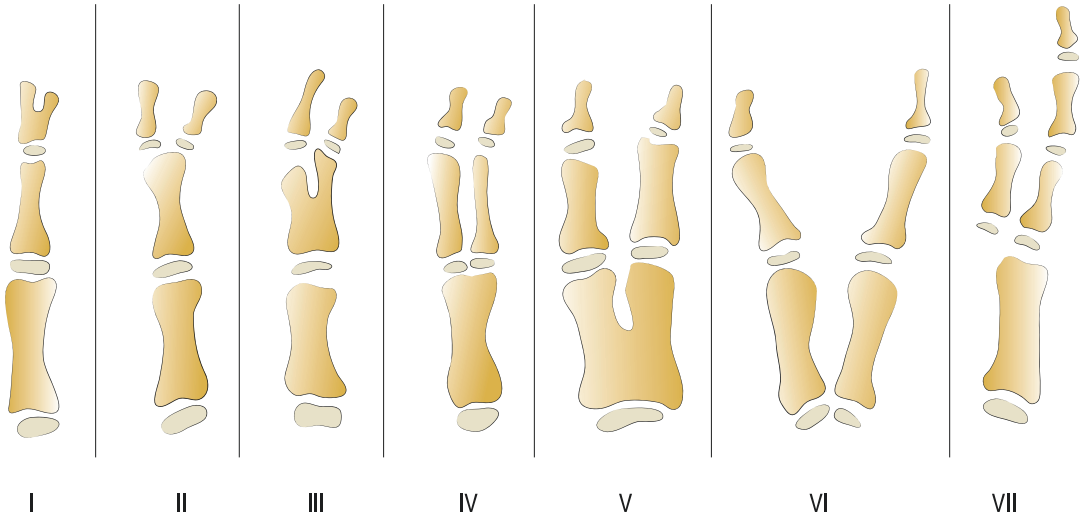



**Fig. 8.1** Wassel classification (1969) for a split thumb. The classification is based on the level of the split, starting in the distal phalanx. *Type I*: splitting of part of the distal phalanx (2%). *Type II*: splitting up to the interphalangeal joint (15%). *Type III*: splitting up to the proximal phalanx (6%). *Type IV*: complete splitting-up of the thumb (43%). *Type V*: splitting up to the metacarpal bone (10%). *Type VI*: splitting up to the carpometacarpal joint (4%). *Type VII*: complete double thumb, in which one of the split thumbs has three phalanges (triphalangeal thumb) (20%) (Redrawn from: Wassel HD. The results of surgery for polydactyly of the thumb. Clin Orthop Relat Res. 1969;64:175–93)

**Table 8.1** Syndromes with a split or extra thumb

| |
|---|
| Acrocephalopolysyndactyly (Carpenter type) |
| Fanconi anemia |
| Holt-Oram syndrome |

See Appendix for characteristics of syndromes

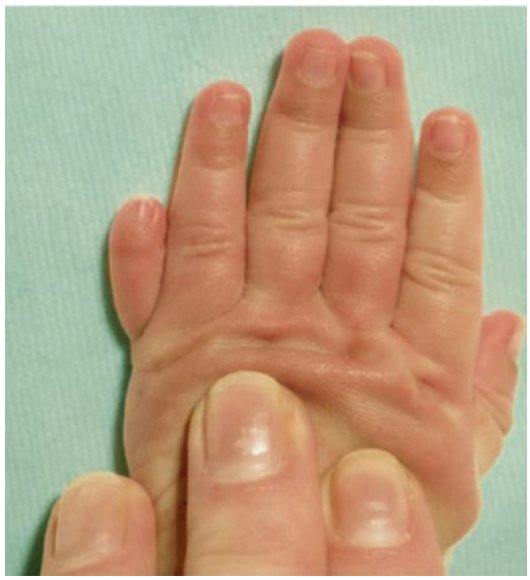


**Fig. 8.2** Extra little finger of the left hand

*Central polydactyly* In a central polydactyly there is an extra index, middle or ring finger (3.5% of all polydactylies). Tendons, nerves and blood vessels of the extra finger tend to be abnormal. The same applies to growth plates of the extra finger, often leading to ulnar or radial deviation.

**Ulnar dimelia** Ulnar dimelia is also known as mirror hand because the ulnar part of the forearm and hand are present as a mirror image. A mirror hand is a very rare and a mostly unilateral anomaly. The entire literature describes fewer than 100 cases. There is a double ulna and ulnar side of the hand, while the radius and the radial part of the hand are absent. The thumb is always absent. Both parts of a mirror hand are usually not entirely symmetrical, and seven fingers are more common than eight (Fig. 8.4). The function of the hand is very limited. The elbow and the wrist are usually widened, and the forearm and upper arm tend to be shorter. There may be irregularities in the shoulder blade, clavicle, humerus and shoulder joint. The capitulum is absent and the trochlea is duplicated in the elbow. The elbow is generally stiff. The biceps and brachialis muscles

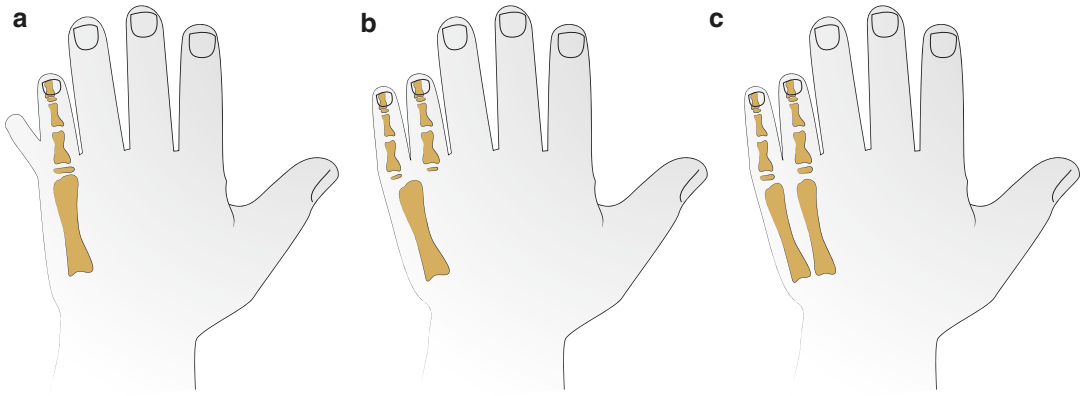



**Fig. 8.3** According to the classification of Stelling (1963) and Turek (1967), there are three types of extra little fingers. In *type A* there is a rudimentary little finger without phalanges, in *type B* there is a completely duplicated little finger, and in *type C* not only the little finger but also a duplicated fifth metacarpal bone (Redrawn from: Stelling F. The upper extremity. In: Fergusion AB, editor. Orthopedic surgery in infancy and childhood. Williams & Wilkins; 1963. p. 304–8; Turek SL. JB Lippincott, Philadelphia. 1967;123)

**Table 8.2** Syndromes with an extra little finger

| |
|---|
| Biemond II syndrome |
| Ellis-van Creveld syndrome |
| Goltz syndrome |
| Jeune syndrome |
| Laurence-Moon-Biedl-Bardet syndrome |
| Orofaciodigital syndrome type I |
| Orofaciodigital syndrome type II |
| Trisomy 13 |

See Appendix for characteristics of syndromes

do not insert into the forearm but into the distal part of the humerus. The musculature and vascular systems of the forearm and hand are also abnormal. Another irregularity that often occurs on its own is fibular dimelia, with a duplication of the fibula and outer toes and absence of the tibia and the tibial part of the foot and the big toe and second toes.

Supplementary assessment: if a mirror hand is suspected, X-rays of the hand, elbow, forearm and wrist (Fig. 8.4).

Primary care treatment: none. Occasionally the general practitioner would sometimes tie a tight string around the base of a rudimentary little finger causing necrosis. This is no longer carried out because of the risk of neuroma formation, but also because it is often difficult for parents to accept this situation. One should be aware of other possible additional congenital anomalies.

When to refer: referral to a hand surgeon between the first and second years of life.

Secondary care treatment: surgery is recommended between 9 and 15 months of age.

**Radial polydactyly** One part of the split thumb tends to be clearly larger than the other part. In such cases, the smallest part is excised (Fig. 8.5). In types I, II and III (Fig. 8.1) the central part is resected and the remaining parts are coapted (Bilhaut-Cloquet procedure). Sometimes it makes sense to limit resection of the central part to the soft parts and nails (Fig. 8.6).

**Ulnar polydactyly** Type A: resection of the rudiment. Type B: ablation of the extra little finger. Type C: resection of the extra little finger and metacarpal.

**Central polydactyly** Resection of the extra finger. This is technically very difficult and it is a tall order to obtain an acceptable appearance and function in the hand in such cases.

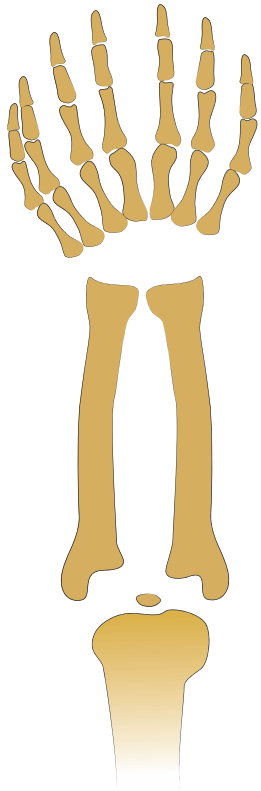


**Fig. 8.4** Ulnar dimelia or mirror hand. This is a duplication of the ulna and the ulnar side of the hand, with an absent radius (starting at the elbow) and radial part of the hand. The thumb is always absent. Both parts of a mirror hand tend not to be entirely symmetrical, and seven fingers are more common than eight (Redrawn from: Green DP. Operative hand surgery. 2nd ed. Churchill Livingstone. New York, Edingburgh, London, Melbourne; 1988)

**Ulnar dimelia** This is extremely difficult, if not impossible to treat, and the procedure can only be performed by a highly experienced hand surgeon specialized in congenital hand abnormalities.

## Gigant Growth of Thumb and/or Finger(s)

Complaint: immediately after birth it is noticeable that the thumb and/or one or several fingers are much larger than normal.

Assessment: overgrowth of the thumb and/or fingers (Fig. 8.7). There is normal sensation. The interphalangeal and metacarpophalangeal joints have normal mobility in children.

Differential diagnosis:

**macrodactyly**
non progressive type
progressive type

Explanatory note: **macrodactyly.** This is a rare, non-hereditary anomaly that accounts for 0.9 % of all congenital hand abnormalities. In 10 % of cases there is a syndactyly. The cause is unknown. The joints of the affected thumb or finger may become stiff during adulthood. There are two types of macrodactyly: non-progressive and progressive.

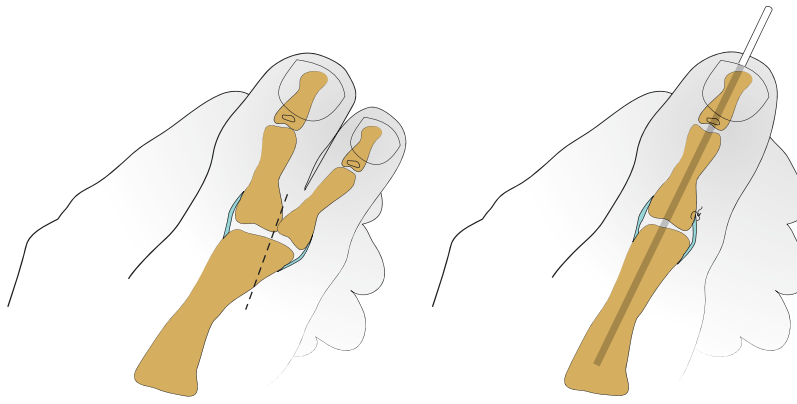


**Fig. 8.5** Treatment of extra (*left*) thumb. Most of the time one part of the split thumb is clearly larger than the other. In such cases, the smallest part is removed. In this particular case, with a completely split thumb, a part of the radial side of the metacarpal head has to be resected, and to prevent instability reconstruction of the radial ligament has to be carried out (Redrawn from: Marks TW, Bayne LG. Polydactyly of the thumb: abnormal anatomy and treatment. J Hand Surg Am. 1978;3:107–16)

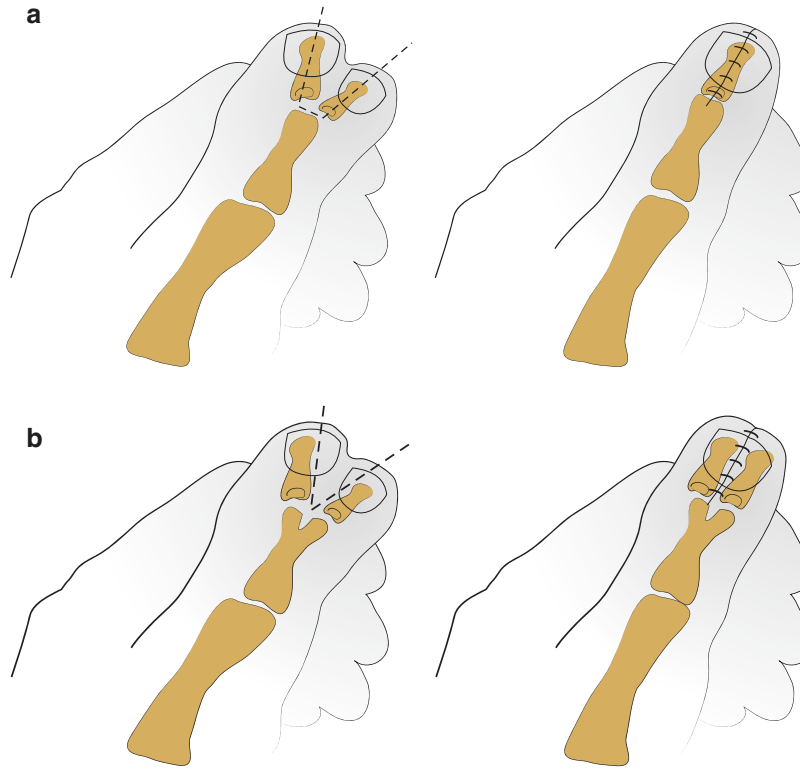



**Fig. 8.6** (**a**) The central part can be resected (Bilhaut-Cloquet procedure) if the splitting is equally large in types I, II and III. (**b**) Sometimes it makes sense to limit resection of the central part to the soft tissue and nails (Redrawn from: Marks TW, Bayne LG. Polydactyly of the thumb: abnormal anatomy and treatment. J Hand Surg Am. 1978;3:107–16)

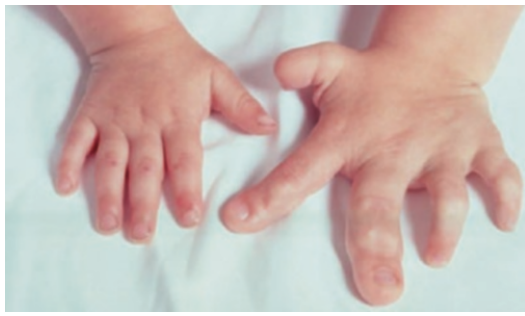


**Fig. 8.7** Macrodactyly of the left hand

*Non progressive type* During growth the thumb and/or finger(s) remain proportional to the normal digits.

*Progressive type* The macrodactyly becomes disproportionately larger than the rest of the hand during childhood.

Supplementary assessment: hand X-rays.

Primary care treatment: none.

When to refer: the parents usually want advice with referral to a specialist (pediatric plastic surgeon or hand surgeon).

Secondary care treatment: **macrodactyly.** Removal of the excess soft parts on one side of the affected thumb or finger(s), and 3 months later on the other side. Simultaneously, all the growth plates of the affected thumb or finger(s) are curetted. The operation is carried out when the thumb and/or finger(s) have reached adult length. It may be helpful to compare the situation with the thumb or finger(s) of the parent of the same sex. There are also shortening methods with segmental resection of the nerves that are then microscopically sutured end-to-end if the above technique is insufficient. The most common complication is recurrent macrodactyly of the thumb or finger(s) and necrosis of the skin. In some cases the entire affected finger and metacarpal bone have to be resected.

## Finger Misalignment

Complaint: crooked or bent finger.

Assessment: crooked or bent finger.

Differential diagnosis:

**camptodactyly**
**Kirner deformity**
**trigger finger**
**clinodactyly**
**delta phalanx**

Explanatory note: **camptodactyly.** This is a congenital flexion contracture of the proximal interphalangeal joint (Fig. 8.8). It affects 1–2 % of the population, usually the little finger. The anomaly is bilateral in two-thirds of cases. When it is unilateral it tends to affect the right hand. There are three types of camptodactyly. Type I: is seen in infancy. Type II: is seen in adolescence. Type III: occurs as part of an underlying syndrome such as the Russel-Silver syndrome.[1]

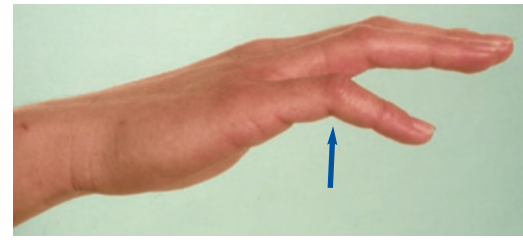


**Fig. 8.8** Camptodactyly. This is a congenital flexion contracture of the proximal interphalangeal joint, usually of the little finger (*arrow*)

**Kirner deformity** This is a rare, progressive palmar and radial curvature of the distal phalanx usually involving the little finger (Fig. 8.9). The cause is unknown. It typically affects preadolescent females, usually bilaterally. This anomaly doesn't usually cause many problems, and treatment is generally not required.

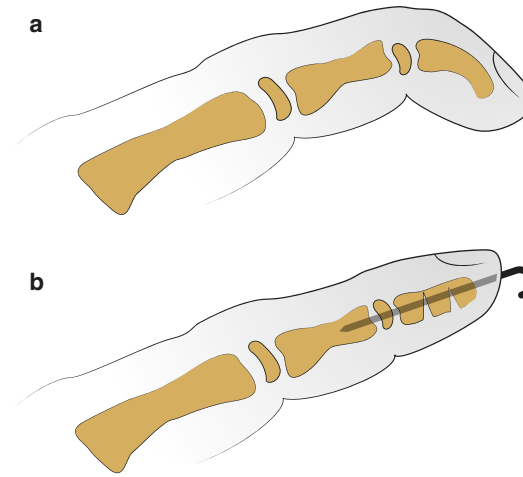



**Fig. 8.9** (**a**) Kirner deformity. This is a progressive, palmar and radial curvature of the distal phalanx of the little finger. (**b**) It is only in rare cases that multiple osteotomies are necessary to correct it (Redrawn from: Carstam N, Eiken O. Kirschner's deformity of the little finger. J Bone Joint Surg. 1970;52A:1663–5)

**Trigger finger** In a trigger finger there is snapping or locking of the digit at the A1 pulley. A trigger finger is usually due to a congenital anomaly of the flexor mechanism.

**Clinodactyly** This is a radial deviation of the distal phalanx at the level of the distal interphalangeal joint. It may affect any finger, but in most cases it involves the little finger (Fig. 8.10), and is frequently bilateral. The anomaly affects between 1 and 10 % of the population.

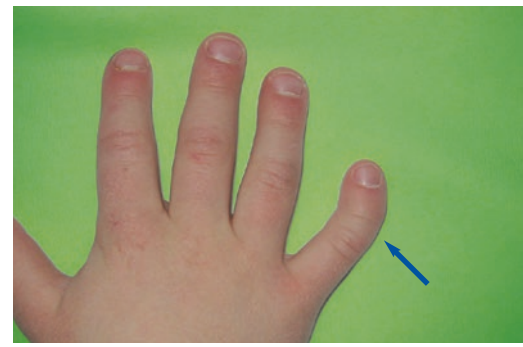


**Fig. 8.10** Clinodactyly of the little finger (*arrow*). This is a radial deviation at the level of the distal interphalangeal joint. It can affect any finger, but in most cases the little finger is involved. This anomaly is often bilateral

[1] See Appendix.

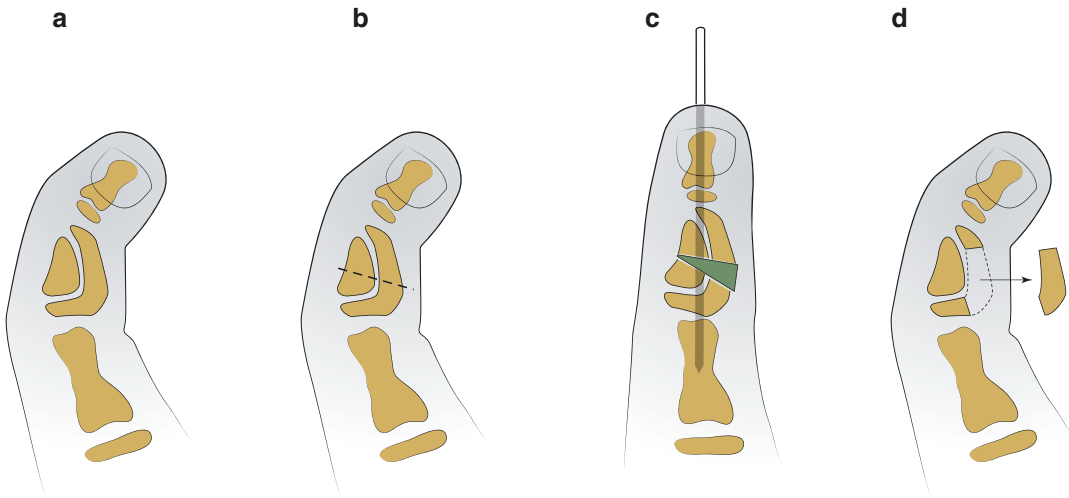



**Fig. 8.11** (**a**) Delta phalanx. This is an abnormal triangular form, mostly of the middle phalanx of one of the fingers, causing radial or ulnar deviation. (**b**, **c**) Opening wedge correction osteotomy of a delta phalanx. (**d**) Removal of the central part of the longitudinal epiphyseal bracket. The defect is filled with fat (Redrawn from: Wood VE, Flatt AE. Congenital triangular bones in the hand. J Hand Surg. 1977;2:179–93)

**Delta phalanx** There is an abnormal triangular delta shape of one of the fingers, mostly involving the middle phalanx. A radial or ulnar deviation occurs due to the longitudinal epiphyseal bracket (Fig. 8.11). This is a rare anomaly.

Supplementary assessment: X-rays of the involved finger if necessary.

Primary care treatment: if there is a camptodactyly, Kirner deformity or delta phalanx with a mild deformity a wait-and-see policy can be adopted. The same applies for clinodactyly when there are almost no functional problems.

When to refer: if there is trigger finger and if a crooked finger causes a functional problem.

Secondary care treatment: **camptodactyly.** The deformity can generally be corrected passively using a splint if this is done early enough.

**Kirner deformity** If the anomaly is painful, temporary immobilization with a splint can be considered. In severe cases one or several osteotomies are necessary to correct the abnormality (Fig. 8.9).

**Trigger finger** Requires operative treatment. A release of A1 pulley alone may not be sufficient — there is a recurrence rate of up to 50 %.

**Clinodactyly** None.

**Delta phalanx** A correction osteotomy can be performed or the central part of the longitudinal epiphyseal bracket can be removed in a radial or ulnar deviation of a finger with a delta phalanx. The defect is filled with fat. The misalignment and length of the phalanx will correct spontaneously after this (Fig. 8.11).

## Short Finger

Complaint: one or several fingers are too short.

Assessment: one or several fingers are too short.

Diagnosis: **brachydactyly**

Explanatory note: **brachydactyly.** This is a shortening of the finger phalanges or one or several metacarpal bones, usually involving the ring finger or little finger (Fig. 8.12). This is generally an autosomal hereditary problem, but may also be part of a syndrome, such as Poland syndrome and Cornelia de Lange syndrome[2].

Supplementary assessment: if necessary X-rays of the hand.

Primary care treatment: none.

When to refer: it is usually a cosmetic problem, and is rarely a functional problem. One can refer the patient if the latter is the case.

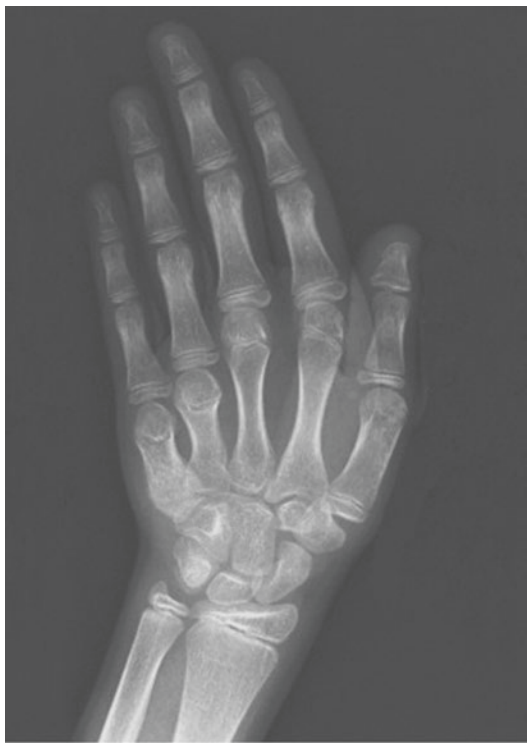


**Fig. 8.12** Brachydactyly of the fourth and fifth metacarpal bones

[2]See Appendix.

Secondary care treatment: **brachydactyly.** Lengthening the affected metacarpal bone. The metacarpal can be lengthened 10 mm in one operation, or up to 30 mm with gradual distraction using external fixation.

## Fused Fingers and/or Thumb

Complaint: two or more fingers are attached to each other.

Assessment: two or more fingers are attached to each other.

Differential diagnosis:

**syndactyly**
simple type
complex type

Explanatory note: **syndactyly**

*Simple type* Only the soft parts are involved. The syndactyly can be incomplete, in which case the fingers are fused at the base but not distally (Fig. 8.13) or complete, in which case the fingers are fused over the whole length (Fig. 8.14).

*Complex type* Even some bones are fused in a complex syndactyly (Fig. 8.15).

Syndactyly is often an isolated abnormality, but can be a part of many syndromes (Table 8.3) (Fig. 8.16). The incidence of syndactyly is about 1 in 2000 live births. This anomaly is 10 times more common among white children than among children of African descent, and it also affects boys twice as often as girls; bilateral and unilateral frequency are the same. Fusion of the middle and ring fingers is the most frequent. Sometimes there is also an extra digit.

Supplementary assessment: hand X-rays.

Primary care treatment: none.

When to refer: as quickly as possible if the thumb is involved.

Secondary care treatment: **syndactyly.** A syndactyly between the thumb and index finger should be treated before the age of 6 months because otherwise the index finger will grow crooked. Separation of a syndactyly between the ring finger and little finger must take place before 1 year of age for the same reason. In other cases the operation is usually postponed until 2 or 3 years of age because the fingers are then larger and the operation is easier to perform.

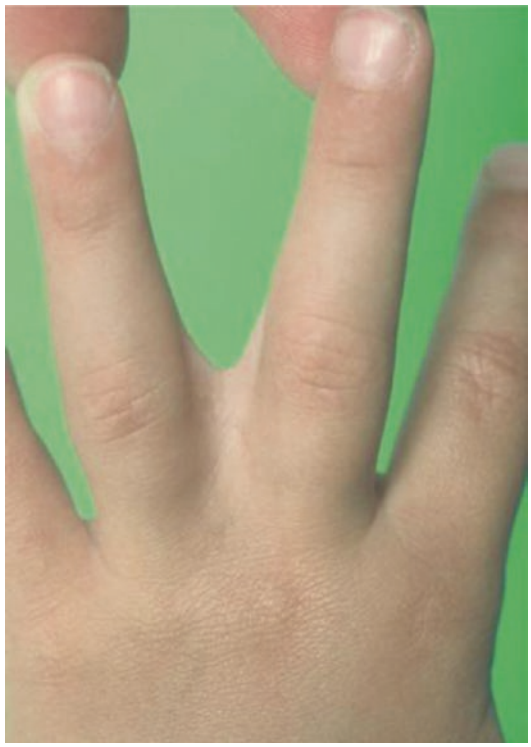


**Fig. 8.13** Incomplete syndactyly

## Thumb Misalignment

Complaint: thumb curvature.

Assessment: there is a slightly flexed position around the interphalangeal joint (Fig. 8.17) or excessive flexion at the metacarpophalangeal I joint, which forces the thumb into the palm of the hand (Fig. 8.18).

Differential diagnosis:

**trigger thumb**
**congenital clasped thumb**

Explanatory note: **trigger thumb.** Passive extension of the interphalangeal joint is not possible or can only occur by using some force, which causes a snapping sensation. A swelling may be observed on the palmar side, at the base of the thumb. The flexor pollicis longus tendon has difficulty in gliding

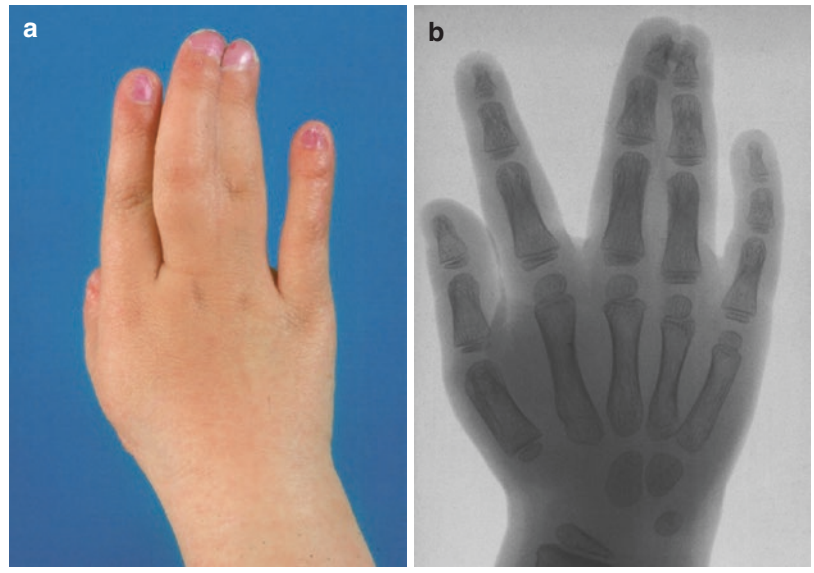



**Fig. 8.14** (**a**) Complete syndactyly of middle and ring finger. (**b**) The X-ray shows that this is a simple type in which only the soft tissues are involved

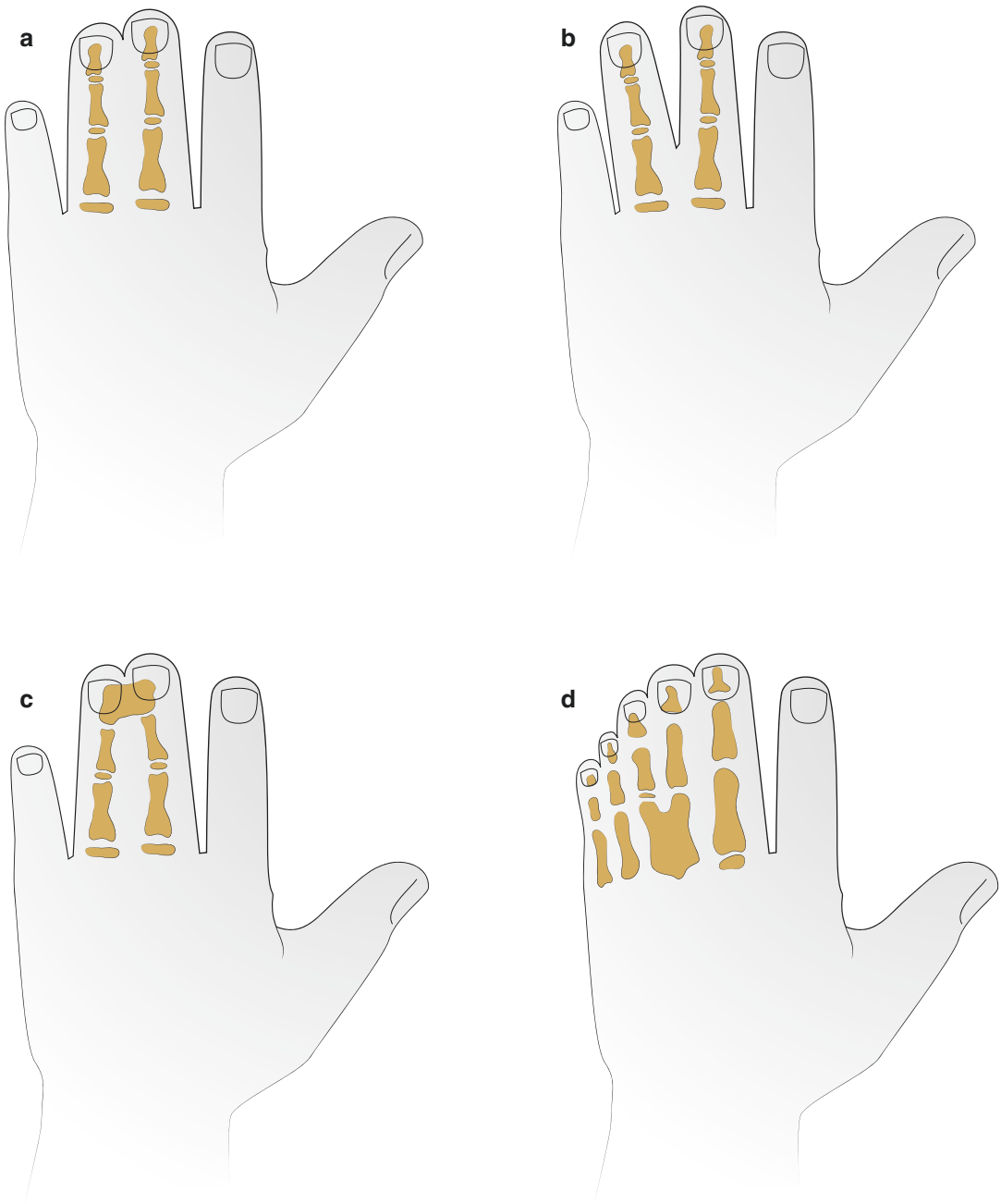



**Fig. 8.15** Syndactyly. (**a**) Simple complete syndactyly of the middle and ring fingers in which the soft tissues are fused over the entire length. (**b**) Simple incomplete syndactyly in which the soft tissues are fused together at the base, but not distally. (**c**, **d**) Complex forms of syndactyly in which the bones of different phalanges have also fused together (Redrawn from: Green DP. Operative hand surgery. 2nd edition. Churchill Livingstone; 1988. p. 347)

**Table 8.3** Syndromes with syndactyly of the fingers

| |
|---|
| Acrocephalopolysyndactyly |
| Acrocephalosyndactyly (Apert syndrome) |
| Biemond II syndrome |
| Laurence-Moon-Biedl-Bardet syndrome |
| Orofaciodigital syndrome type I |
| Orofaciodigital syndrome type II |
| Poland syndrome |
| Rothmund-Thomson syndrome |
| Rubinstein-Taybi syndrome |
| Trisomy 13 |
| Trisomy 18 |

See Appendix for characteristics of syndromes

through the tendon sheath at the level of the proximal annular ligament (A1 pulley). Because of this impediment the tendon thickens, which makes it increasingly difficult to glide through the tendon sheath. The anomaly develops during the first 2 years of life. In 25 % of cases the anomaly is bilateral.

**Congenital clasped thumb** The extensor pollicis brevis muscle is underdeveloped or absent. The extensor pollicis longus muscle may also be absent. The thumb looks normal during the first 3 months after birth. The diagnosis is usually made after 3 months of age. A flexion contracture at the metacarpophalangeal I joint develops at this age.

Supplementary assessment: none.

Primary care treatment: none.

When to refer: trigger thumb: before 3 years of age. Congenital clasped thumb: as soon as possible.

Secondary care treatment: **trigger thumb.** A thumb that can still be manually extended can be splinted for 6 weeks in an extended position. If this doesn't help, or in the case of a fixed flexion of the interphalangeal joint, the proximal annular ligament (A1 pulley) can be released in children older than 1 year of age.

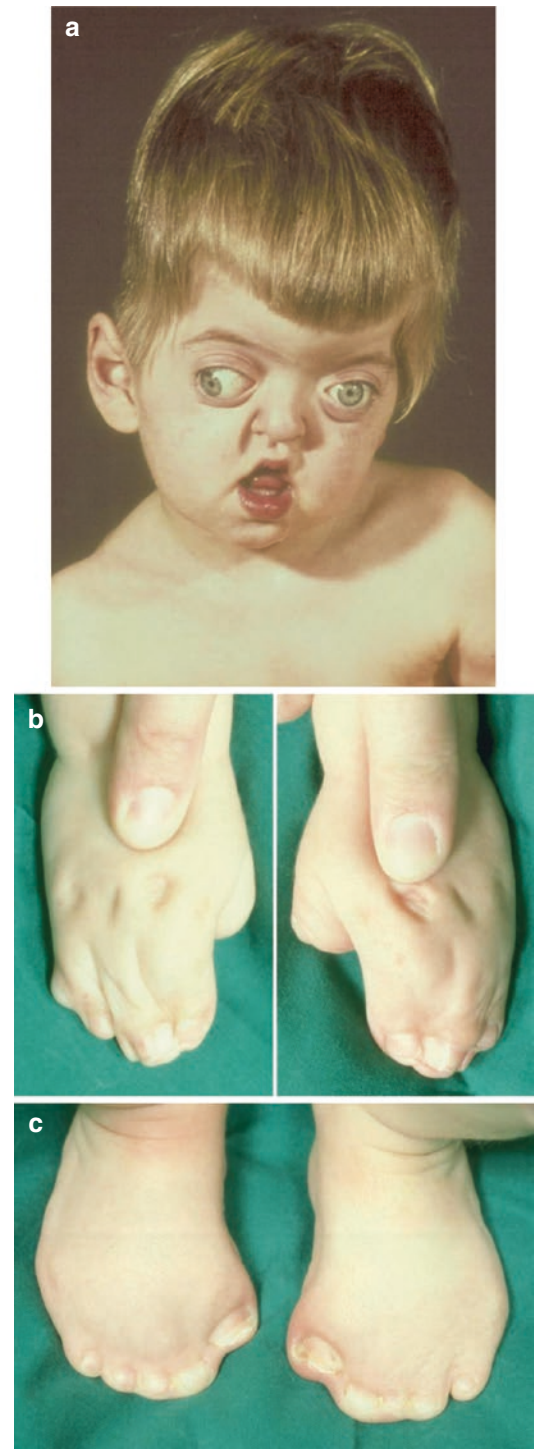



**Fig. 8.16** (**a**) Acrocephalosyndactyly (Apert syndrome) in which there is a syndactyly of both hands (**b**) and both feet (**c**) (see appendix)

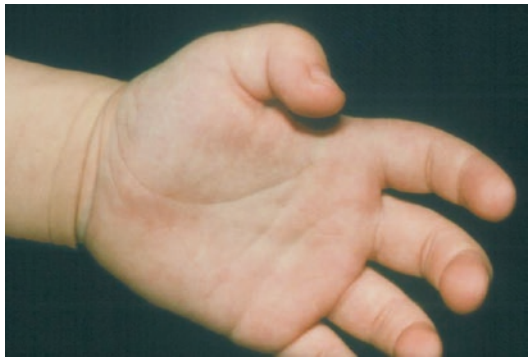


**Fig. 8.17** Trigger thumb with a flexed interphalangeal joint

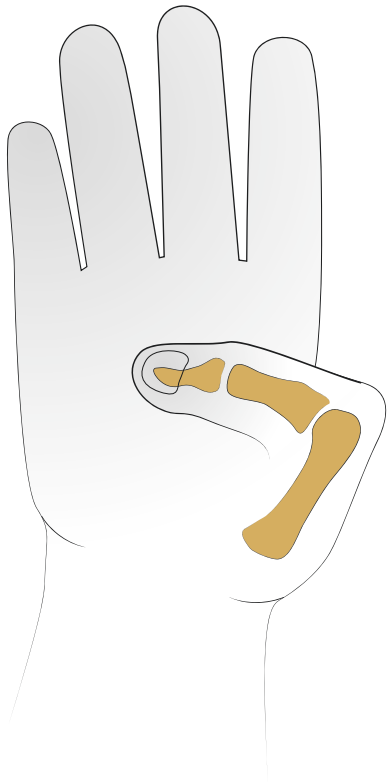


**Fig. 8.18** Congenital clasped thumb with excessive flexion of the metacarpophalangeal I joint in which the thumb lies in the palm of the hand

**Congenital clasped thumb** Splintage or serial casting between 3 and 6 months of age is usually sufficient. Extension of the thumb must be restored if splintage or serial casting doesn't help. A tendon transfer of the extensor indicis proprius or the flexor digitorum superficialis muscles of the ring finger is generally carried out in order to achieve this.

## Long Thumb

Complaint: the thumb is longer than normal.

Assessment: the thumb is longer than normal and has three joints. The distal phalanx can have an abnormal posture. The thumb may even look like a finger (Fig. 8.19).

Differential diagnosis:

**triphalangeal thumb**
type I
type II
type III

Explanatory note: **triphalangeal thumb.** In a triphalangeal thumb the thumb has three instead of two phalanges. There are three types:

*Type I* The middle phalanx has a delta or other shape.

*Type II* The thumb has three phalanges but not withstanding this the function is more or less normal. Opposition is possible but a pinch grip is less refined than in a normal thumb.

*Type III* In this case the thumb is actually a fifth finger. We call this a five-fingered hand.

A triphalangeal thumb is often an autosomal dominant hereditary condition. Other anomalies that can accompany this are polydactyly, cleft hand, cleft foot and tibial hypoplasia or aplasia. This can also be part of the Holt-Oram syndrome[3] or a Fanconi anemia[3].

Supplementary assessment: anteroposterior and lateral X-rays of the hand.

Primary care treatment: none.

[3] See Appendix.

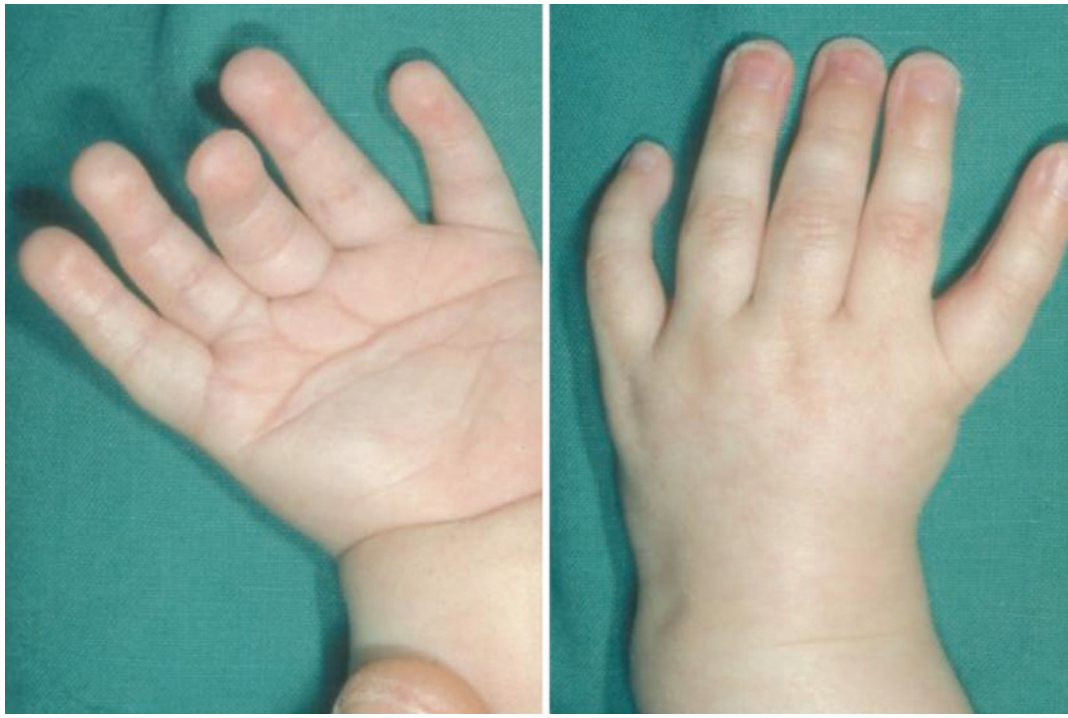


**Fig. 8.19** Five-fingered hand

When to refer: refer to a pediatric plastic surgeon or hand surgeon, preferably before 1 year of age.

Secondary care treatment: **triphalangeal thumb.**

*Type I* Resection of the middle phalanx. This must be carried out before the child's first birthday. The extensor pollicis longus and flexor pollicis longus tendons have to be shortened. The thumb is stabilized with a Kirschner wire for 6 weeks, then immobilized with a thumb splint for 2 months.

If resection of the middle phalanx is performed at an older age an ankylosis of the neo-interphalangeal joint of the thumb will occur. A resection arthrodesis of the distal interphalangeal joint may be considered as an alternative. Such an operation must be postponed for as long as possible because a growth disorder in the future distal phalanx can develop. A Z-plasty may be required in the case of contractures of the skin between thumb and index finger.

*Type II* Patients indicate that there isn't a major functional problem with this thumb, but it is cosmetically unattractive. About half of these patients have a contracture of the skin between the thumb and index finger which limits grasping large objects. A Z-plasty of the skin between thumb and index finger can increase the grasp function. The middle phalanx can also be operatively removed, or a resection arthrodesis of the distal interphalangeal joint may be carried out.

*Type III* Pollicization of the most radial localized finger may be carried out if required.

## Short Thumb

Complaint: the thumb is too short or is absent.

Assessment: the thumb is too short but has good mobility. There may be abnormal adduction or abduction. There may be a flaccid thumb that cannot be actively moved (Fig. 8.20), or the thumb may be absent.

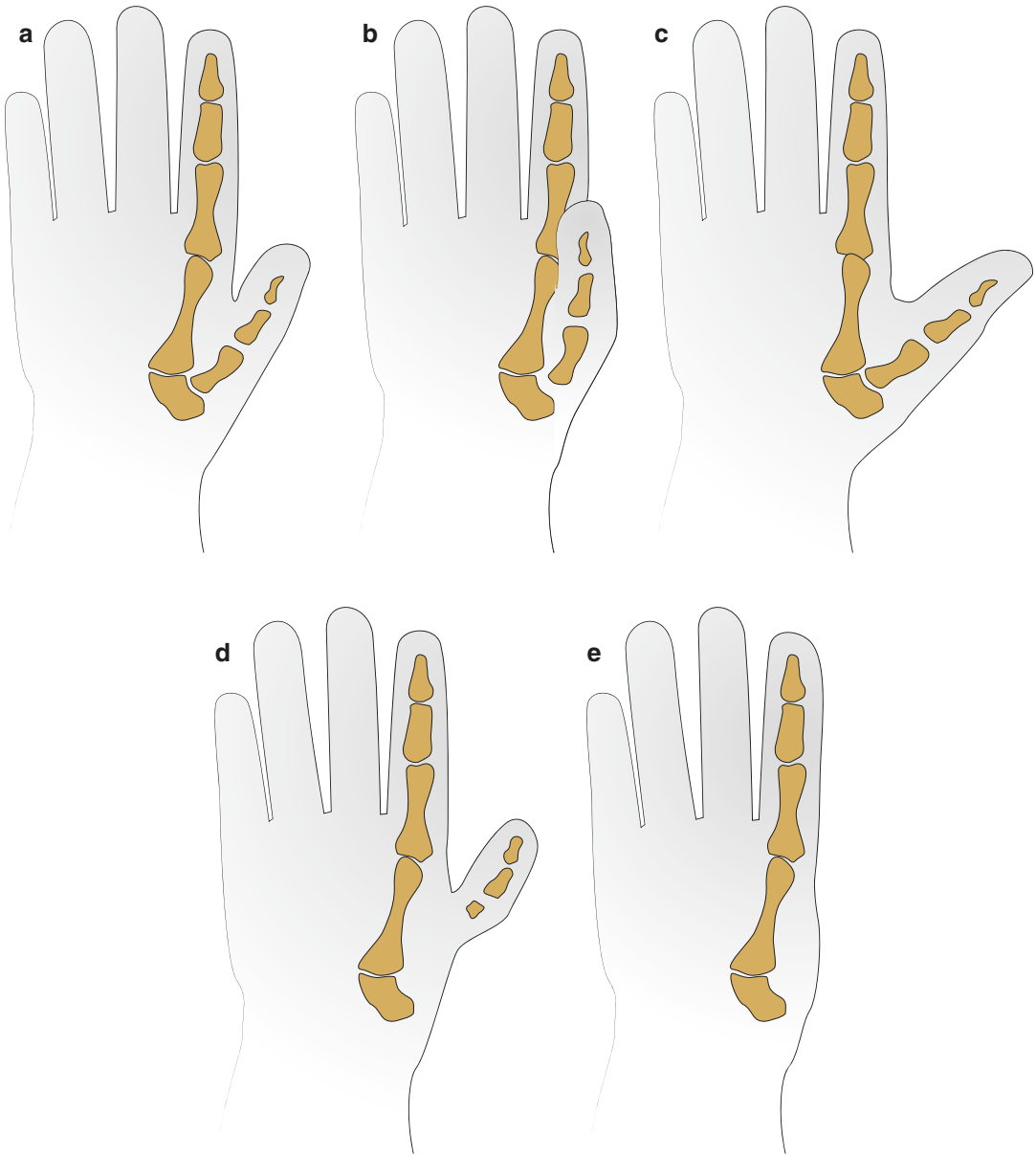



**Fig. 8.20** Hypoplasia and aplasia of the thumb. (**a**) Hypoplastic thumb. (**b**) Hypoplastic adducted thumb. (**c**) Hypoplastic abducted thumb. (**d**) Hypoplastic floating thumb. (**e**) Aplasia of the thumb (Redrawn from: Blauth W. Der hypoplastische Daumen. Arch Orthop Unfallchir. 1967;62:225–46)

Differential diagnosis:

**hypoplasia or aplasia of the thumb**

- hypoplastic thumb
- hypoplastic adducted thumb
- hypoplastic abducted thumb
- hypoplastic floating thumb
- aplasia of the thumb

Explanatory note: **hypoplasia or aplasia of the thumb.** All variations are possible: from a minimal shortening to a fully absent thumb, and from a normal to a flaccid, afunctional thumb. Thumb hypoplasia and aplasia comprises 5 % of all congenital hand abnormalities.

**Table 8.4** Syndromes of a shortened thumb and associated syndromes

| Shorter and narrower metacarpal I | Shortened and widened metacarpal I | Proximal phalanx of the thumb shortened and widened | Distal phalanx of the thumb shortened and widened |
|---|---|---|---|
| Isolated variant | Cornelia de Lange syndrome | Brachydactyly | Brachydactyly |
| Fanconi anemia | Hand, foot and uterus syndrome | | Rubinstein-Taybi syndrome |
| Holt-Oram syndrome | Diastrophic dwarfism | | Acrocephalosyndactyly (Apert syndrome) |
| Jubert-Hayward syndrome | Progressive myositis ossificans | | Acrocephalopolysyndactyly (Carpenter type) |

See Appendix for characteristics of syndromes

**Table 8.5** Tumors in the hand area. The § sign identifies tumors that aren't very common

| Benign bone tumors | Malignant bone tumors | Benign soft tissue tumors | Malignant soft tissue tumors |
|---|---|---|---|
| Enchondroma | Ewing sarcoma | Ganglion | Rhabdomyosarcoma |
| Aneurysmal bone cyst | Osteosarcoma § | Fibromatosis | |
| Osteoid osteoma | Hemangioendothelioma § | | |
| Osteoblastoma § | | | |
| Fibrous dysplasia § | | | |

See Appendix for characteristics of syndromes

*Hypoplastic thumb* The thumb is shortened. The underdevelopment is primarily bony. Loss of function is minimal and treatment is seldom needed. Apart from being shortened, the thumb may be too narrow or parts of it may be too wide. A hypoplastic thumb can also be part of a syndrome (Table 8.4).

*Hypoplastic adducted thumb* The thumb is shortened and underdeveloped. There is a skin contracture between the thumb and index finger. Flexion of the interphalangeal joint is limited due to an abnormal insertion of the flexor pollicis longus muscle. Flexion in the metacarpophalangeal joint is possible. Extension of the thumb is usually normal.

*Hypoplastic abducted thumb* The thumb is shortened and underdeveloped. There is instability in the metacarpophalangeal joint when the thumb is abducted. The thenar musculature is deficient.

*Hypoplastic floating thumb* The thumb is flaccid and afunctional. There may be two underdeveloped phalanges and the metacarpal bone is absent. The tendons of intrinsic and extrinsic muscles are absent.

*Aplasia of the thumb* The thumb is absent. An absent thumb can also occur in several syndromes (Table 8.5), and as part of a hypoplasia or aplasia of the radius or ulna.

Supplementary assessment: anteroposterior and lateral X-rays of the hand.

Primary care treatment: one should consider the possibility of a syndrome in which an abnormal thumb may be present.

When to refer: always refer because a hypoplastic or absent thumb tends to be part of a syndrome. Referral should be made as early as possible because some operations have to be carried out at an early age

Secondary care treatment: **hypoplastic thumb.** None.

**Hypoplastic adducted thumb** Correction using a Z-plasty and/or skin graft between the thumb and index finger.

**Hypoplastic abducted thumb** Arthrodesis of the metacarpophalangeal joint and correction

of the skin contracture between thumb and index finger with a Z-plasty or if necessary skin graft. A transposition can be carried out using the abductor digiti minimi tendon or of the flexor superficialis of the middle or ring finger to the thumb to improve the opposition if necessary.

**Hypoplastic floating thumb and aplasia of the thumb** Pollicization. The index finger is made into a thumb in order to make opposition possible. Some surgeons want to carry this out in babies between 6 and 9 months of age, others prefer to do the procedure between the ages of 3 and 4. The reason for the early surgery is that it produces better thumb function because the child gets used to the pollicization at a young age. It's a difficult operation with a substantial risk of complications, which may even include the loss of the pollicized finger because of vascular damage. This could be the argument for operating at a slightly older age when the structures are larger and stronger with less risk of vascular complications.

## Hand Swelling

Complaint: hand swelling.

Assessment: hand swelling (Fig. 8.21).

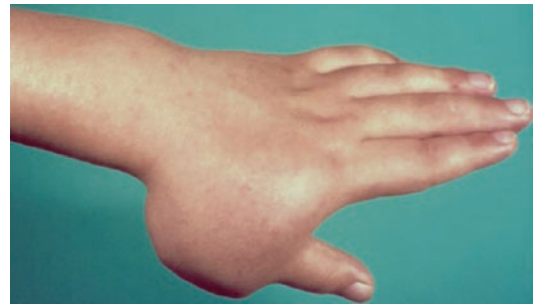


**Fig. 8.21** Hand tumor

Differential diagnosis:

**bone tumor**
**soft tissue tumor**

Explanatory note: see Table 8.5, p. 73.

Supplementary assessment: X-ray, MRI, CT-scans of the upper extremity

Primary care treatment: none

When to refer: when the X-ray or MRI show a bone or soft tissue lesion.

Secondary care treatment: **bone tumor/soft tissue tumor.** Treatment should take place in a specialized medial center. In case of a malignancy surgical removal of the tumor with, as indicated, radiotherapy and/or chemotherapy. If necessary in case of malignant tumors, and in even in some cases with large benign tumors (partial) amputation may be considered.

**Differential Diagnosis: Thumb and Finger Abnormalities**

| **Extra thumb or finger(s)** | **Polydactyly** |
|---|---|
| Extra thumb. | Radial polydactyly (preaxial polydactyly, extra thumb) |
| Extra little finger. | Ulnar polydactyly (postaxial polydactyly, extra little finger) |
| Extra index, middle or ring finger. | Central polydactyly (extra finger) |
| Seven or eight fingers with an absent thumb. | **Ulnar dimelia (mirror hand)** |
| **Gigant growth of thumb and/or finger(s)** | **Macrodactyly** |
| Non progressive during growth. | Non progressive type |
| Progressive during growth. | Progressive type |
| **Finger misalignment** | |

| | |
|---|---|
| Flexion contracture of proximal interphalangeal joint. | **Camptodactyly** |
| Progressive palmar and radial curvature of the little finger. | **Kirner deformity** |
| Snapping or locking of the digit at A1 pulley. | **Trigger finger** |
| Radial deviation of the distal interphalangeal joint. | **Clinodactyly** |
| Radial or ulnar deviation of the middle phalanx. | **Delta phalanx** |
| **Short finger** | |
| One or several fingers are too short. | **Brachydactyly** |
| **Fused fingers and/or thumb** | |
| Two or several fingers have grown together. | **Syndactyly** |
| | Simple type |
| | Complex type |
| **Thumb misalignment** | |
| Snapping or locking of the thumb at A1 pulley. | **Trigger thumb** |
| Flexion contracture of the metacarpophalangeal joint. | **Congenital clasped thumb** |
| **Long thumb** | |
| The thumb is made up of three phalanges | **Triphalangeal thumb** |
| Abnormal extra middle phalanx | Type I |
| Normal extra middle phalanx, opposition is possible | Type II |
| Five-fingered hand, opposition is not possible | Type III |
| **Short thumb** | **Hypoplastia or aplasia of the thumb** |
| Shortened thumb. | Hypoplastic thumb |
| Shortened adducted thumb. | Hypoplastic adducted thumb |
| Shortened abducted thumb. | Hypoplastic abducted thumb |
| Shortened flaccid thumb. | Hypoplastic flaccid thumb |
| Absent thumb. | Aplasia of the thumb |
| **Hand swelling** | |
| Swelling in the hand area. | **Bone tumor** |
| | **Soft tissue tumor** |

# Hip

# 9

## Hip Movement Limitations in Babies and Infants

### Hip Movement Limitation Away from the Midline of the Body

Complaint: the mother has the feeling that there is something wrong with the hips of her 3-month-old child. Changing diapers is difficult and one or both leg(s) do not abduct easily. The child has no pain.

Assessment: there is an abduction limitation of the hip(s) and an extra buttock fold (Fig. 9.1).

Differential diagnosis:

**developmental dysplasia of the hip (congenital dysplasia of the hip)**
dislocatable hip (neonatal hip instability)
acetabular dysplasia
subluxation/dislocation of the hip

Explanatory note: **developmental dysplasia of the hip.** Developmental dysplasia of the hip refers to the complete spectrum of pathological conditions involving the developing hip, ranging from dislocatable hip to acetabular dysplasia, subluxation and dislocation. The clinical presentation varies with age.

*Dislocatable hip* The number of dislocatable hips in newborns is 10–20 per 1000 births. A dislocatable hip can be easily dislocated and reduced in the first days of life. It is caused multifactorially by laxity limited to the hip joint capsule, and is influenced by hormonal and genetic factors. This laxity has a natural tendency to restore itself. A dislocatable hip corrects itself in 60 % of cases within 1 week and in 90 % of cases within 2 months after birth.

*Acetabular dysplasia* In some cases, without a subluxation/dislocation, the acetabular roofing of the femoral head will be insufficient (Fig. 9.2). This situation is known as acetabular dysplasia. It affects 10–20 out of 1000 3-month-olds. In contrast to a dislocatable hip and a dislocation, with acetabular dysplasia physical assessment tends to show no anomalies. Acetabular dysplasias are mostly diagnosed because children with increased risk of developmental dysplasia of the hip undergo supplementary assessments (Table 9.1).

*Subluxation/dislocation of the hip* A permanent subluxation/dislocation develops in 10 % of dislocatable hips, thus affecting 1–2 in 1000 children. By the time the child is 2–3 months old, secondary changes develop. An arbitrary distinction can be made between a subluxation and a dislocation. The transition from a subluxation to a dislocation is gradual. The femoral head will have respectively no full contact or no contact at all with the acetabulum.

In a hip dislocation the femoral head is dislocated backwards and upwards (Fig. 9.3). The


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_9

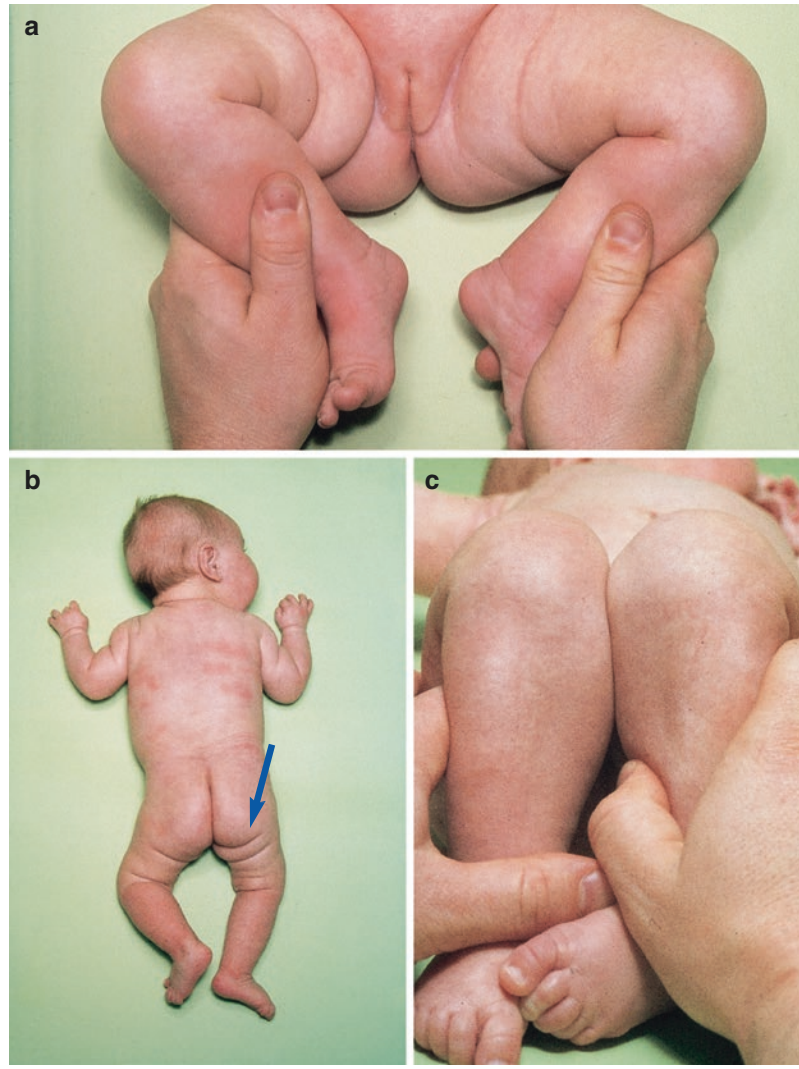



**Fig. 9.1** (**a**) Three-month-old girl with an abduction limitation in a right hip dislocation. (**b**) Extra buttock fold on the right side. The extra buttock fold is the most proximally located (*arrow*). (**c**) Galeazzi test. At the age of 3 months the difference in leg length is not so pronounced and is easily missed

superior and posterior side of the acetabulum is flattened due to pressure from the femoral head, and on the upper side of the normal or true acetabulum a new cavity is formed in the iliac bone, the new acetabulum or neoacetabulum (Fig. 9.2). The angle between the femoral head or neck and the femoral shaft is increased (enhanced valgus position) (Fig. 9.4). The femoral head has an enhanced anteversion (Fig. 9.5). In a complete dislocation the inverted hypertrophied acetabular labrum, also called limbus, obstructs the joint (labrum is Latin for lip and limbus for edge). In a subluxation this is not the case, but the superior and posterior part of the acetabular labrum is pressed flat by the femoral head against the iliac bone. The cavity is smaller, and the transverse acetabular ligament, which crosses the acetabular notch, is shortened. The ligament of the head of the femur, also known as teres ligament (teres is Latin for round and long), which runs from the acetabular fossa to the femoral head, is thicker and longer than normal, and the fat in the acetabular fossa has hypertrophied (Fig. 9.6). Between the femoral head and the acetabulum lies the tendon of the shortened iliopsoas muscle (Fig. 9.7), that constricts the joint capsule which acquires an hourglass shape. There is also a shortening (contracture) of the hip adductors, especially the adductor longus. Without anesthesia the hip joint cannot be reduced manually.

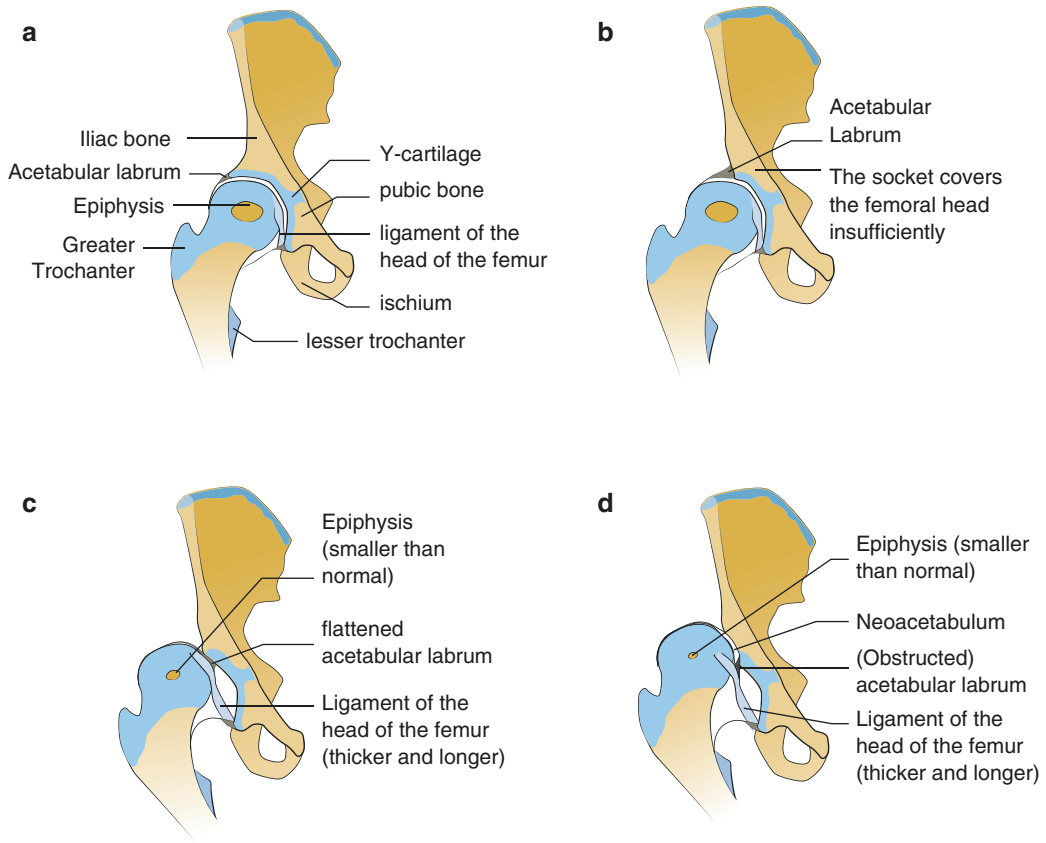



**Fig. 9.2** (**a**) Normal hip joint. (**b**) Acetabular dysplasia: there is insufficient coverage of the femoral head by the acetabulum. (**c**) Subluxation: the femoral head has lost contact with the acetabulum. The acetabular labrum, a ring-shaped connective tissue attached to the acetabular rim, also known as limbus, has been stretched superiorly and posteriorly by the migration of the femoral head and is pressed flat against the ilium by the femoral head. (**d**) Dislocation: a neoacetabulum (new acetabulum) has been formed around the iliac bone. The acetabular labrum is obstructing the joint

**Table 9.1** Risk factors for developmental dysplasia of the hip

| |
|---|
| Positive family history |
| Full breech presentation |
| Incomplete breech presentation |
| Torticollis |

## Incidence of Developmental Dysplasia of the Hip

Among Caucasians, subluxations/dislocations occur in 1–2 in 1000 children. Among Laplanders and Navajo Indians this is 20 per 1000 births. At a rate of 1 in 10,000, Africans and Asians have a relatively low incidence of developmental dysplasia of the hip. In African Bantu babies developmental dysplasia of the hip has almost never been seen (Fig. 9.8).

The left hip is affected twice as often as the right hip. This anomaly occurs in girls four more times than in boys. If developmental dysplasia of the hip occurs with no parent involved, a child has a 6 % risk of having it; with at least one parent involved the risk is 12 % and with one parent and a sibling 36 %. In a complete breech presentation the chances of a dislocation are 2 % and in an incomplete breech presentation 20 % (Fig. 9.9). According to the literature, in children with torticollis the chances of developmental dysplasia of the hip vary from 2 to 29 %.

## Syndromal and Neurogenic Hip Dislocation

Developmental dysplasia of the hip is distinguished from syndromal and neurogenic hip

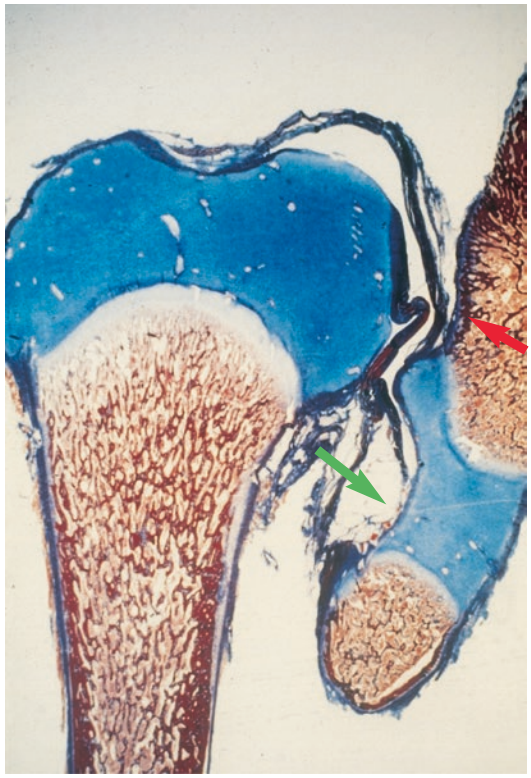


**Fig. 9.3** Anatomical preparation of a frontal cross-section of the right hip in a premature with a hip dislocation. The *green arrow* identifies the true acetabulum. In a hip dislocation the femoral head is dislocated posteriorly and superiorly, forming a neoacetabulum (new acetabulum) (*red arrow*) (Image received in 1985 from Prof. A. Campos da Paz Jr., M.D. Ph.D., Brasil.) (Figs. 9.3, 9.6 and 9.7 are published in: Campos da Paz A, Jr., Kalil RK. Congenital dislocation of the hip in the newborn. A correlation of clinical, roentgenographic and anatomical findings. Ital J Orthop Traumatol. 1976;2:261–72)

dislocation. In some syndromes (Table 9.2) and neurological anomalies, such as spina bifida, there is an increased chance of dislocations. These types of hip dislocation will not be discussed further.

## Natural History

What happens if a child with a subluxation/dislocation or an acetabular dysplasia is not treated?

If a child with a subluxation/dislocation is not treated, an unsightly limp will develop and the child will have to forgo many sports and recreational activities. In addition, in a one-sided subluxation/dislocation there is a difference in leg length that can amount to about 6 cm in a complete dislocation (Fig. 9.10). A complete dislocation does not cause any pain. A subluxation forms a new acetabulum in the iliac bone. A neoacetabulum will be painful in adulthood.

When there is acetabular dysplasia, chances of early arthrosis at ages 40–50 are about 70 %. Prematurely, between the ages of 20 and 30 many develop pain after prolonged walking, especially when strolling (Fig. 9.11).

## Physical Assessment

The anomalies found on physical assessment for a subluxation/dislocation vary with age (Table 9.3).

## Physical Assessment in a Newborn

**Ortolani test** In the Ortolani test the dislocated hip is reduced manually (Fig. 9.12). This test can only be carried out if the child is relaxed. A child that is resisting tightens the hip adductors, which makes it more likely for the test to fail. The child must be placed with the back on the mother's lap. If possible, let the mother bottle-feed the child.

The hips are bent 90°. Now the hips are examined individually. If the examiner begins with the right hip, his left hand will encompass the child's right upper leg. The thumb will lie against the inner side and the index and middle fingers against the outer side. The right hand of the examiner fixes the left upper leg and the pelvis. By abducting the right leg (starting from the midline), the dislocated femoral head is pushed back over the posterior edge of the acetabulum into the socket. The examiner will have the feeling that something has shifted with a "jerk". About the same thing is experienced if one rubs the knuckles of the fists over each other. This is incorrectly described as a "click". The "jerk" indicates that the femoral head has been reduced over the posterior edge of the acetabulum. Ortolani describes this phenomenon as a "segno dello scatto" (sign of a ridge), in this way trying to indicate that the phenomenon should be felt much rather than heard. Audible clicking coming from tendons, muscles, bone or cartilage is common and doesn't mean anything.

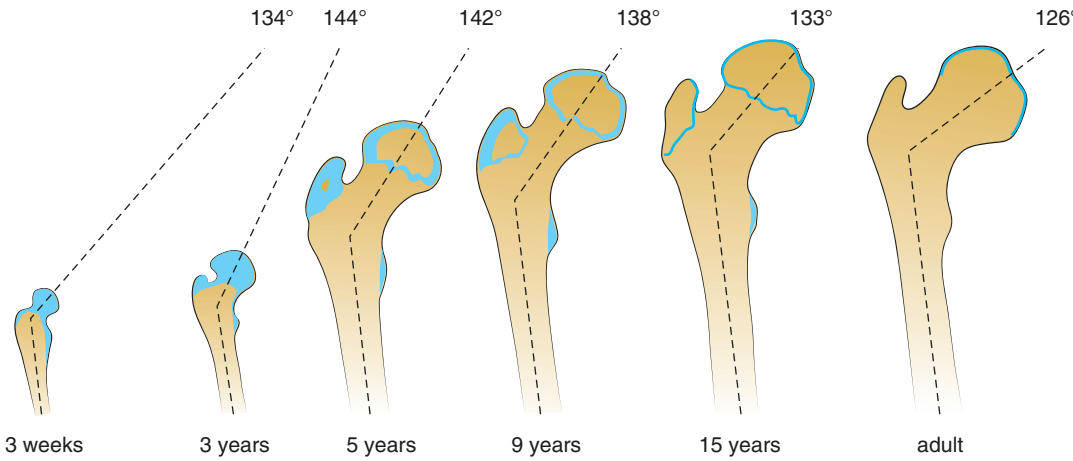



**Fig. 9.4** The neck-shaft angle, is on average 134° at birth, increasing in the first years of life to an average of 144° and diminishing gradually to an average of 126° in adulthood. In a congenital hip dislocation there is an enhanced valgus position of the proximal part of the femur (Redrawn from: Lanz T von. Über umwegige Entwicklungen am menslichen Hüftgelenk. Schweiz. Med Wschr. 1951;81: 1053–65)

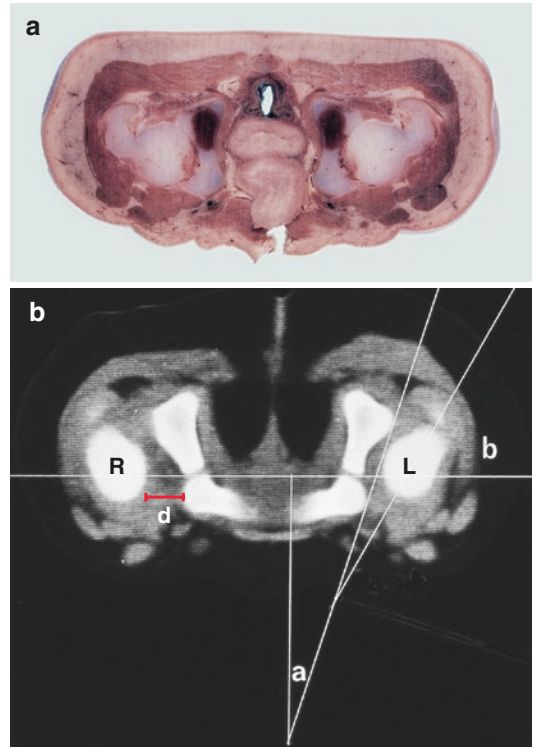



**Fig. 9.5** (**a**) Transverse crossection of the hip joint in a premature with a normal hip joint (anatomical preparation). The anteversion of the acetabulum is on average 7° shortly after birth, and 16.5° in adulthood. In a newborn the anteversion of the proximal part of the femur is on average 31°, in an adult 11°. (**b**) CT-scan of an 18-month-old boy with a right side (*R*) congenital hip dislocation. The distance between the right femoral head and the acetabulum (*d*) is larger than that of the left side (*L*). The edge of the posterior side of the right acetabulum is flattened. Acetabular anteversion (*a*) is 20° on both sides and femoral anteversion (*b*) 60° on both sides. In a dislocation an enhanced femoral anteversion is usually 50–60°

**Barlow test** This test consists of two manoeuvres (Fig. 9.13). In the first manoeuvre the hip is dislocated and in the second it is reduced. It is a test to find out whether a non-dislocated hip can be dislocated. The hips are flexed 90° and the knees are maximally flexed. The examiner encompasses the leg to be examined in the same way as is done in the Ortolani test, but in the first part of the test one presses with the thumb on the inner side of the upper part of the thigh. If there is laxity of the joint

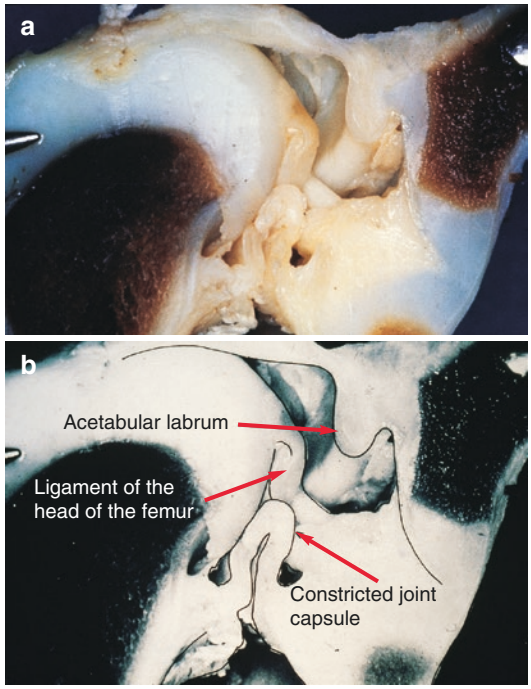



**Fig. 9.6** (**a**) Anatomical preparation of right-side dislocation. The acetabular labrum is obstructing the joint. The ligament of the femoral head is thicker and longer than normal. The joint capsule is constricted in an hourglass shape because the shortened iliopsoas tendon is located between the femoral head and the hip socket. (**b**) The most important structures are outlined (Images received in 1985 from Prof. A. Campos da Paz Jr., M.D. Ph.D., Brasil)

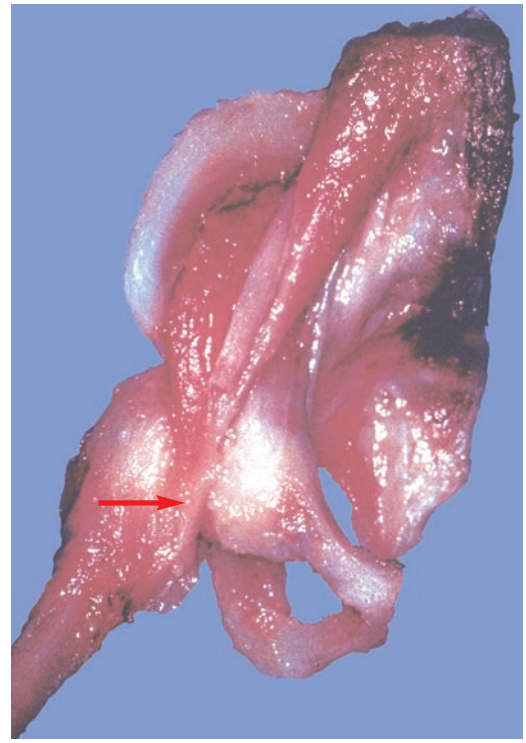


**Fig. 9.7** The shortened iliopsoas tendon (*arrow*) is located between the right femoral head and the acetabulum (Image received in 1985 from Prof. A. Campos da Paz Jr., M.D. Ph.D., Brasil)


**Fig. 9.8** (**a**) In some cultures children are wrapped up; Navajo Indians carry their children on their backs using a cradle board (Tikonagan). In this way the hips are brought into adduction and extension, increasing the risk of developmental dysplasia of the hip. (**b**) African mothers carry their child on their back, which bends and abducts the hips. This is the best position to counteract developmental dysplasia of the hip

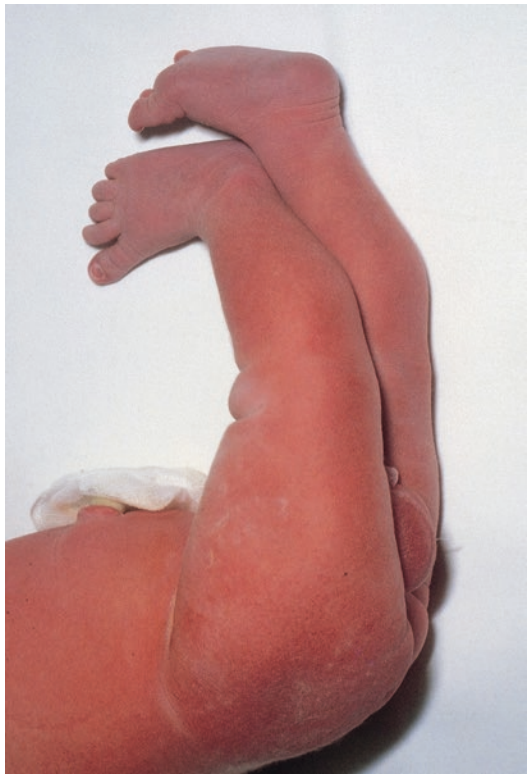


**Fig. 9.9** Hip dislocations are frequent with breech presentations especially in an incomplete breech presentation when there are hyperextended knees

**Table 9.2** Syndromes with a highly increased risk of hip dislocation

| |
|---|
| Arthrogryposis |
| Diastrophic dwarfism |
| Down syndrome |
| Ehlers-Danlos syndrome |
| Larsen syndrome |
| Hunter syndrome |
| Hurler syndrome |

See Appendix for characteristics of syndromes

capsule the femoral head can be easily displaced over the posterior edge of the acetabulum. In the process one can feel the "jerk" when dislocation of the hip is possible. In the second manoeuvre the middle finger is pressing on the outer side of the greater trochanter, so that feeling the "jerk" means that the femoral head has been reduced over the posterior edge of the acetabulum.

**Fig. 9.10** Leg length difference of 2 cm in the left thigh as a result of a developmental dysplasia with a dislocated left hip. The leg length difference can increase in adulthood up to 5 or 6 cm

Why is a positive Ortolani test found in some cases, and in other cases a positive Barlow test? This depends on whether a dislocatable hip in a newborn is dislocated or not. This is probably influenced by the position of the legs during the assessment. If a child with a dislocatable hip lies supine with the hips in flexion and in 80–90° abduction — a normal resting position — then the hip can be reduced. In that case there is a positive Barlow test. A joint can be dislocated when the child lies with extended and adducted legs, that means that there is a positive Ortolani test.

One should start by slowly extending the legs and adducting them if only the Ortolani test is to be done. The hip will dislocate. Next, perform the test as described above. It is recommended to

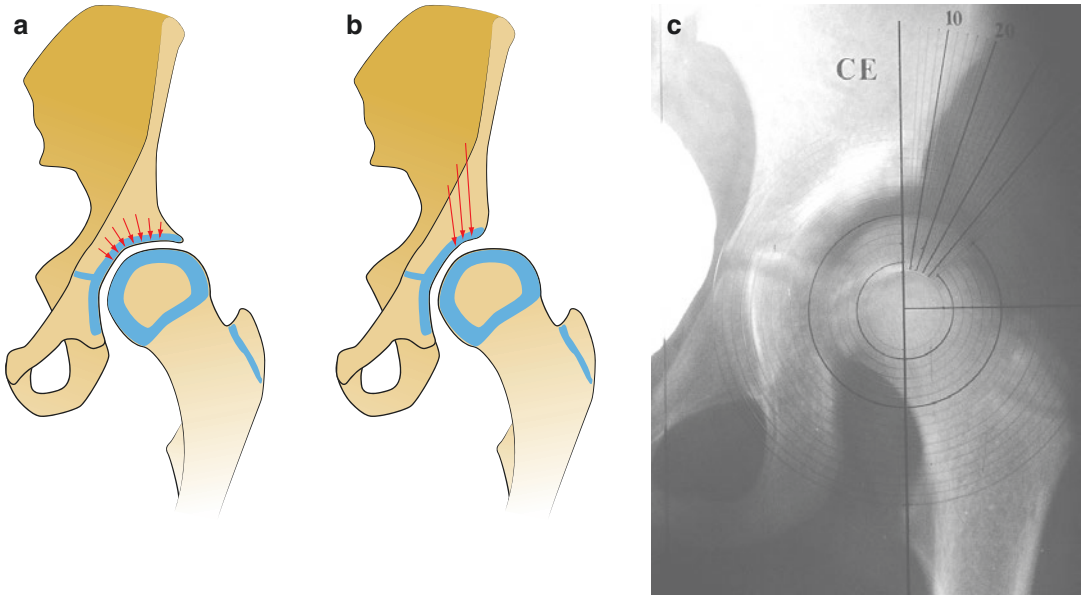



**Fig. 9.11** (**a**) Normal hip. Good distribution of the load on the hip joint. (**b**) Acetabular dysplasia. Abnormally large load on a part of the hip joint with chances of degenerative changes. (**c**) In older children and adults, in an anteroposterior X-ray of the hip the relation between the acetabulum and the femoral head is displayed by means of the CE angle of Wiberg. C stands for "center" and E for "lateral Edge of the acetabular roof". The CE angle is formed by a line drawn from the middle point of the head parallel to the median line, and a line drawn from the middle point of the head to the lateral edge of the acetabulum. The CE angle is 25° or more when the acetabulum properly covers the femoral head. A CE angle of 20° or less means that there is acetabular dysplasia. An angle between 20° and 25° is a gray area and not clearly dysplastic. In this case the CE angle is 10° and the acetabulum is clearly dysplastic

**Table 9.3** Examination findings for developmental dysplasia of the hip by age

| Age | Physical examination |
|---|---|
| 0–2 months | Positive Ortolani and/or Barlow test(s) |
| 2–9 months | Abduction limitation |
| Around 1 year | Difference in leg length |
| After the child starts walking | Waddling gait |
| Acetabular dysplasia | Physical assessment tends to show no anomalies |

start with the Ortolani test, and if it turns out negative then do the Barlow test.

Doing a hip assessment on a newborn is not a simple matter. Ample experience is required. Occasionally there is a dislocation in newborns, irreducible when doing the Ortolani test, as secondary intrauterine changes will have already taken place in terms of contractures of the iliopsoas muscles and the adductors. This anomaly is called teratogenic hip dislocation.

## Physical Assessment at the Age of 3–4 Months

As a rule, the Ortolani and Barlow tests for a dislocatable hip are positive in the first days to a few months after birth. Three situations are possible after that:

- Ligamental laxity of the hip joint capsule does not correct itself after a few months. The Ortolani and Barlow tests remain positive for a longer period of time. This is very rare.
- Ligamental laxity of the hip joint capsule corrects itself within several months, the femoral head sits in the joint cavity, and the hip develops normally. In some cases, despite the fact that the femoral head is in the acetabulum, the development of the hip joint will not be entirely optimal. The acetabular coverage of the femoral head is not ideal, and there is acetabular dysplasia. A child with acetabular dysplasia does not necessarily show physical anomalies.

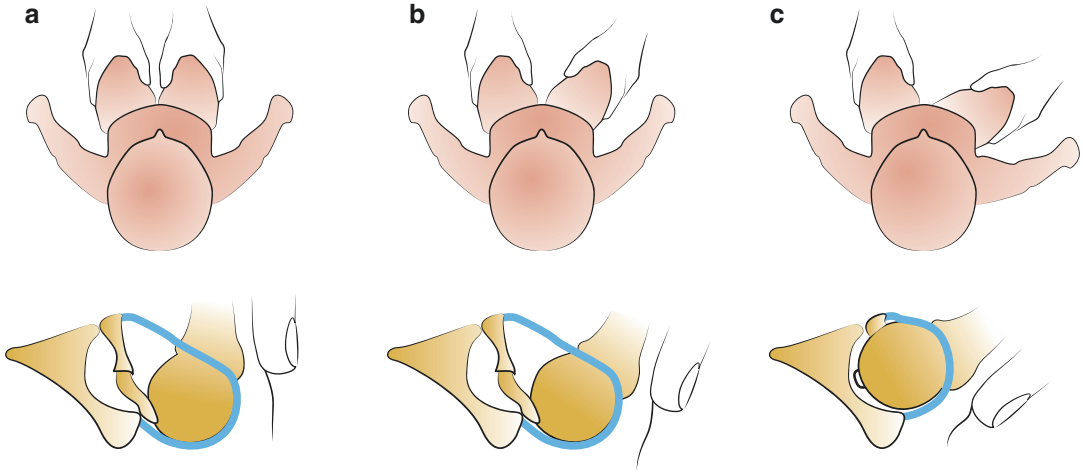



**Fig. 9.12** Ortolani test. "Jerk of entry". (**a**) The hip joint is dislocated. The hips are bent at 90°. (**b**) By abducting the leg the hip is repositioned over the posterior edge of the hip socket. (**c**) By increasing abduction the examiner feels that something shifts with a "jerk"

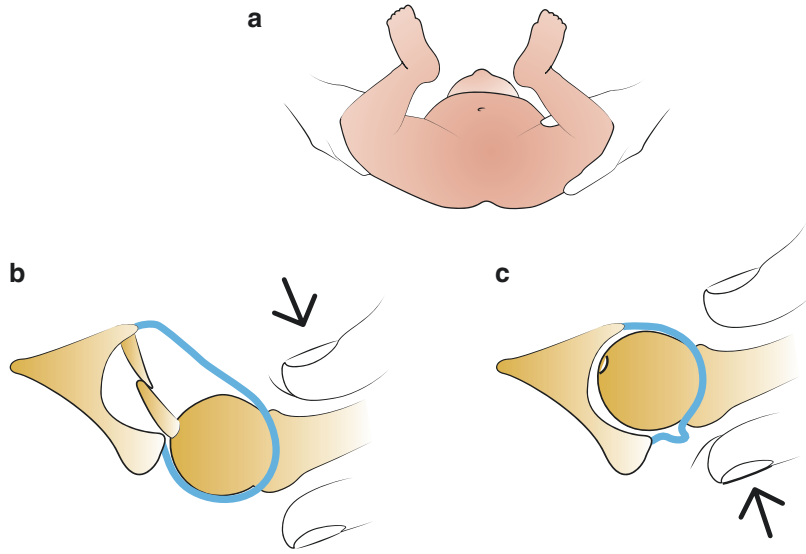



**Fig. 9.13** Barlow test. This test is used to ascertain whether a non-dislocated hip joint can be dislocated. (**a**) The hips are bent 90° and the knees are maximally flexed. (**b**) If there is ligamental laxity, the hip can be dislocated with pressure of the thumb on the inner side of the proximal part of the thigh. At that moment the examiner feels that something changes place with a"jerk". (**c**) In the second part of the manoeuvre the hip is reduced by pressing on the greater trochanter with the index and middle fingers. The moment that the hip is reduced the examiner feels once more that something changes position with a "jerk"

- The hip remains dislocated causing in the first 2 or 3 months after birth contractures of the iliopsoas muscle and the adductors, especially the adductor longus muscle. Due to the contractures the hip cannot be reduced with the Ortolani test. At first the adduction contracture is not that obvious and can easily be missed. Sometimes it is first noticed by the mother or carers, because changing diapers becomes more difficult and the legs cannot be sufficiently abducted (Fig. 9.1). Normally, at the age of 3 months legs can still be abducted

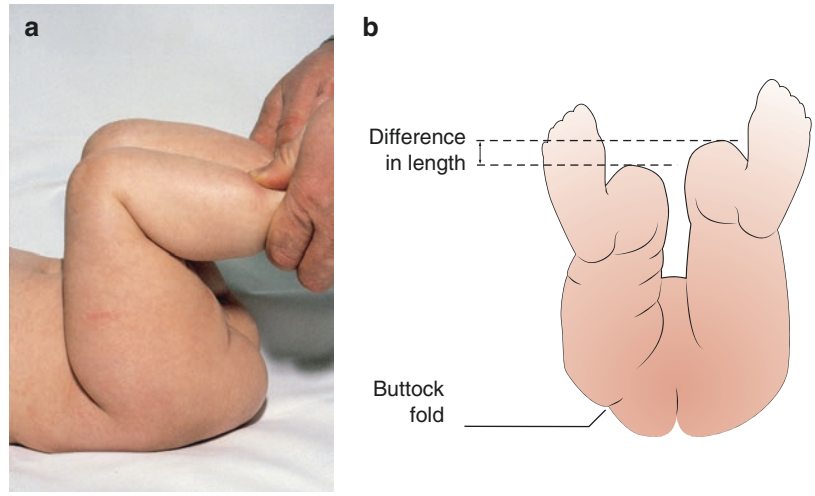



**Fig. 9.14** (**a**) The difference in length between the thighs can also be observed by flexing the hips and knees 90°. (**b**) This is in principle a better method than the Galeazzi test (Figs. 9.1a and 9.15c), because the Galeazzi test is also positive if there is a difference in leg length below the knees. This examination only measures the difference in the thighs

80–90° when the hips are flexed 90°. Abduction of less than 60° should be considered as abnormal and further radiological assessment is indicated. ❗

An extra buttock fold is noticeable in children who are 2–3 months old and have a dislocated hip joint. The extra fold is caused by the dislocation, because the thigh at the level of the dislocated hip becomes relatively too short and a surplus of soft tissue occurs which causes the extra fold. This extra buttock fold is close to the perineum, and not somewhere else on the posterior side of the thigh ❗ (Fig. 9.1). Such folds are present in 20% of children without hip disorders, hence an extra buttock fold without an abduction limitation has no relevance.

The normal groin and buttock folds on the dislocated side are longer and deeper than on the non-dislocated side. In unilateral dislocations there is also a difference in length of the thighs. This can be demonstrated with the Galeazzi test (Fig. 9.1). Here the knees and hips are flexed to a degree such that the soles of the feet are approximated on the examination table. If there is a difference in knee lengths, to the disadvantage of the dislocated side, one speaks of a positive Galeazzi test. The difference in length between the thighs can also be observed by flexing the hips and knees to 90° (Fig. 9.14). This is actually a better method, because the Galeazzi test is also positive when there is a difference in length below the knees. At the age of 2–3 months the difference in leg length is not that pronounced though, so it can easily be missed. ❗ In bilateral dislocations there is of course no difference in leg length, and the abduction is symmetrically limited. ❗

### Physical Assessment Around the First Year of Life

As the child becomes older, the abduction in the non-dislocated hip diminishes while the adduction contracture of the dislocated hip decreases. In other words, the difference in abduction becomes less pronounced and more difficult to see. At the same time, the extra buttock fold disappears and the normal groin and buttock folds on the dislocated side become as long and as deep as those on the non-dislocated side. The soft parts adapt themselves. By contrast, the difference in leg length increases (Fig. 9.15).

### Physical Assessment After the Child Can Stand and Walk

Occasionally we see that a congenital hip dislocation is identified at an older age. This usually involves a dislocation of both hip joints. In such cases there is a symmetrical abduction limitation that is not as easily noticeable as an unilateral abduction limitation, as is the case in unilateral dislocations. In addition, there is no difference in leg length between the thighs because both hip joints are dislocated, so in most cases there is a

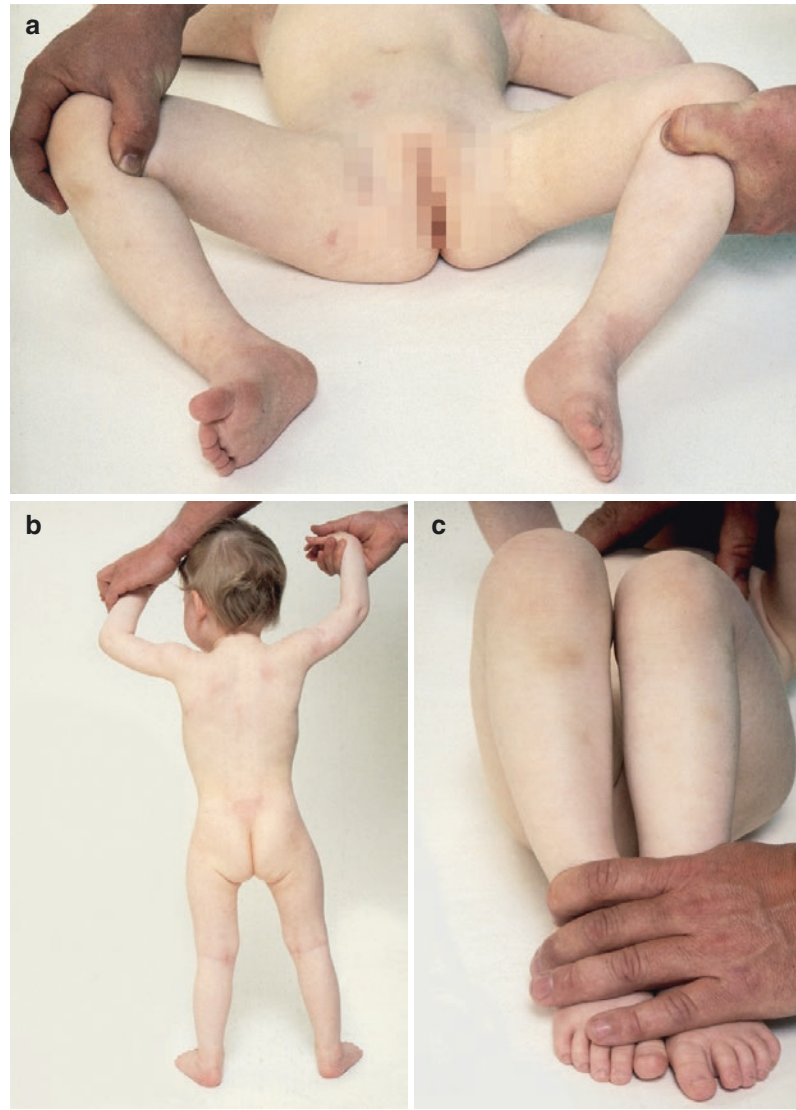



**Fig. 9.15** (**a**) 16-month-old girl with a left-sided hip dislocation. At this age the difference in abduction is less pronounced. (**b**) The same patient as in Fig. 9.15a. The extra buttock fold is absent or gone. (**c**) The same girl as in Fig. 9.15a. With age, the difference in leg length in a one-sided hip dislocation becomes more noticeable. The Galeazzi test shows a clear difference in knee height to the disadvantage of the dislocated left side

similar shortening of both thighs. When these children stand up, the air space between the legs, around the perineum, is widened (also called thigh gap). There is an abnormal hollow back (hyperlordosis) and the pelvis is tilted forwards (Fig. 9.16). Parents often complain that the child has a waddling gait. The Trendelenburg test is positive (Fig. 9.17). The Trendelenburg test serves to assess the strength of the hip abductors. One can ask the patient to lift the left leg. When there is normal strength in the right hip abductors, the left side of the pelvis is lifted. In such cases the Trendelenburg test is negative. In a hip dislocation the distance between the origin and insertion of the hip abductors is shortened. This makes the hip abductors relatively too long and therefore less effective when the child stands on the leg with the hip dislocation, when lifting the other (contralateral) leg the pelvis on the contralateral side will drop downwards. In this case the Trendelenburg test is positive (Fig. 9.17).

Supplementary assessment: When should an ultrasound assessment of the hips take place or anteroposterior X-rays of the pelvis be taken? This is done if the physician, upon physical examination, finds indications of a possible developmental dysplasia of the hip. Additional radiological assessment is recommended

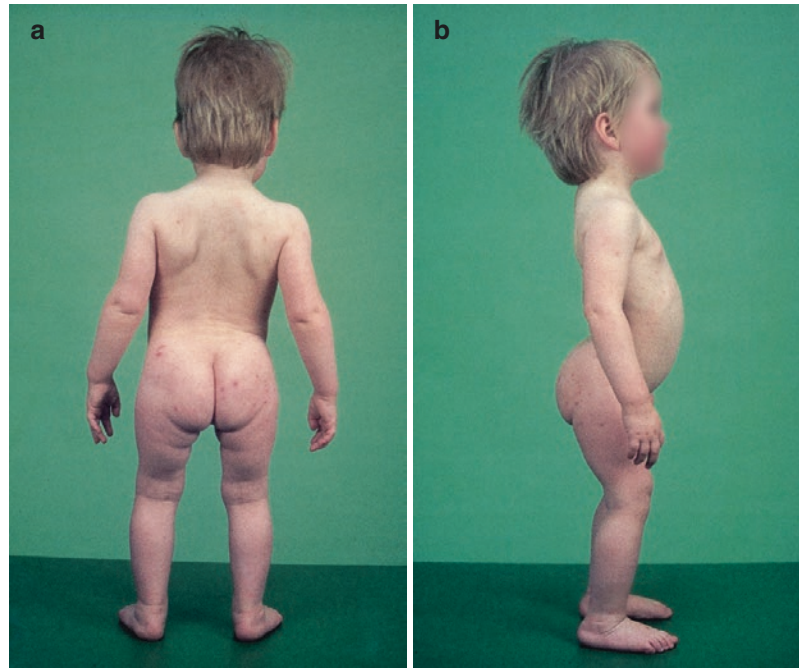



**Fig. 9.16** (**a**) Girl aged age 2 years and 3 months. There is a bilateral hip dislocation with a widened air space just below the perineum (thigh gap). (**b**) There is enhanced lumbar lordosis with a forward-tilting pelvis

if physical examination shows no irregularities but there is developmental dysplasia of the hip in the family or there has been a breech birth presentation, or there is a torticollis (Table 9.1).

Ultrasound or echographic assessment makes use of sound waves. This type of assessment is useful until about the age of 1 year. In older children it is unreliable. The advantage of ultrasound is that no radiation is involved. A disadvantage of ultrasound is that the professional doing the examination must have ample experience with ultrasound assessment of children's hips in order to establish the proper diagnosis. If this person lacks sufficient experience with such assessments, there is a risk that the hip irregularity will not be recognized or that hip anomalies will be seen that aren't there (Fig. 9.18). The advantage of an X-ray is that it easier to assess than an ultrasound (Fig. 9.19). The disadvantage is of course that radiation is involved.

A positive Ortolani or Barlow test in a newborn can be easily visualized with ultrasound technology because it is a dynamic assessment. If after the age of 1–2 months the Ortolani and Barlow tests are negative but physical examination points to other possible hip anomalies (for example abduction limitation), then ultrasound assessment of the hips is advisable. The same applies if physical examination shows no abnormalities despite an increased risk of developmental dysplasia (Table 9.1). Anteroposterior X-rays of the pelvis are made right away if the ultrasound shows abnormalities.

The degree of acetabular dysplasia is determined on the basis of the acetabular angle (Fig. 9.19) and the Tönnis and Brunken table (Table 9.4).

Primary care treatment: In primary care all children, in any event at the age of 3 months, should be examined for developmental dysplasia of the hip. Radiological assessment should be requested if physical examination shows signs that point to developmental dysplasia of the hip as well as in children with increased chances of this condition (Table 9.1).

When to refer: If physical and/or radiological assessment point to signs of developmental dysplasia of the hip[1].

[1] A child that shows an abduction limitation of the hip(s) upon physical examination, but whose hip ultra sound assessment or an anteroposterior X-ray of the pelvis show no abnormalities, does not need to be referred. This involves a postural anomaly that corrects spontaneously.

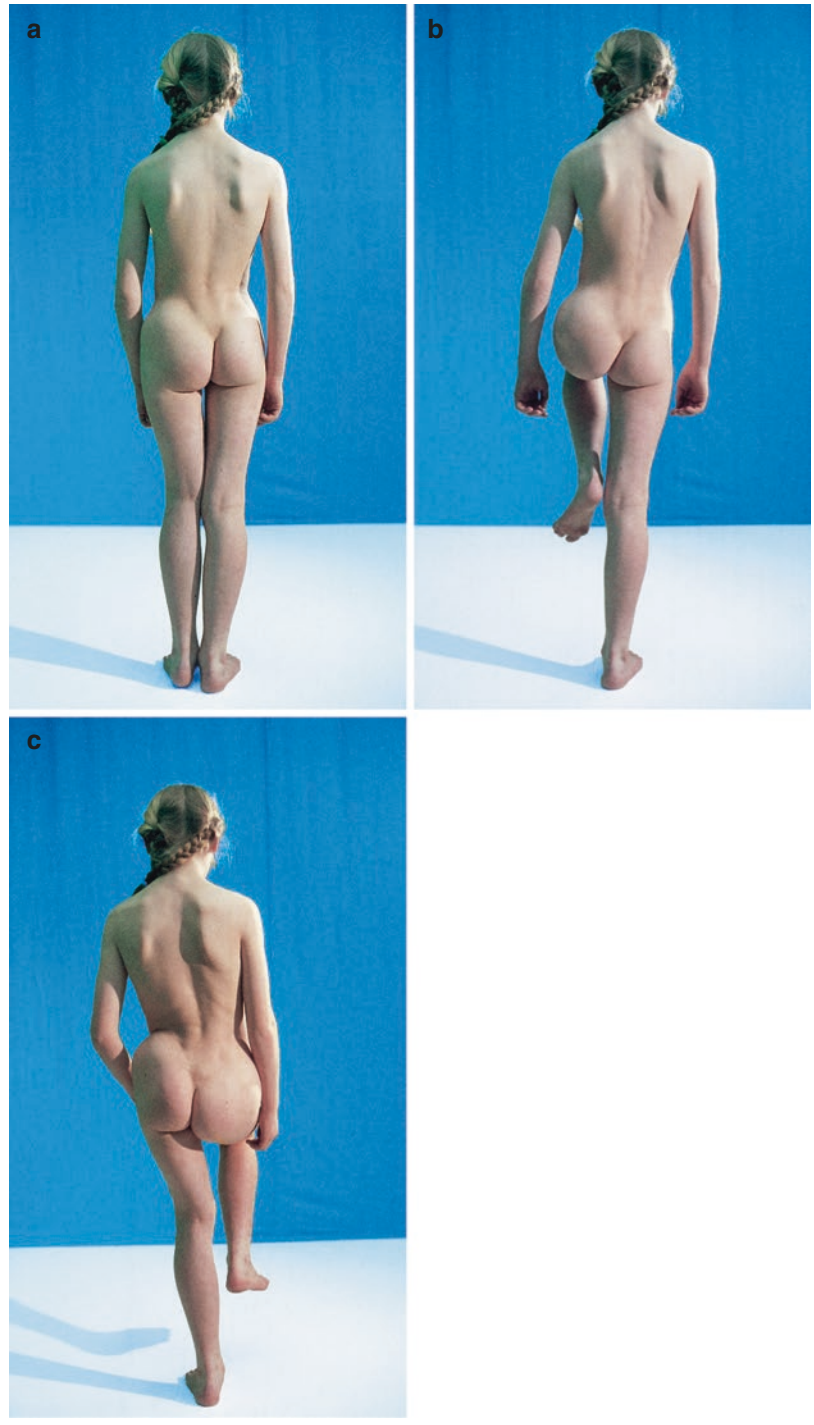



**Fig. 9.17** Trendelenburg test. (**a**) A patient with a leftsided hip dislocation stands with both legs on the ground. (**b**) The patient is asked to lift the left leg. If there is normal strength in the hip abductors of the right hip the left side of the pelvis is lifted. The Trendelenburg test is negative. (**c**) When lifting the right leg the pelvis drops on the right side. The Trendelenburg test is positive. There is weakness in the hip abductors of the left hip

Secondary care treatment: **dislocatable hip.** Hip abduction treatment for 8 weeks using a Pavlik harness (Fig. 9.20).

**Acetabular dysplasia** We will treat it, if the anteroposterior X-ray of the pelvis shows acetabular dysplasia. However, this can sometimes be overtreated.

Up to the age of 3 months treatment is carried out using a Pavlik harness. For babies 3–6 months old one can choose a Pavlik harness and a Visser

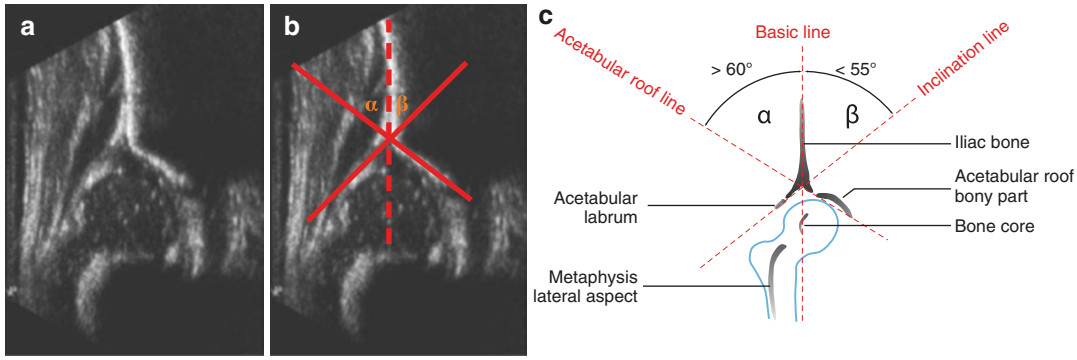



**Fig. 9.18** (**a**) Ultrasound of the right hip in a 3-month-old child. The bone core of the femoral head is not present yet. (**b**, **c**) The α angle between the basic line (iliac bone) and the acetabular roof line shows the degree of bony development of the acetabulum, and in a normally developed hip it exceeds 60°. The β angle that is formed by the basic line and the inclination line gives an impression of the cartilaginous development of the acetabulum. In a normal hip this angle is smaller than 55°. Graf came up with several classifications based on the ultrasound. The simplest classification is: class I, normal hip with an α angle larger than 60° and β angle smaller than 55°; class II, dysplasia with an α angle between 43° and 60° and a β angle between 55° and 77°; class III: subluxation with an α angle smaller than 43° and a β angle larger than 77°; class IV: dislocation, α and β angles cannot be measured

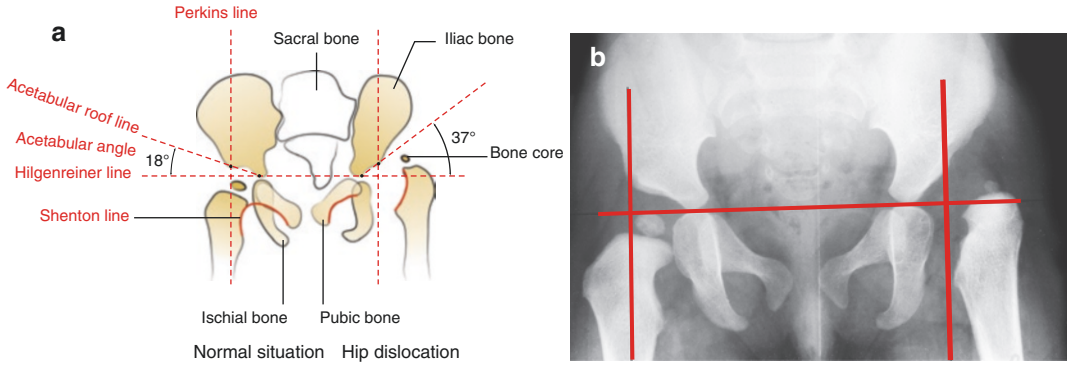



**Fig. 9.19** (**a**) Graphic depiction of an anteroposterior X-ray of the pelvis. To assess a hip dislocation and acetabular dysplasia a number of lines are drawn. A horizontal line is drawn from the y-shaped cartilage of both femoral heads: the Hilgenreiner or y-line. The y-shaped cartilage is the common growth plate of the iliac, ischial and pubic bones. The Perkins lines are dropped, from the lateral edge of the acetabula which lie perpendicular to the Hilgenreiner line. In normal hips and in acetabular dysplasia the medial edge of the proximal femoral metaphysis lies within the Perkins line, in a dislocation it lies outside. The degree of acetabular dysplasia is the acetabular angle, often abbreviated as AC angle and also called acetabular index. The acetabular angle is formed by the Hilgenreiner line and a line through the acetabular roof (acetabular roof line). In the figure the right hip displays a normal acetabular angle and the left hip a congenital hip dislocation with an enlarged acetabular angle. In a hip dislocation there is always an underdevelopment of the acetabulum, and the ossification of the epiphysis is smaller than on the normal contralateral side. The normally flowing line of Shenton, set by the arch of the lower edge of the superior ramus of the pubic bone and the medial edge of the medial border of the metaphysis, is interrupted in a dislocation (and in an enhanced anteversion-valgus position of the proximal part of the femur). (**b**) Anteroposterior image of the pelvis in a 6-month-old child. The lines of Hilgenreiner and the lines of Perkins are drawn. The right side displays a normal hip, the left side a congenital hip dislocation. A hip dislocation is always accompanied by an underdevelopment of the acetabulum, (acetabular dysplasia)

hip abduction orthosis (Fig. 9.21). Small babies get a Pavlik harness at this age, and larger children a Visser hip abduction orthosis. The Pavlik harness is too small for older children. Children with acetabular dysplasia are treated using a dynamic hip abduction orthosis (Visser hip abduction orthosis) between the ages of 6 and 12 months.

**Table 9.4** Threshold values of the acetabular angle in degrees, for the diagnosis of mild or severe acetabular dysplasia, by age and gender according to Tönnis and Brunken (1968), based on a study of 2294 acetabular angles

| | Acetabular dysplasia girls | | | | Acetabular dysplasia boys | | | |
|---|---|---|---|---|---|---|---|---|
| | Mild | | Severe | | Mild | | Severe | |
| Age | Right | Left | Right | Left | Right | Left | Right | Left |
| Months | | | | | | | | |
| 1+2 | 35.8 | 36.1 | 41.6 | 41.6 | (27.7) | 31.2 | (31.8) | 35.2 |
| 3+4 | 31.4 | 33.2 | 36.3 | 38.7 | 27.9 | 29.1 | 32.4 | 33.7 |
| 5+6 | 27.3 | 29.3 | 31.8 | 34.1 | 24.2 | 26.8 | 29.0 | 31.6 |
| 7–9 | 25.3 | 26.9 | 29.4 | 31.1 | (24.6) | 25.4 | 28.9 | 29.5 |
| 10–12 | 24.7 | 27.1 | 28.6 | (31.4) | 23.2 | 25.2 | 27.0 | 29.1 |
| 13–15 | 24.6 | 26.9 | (29.0) | (31.7) | 23.1 | 24.0 | (27.5) | 27.7 |
| 16–18 | (25.0) | (26.1) | (29.3) | 30.4 | (23.8) | (25.8) | (28.1) | (30.0) |
| 19–24 | 24.1 | 26.4 | 28.4 | (30.8) | 20.6 | 23.2 | 24.4 | 27.3 |
| Years | | | | | | | | |
| 2–3 | 21.8 | 23.3 | 25.6 | 27.1 | 21.0 | 22.7 | 25.3 | 26.9 |
| 3–5 | (17.9) | 21.2 | 21.3 | 25.8 | 19.2 | 19.8 | 23.5 | 23.8 |
| 5–7 | 19.3 | 19.8 | 23.4 | 23.8 | 16.8 | 19.3 | 20.9 | 23.2 |

Mild acetabular dysplasia: Threshold values equal to the average plus one time the standard deviations
Severe acetabular dysplasia: Threshold values equal to the average plus twice the standard deviation
Hips with an acetabular angle between the average and once-to-twice the standard deviation (mild acetabular dysplasia) develop normally in 40 % of cases, present an elevated normal value in 40 % of cases, and in 20 % of cases have acetabular dysplasia. Hips with an acetabular angle of the average plus two times the standard deviation or more (severe acetabular dysplasia) do not develop normally (persistent acetabular dysplasia) if no treatment is deployed. The numbers in parentheses are higher than in the current curve trajectory one would expect. A number should be taken that lies between the number above and the number below

**Fig. 9.20** Pavlik harness. A disadvantage of the Pavlik harness is that parents sometimes have difficulty fitting it

Children between the ages of 1 and 2 years get a Hilgenreiner orthosis (Fig. 9.22), generally for 6 months. This orthosis allows the child to walk. For the conditions that have to be met by a hip abduction orthesis to treat developmental dysplasia of the hip, see Table 9.5. Abduction treatment in children older than 2 years produces no better results than the natural recovery. A natural recovery is waited for until about the age of 4 years. After this age, a few degrees of improvement in the acetabular angle may be expected. If at 4 years of age the acetabular angle is not expected to normalize a pelvic osteomy must be carried out in order to improve coverage of the femoral head (Table 9.6, Figs. 9.23, 9.24, 9.25, 9.26, 9.27, and 9.28).

In the indication for hip osteotomy several principles should be taken into consideration. The largest deformity is corrected first. This is usually the acetabulum. If the deformity is located in the acetabulum as well as in the proximal femoral part, the pelvic osteotomy is combined with a proximal femoral osteotomy. In a Pemberton pelvic osteotomy the shape of the acetabulum is deformed because the center of rotation of this osteotomy is located at the level of the y-cartilage. A Pemberton pelvic osteotomy can be performed only up to the age of 6. Above this age the acetabulum has insufficient time to remodel itself.

A Salter pelvic osteotomy can be performed at any age but is only suitable for mild acetabular dysplasia.

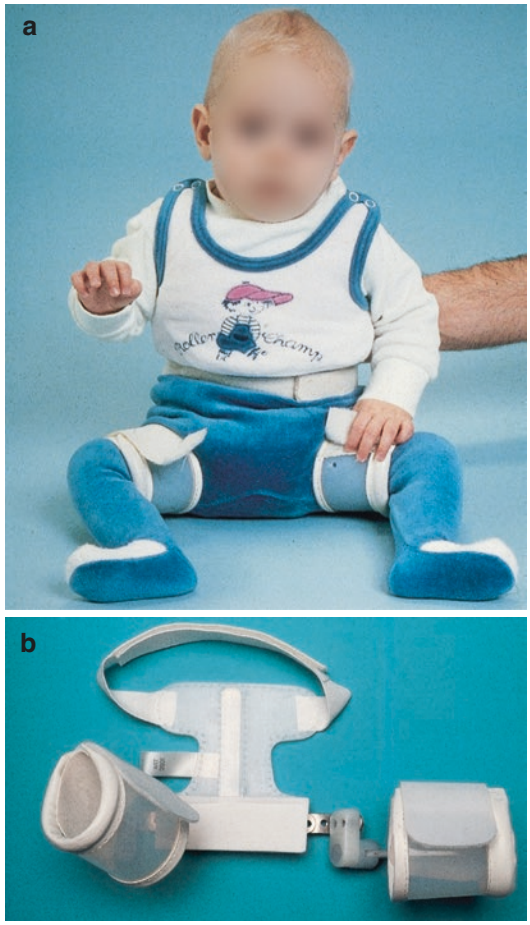



**Fig. 9.21** (**a**, **b**) Visser hip abduction orthosis. (**b**) The Visser hip abduction orthosis has hinges, which allows mobility in the hip joints within certain limits (greater than 90° flexion and 30° abduction)

**Fig. 9.22** Hilgenreiner orthosis. One can walk with this orthosis in contrast to a Pavlik harness and a Visser hip abduction orthosis. A Pavlik harness and a Visser hip abduction orthosis are stock items, a Hilgenreiner orthosis has to be made to size

**Table 9.5** Conditions that a hip abduction orthosis for treatment of developmental dysplasia of the hip must meet

| |
|---|
| The hips are held in flexion to neutralize the contracture of the iliopsoas muscle. |
| The hips are held in 90° flexion and 30°–60° abduction for optimal centralization of the femoral head in the hip socket. |
| Forced extreme abduction should be avoided to prevent ischemic femoral head necrosis. |
| The hips should be movable within certain limits for an optimal nutritional condition of the cartilage. |
| The orthosis should be user-friendly. The materials of the orthosis should not cause hypersensitivity. |

Sutherland and triple osteotomies are usually performed on children older than 10, and a Ganz pelvic osteotomy once a child has reached adulthood.

Pemberton, Salter, Sutherland, triple and Ganz pelvic osteotomies can only be performed on a congruent joint. For severe dysplasias that still have a congruent joint there is no adequate solution for children aged 6–10. In most cases one waits until the child is past age 10 to do a triple pelvic osteotomy, or a Ganz pelvic osteotomy can be performed in adulthood.

A Chiari osteotomy for lateralization or subluxation of the hip, a shelf arthroplasty or a Chiari pelvic osteotomy for acetabular dysplasia with a discongruent joint are generally performed on children older than 10.

*Pemberton pelvic osteotomy* This osteotomy can be performed for mild and severe acetabular dysplasia until the maximum age of 6. It is an osteotomy of the ilium. The osteotomy begins just above the lateral edge of the acetabulum and runs through up to the y-cartilage. The y-cartilage is the center of rotation in this osteotomy.

**Table 9.6** Treatment followed by the authors for a dislocatable hip, acetabular dysplasia with or without a preceding treatment for dislocation, persistent subluxation and a discongruent joint

| Anomaly | Age | Treatment |
|---|---|---|
| Dislocatable hip | 0–2 months | Pavlik harness |
| Dysplasia | 0–6 months | Pavlik harness |
| Dysplasia | 3–12 months | Visser hip abduction orthosis |
| Dysplasia | 1–2 years | Hilgenreiner orthosis |
| Dysplasia | 2–6 years | Pemberton pelvic osteotomy |
| Mild dysplasia | 6 years – fully grown up | Salter pelvic osteotomy |
| Severe dysplasia | 6 years – fully grown up | Wait until the child is fully grown |
| Mild to severe dysplasia | >10 years | Triple osteotomy |
| | Fully grown up | Ganz pelvic osteotomy |
| Severe dysplasia | Fully grown up | Ganz pelvic osteotomy |
| Persistent subluxation | >10 years | Chiari pelvic osteotomy |
| Dysplasia with discongruent joint | Mostly > 10 years | Shelf arthroplasty or Chiari pelvic osteotomy |
| (Nonspherical femoral head and/or acetabulum) | | |

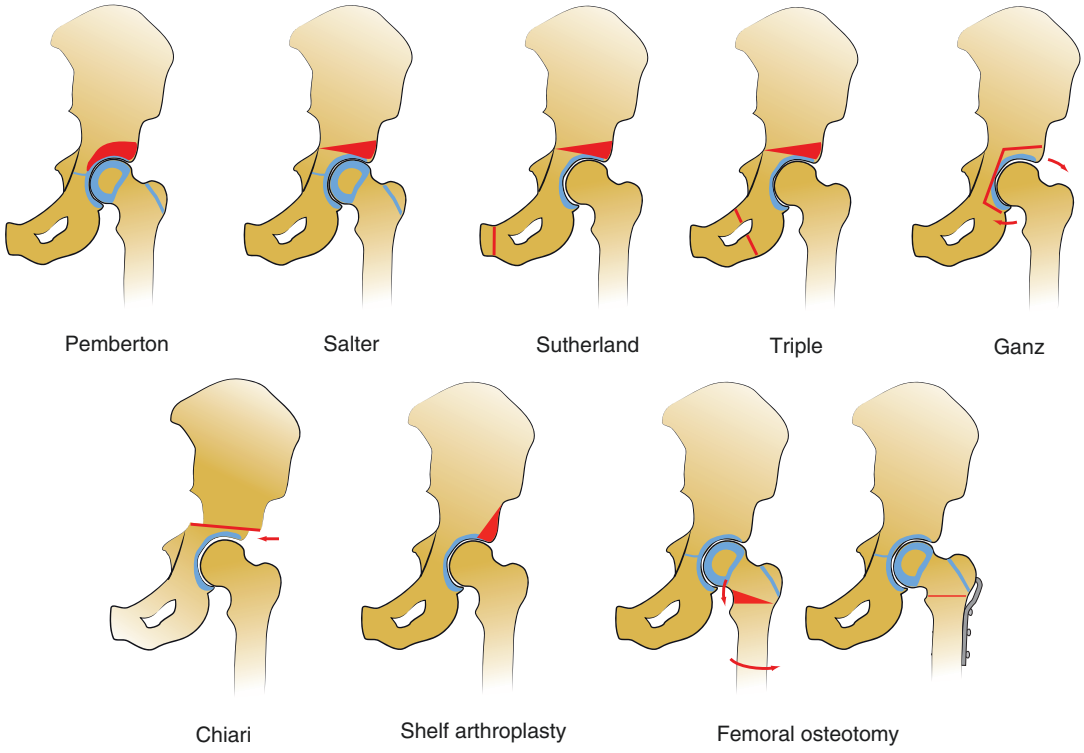



**Fig. 9.23** Pelvic and femoral osteotomies for treatment of acetabular dysplasia (Redrawn from: Staheli RT. Fundamentals of pediatric orthopaedics. 4th ed. Philadelphia/Baltimore/New York/London/Buenos Aires/Hongkong/Sydney/Tokyo: Lippincot Williams & Wilkins. 2008. p. 217)

*Salter pelvic osteotomy* The Salter pelvic osteotomy is suitable for mild acetabular dysplasia and can be performed at any age. The procedure improves the acetabular angle by a maximum of 10–15° and the CE angle by no more than 10°. The symphysis is the center of rotation of this ilium osteotomy.

*Sutherland pelvic osteotomy* Double osteotomy through the ilium and the pubic bone. This

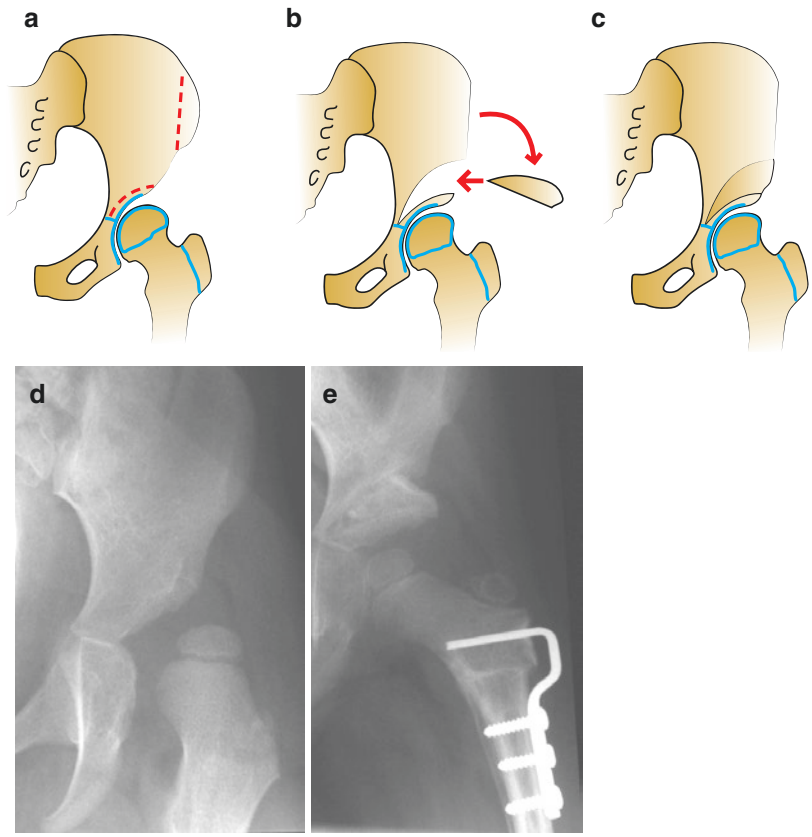



**Fig. 9.24** *Left side*. (**a**–**c**) Schematic representation of a Pemberton pelvic osteotomy. The piece of bone obtained from the left iliac crest is wedged in-between the osteotomy surfaces. (**d**) Acetabular dysplasia with an enhanced anteversion/valgus position of the proximal part of the left femur. (**e**) Result after a Pemberton pelvic osteotomy and a proximal femoral varus external rotation osteotomy

procedure is rarely performed because of the limited correction that can be achieved here.

*Triple osteotomy* Osteotomy through the ilium, superior and inferior ramus of pubis.

*Ganz pelvic osteotomy* The Ganz osteotomy is a technically difficult operation. The osteotomy is done around the acetabulum, avoiding the y-cartilage. For this reason, the operation can only be done if the y-cartilage is closed. A greater correction can be achieved with the Ganz osteotomy than with a triple osteotomy.

*Chiari osteotomy* This osteotomy is performed if there is a lateralization of the femoral head or a discongruent joint (nonspherical head or socket). This is an osteotomy of the iliac bone, after which the femoral head is pressed inward with the distal part of the pelvis. It is a bony coverage without cartilage. The procedure is performed if the previously listed osteotomies aren't possible.

*Shelf arthroplasty* A shelf arthroplasty is performed when there is a discongruent joint. Bone from the iliac crest is grafted onto the iliac bone above the femoral head. In this procedure there is no joint cartilage between the femoral head and the newly covered area either.

*Femoral osteotomy* A proximal femoral varus external rotation osteotomy can be performed when there is a severe enhanced valgus-anteversion position of the proximal femoral part. In this case a wedge of bone is removed from the proximal part of the femur on the medial side after which the distal part of the thigh is adducted and externally rotated and fixed with osteosynthetic material (in this case a hook plate).

**Subluxation/dislocation of the hip up to the age of 6 months** In 80–90 % of cases, repositioning in children up to the age of 6 months can be achieved by using a hip abduction orthosis (Visser hip abduction orthosis) or a Pavlik harness. After 2 weeks with the orthosis or harness,

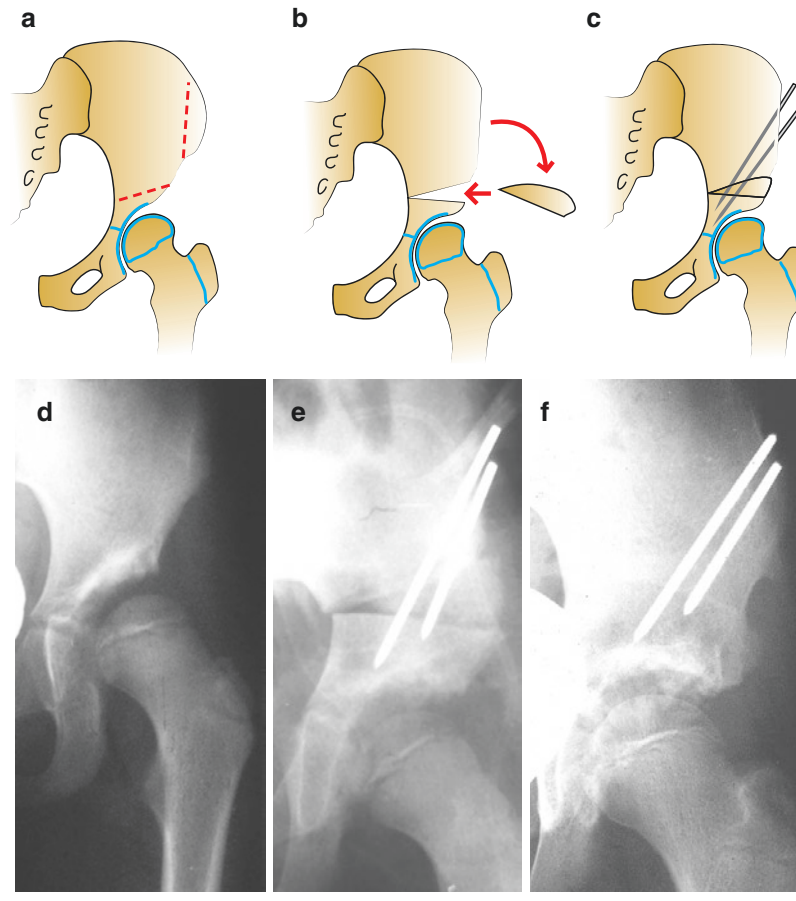



**Fig. 9.25** (**a**–**c**) Schematic representation of a Salter pelvic osteotomy. The piece of bone obtained from the iliac crest is grafted into the openwedge osteotomy and fixed using two Kirschner wires. (**d**) Acetabular dysplasia. (**e**) Situation immediately and (**f**) 3 monthes after a Salter pelvic osteotomy

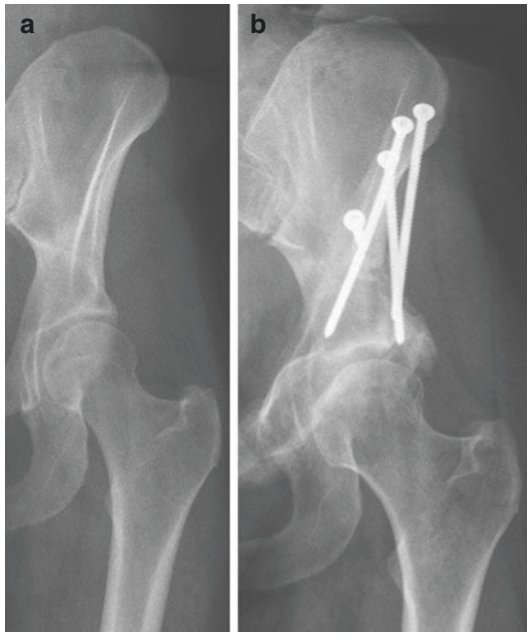



**Fig. 9.26** (**a**) Left-sided acetabular dysplasia in an adult. (**b**) Situation after a Ganz pelvic osteotomy

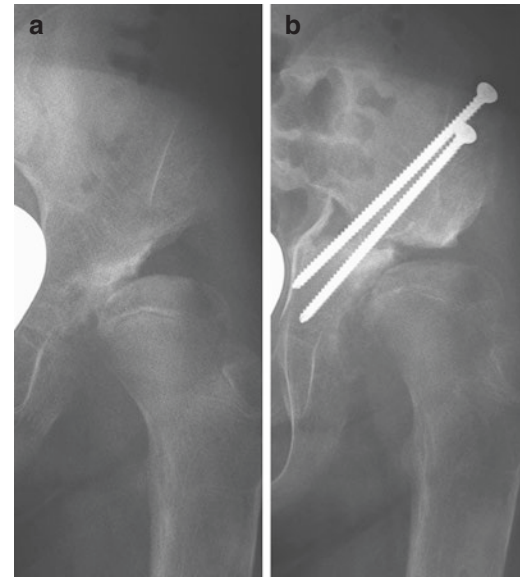



**Fig. 9.27** (**a**) In a congenital hip dislocation it is not always possible to obtain an optimal result. There is a subluxation of the left hip. (**b**) Situation after a Chiari pelvic osteotomy

the success of the reduction can be assessed on an anteroposterior X-ray of the pelvis or an ultrasound of the hip (Fig. 9.29). For the anteroposterior X-ray of the pelvis a X-ray permeable orthosis is used. If reduction is achieved, the abduction treatment with orthosis or harness is continued until the acetabular angle on an anteroposterior X-ray of the pelvis is normalized (as a rule 6 months). If it isn't possible to get the hip reduced (Fig. 9.30), noninvasive reduction under anesthesia is indicated, followed by spica cast immobilization with the hips in 90–100° flexion and 45–60° abduction (Fig. 9.31) for a 3-month period, followed by abduction orthosis treatment until anteroposterior X-ray of the pelvis show a normalized acetabular angle. This treatment is continued until the age of 2 years as needed. In some cases a tenotomy of the adductor longus and psoas muscles is necessary to achieve reduction. Open reduction is rarely necessary at this age (Flowchart 9.1).

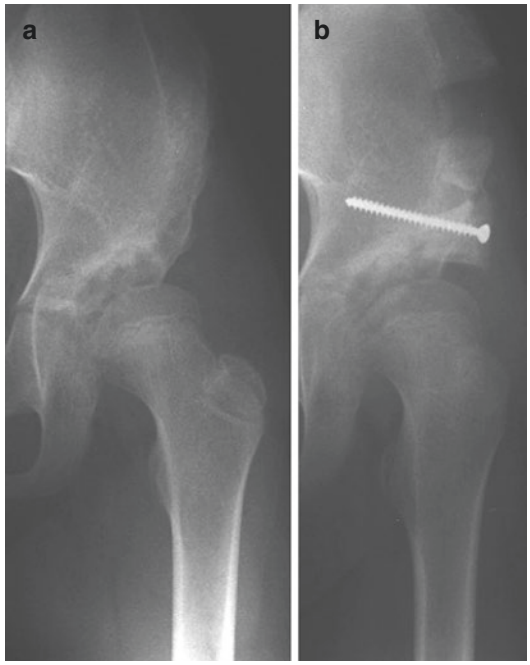



**Fig. 9.28** (**a**) Acetabular dysplasia of the left hip with an enlarged femoral head. (**b**) Situation after a shelf arthroplasty. Fixed with a screw

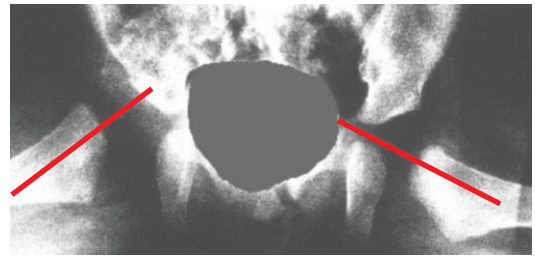


**Fig. 9.30** Anteroposterior image of the pelvis. If the axis of the femoral neck does not point in the direction of the y-cartilage, like it does for the right hip, the hip has not been reduced

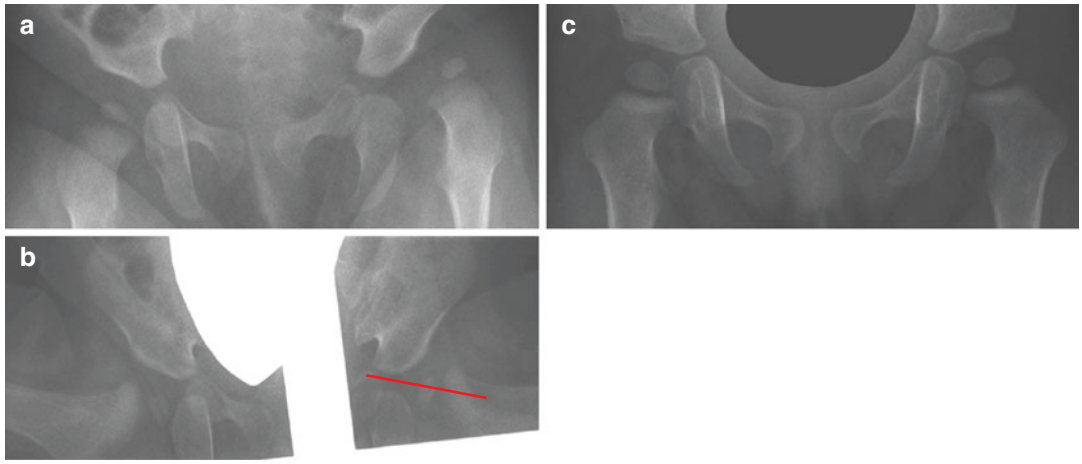
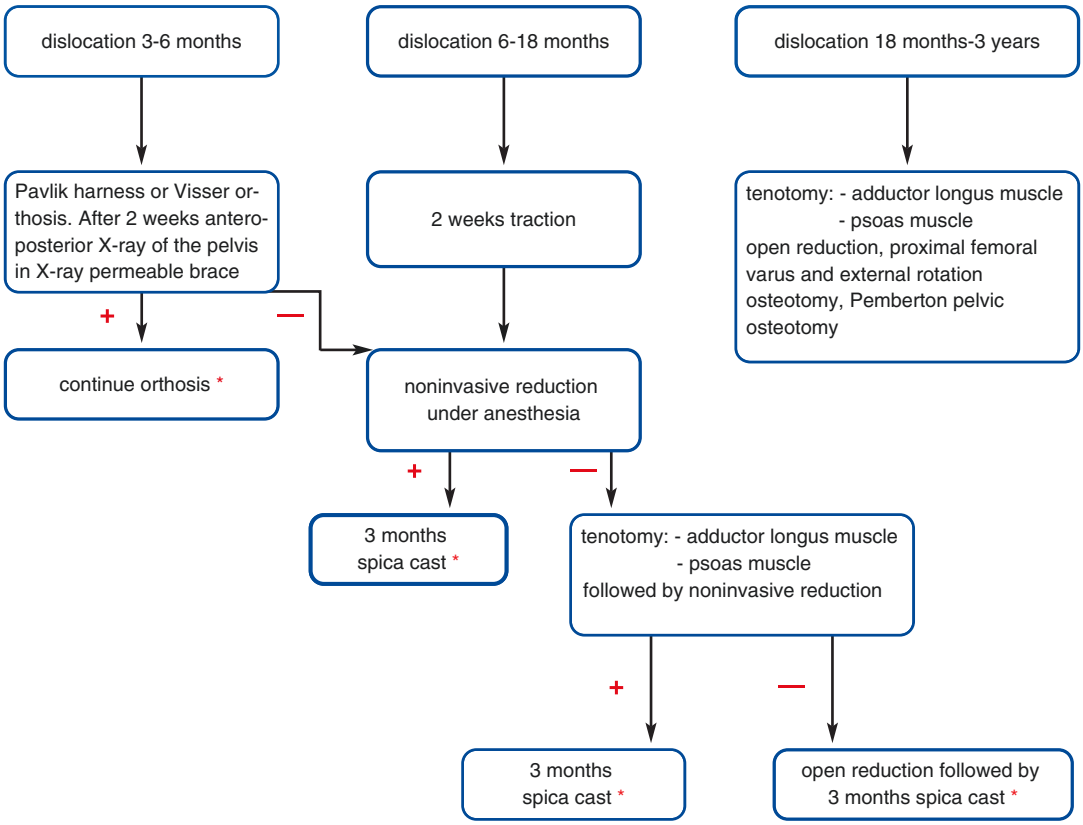



**Fig. 9.29** In 80% of cases of children between the ages of 3 and 6 months reduction can be achieved using a Visser hip abduction orthosis or a Pavlik harness. (**a**) Left hip dislocation. (**b**) Two weeks after starting treatment with a Visser hip abduction orthosis an anteroposterior X-ray of the pelvis (with the orthosis or harness on) can be taken to assess whether reduction has been successful. If the reduction was successful, as is the case here, the axis of the femoral neck points in the direction of the y-cartilage. (**c**) Situation after 6 months

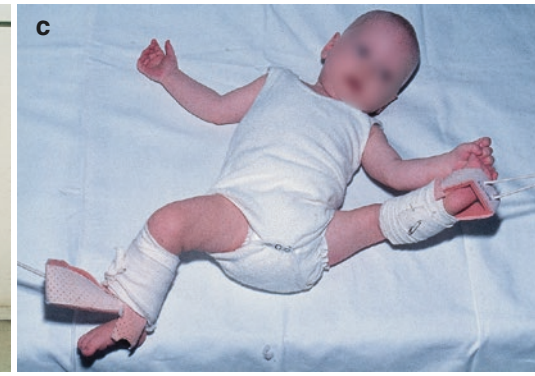
**Subluxation/dislocation of the hip in children 6–18 months** In case of stiff dislocated hips in which the abduction is less than 30°, the adduction contracture is first treated with a balanced suspensory traction. During this traction the hips are flexed 90° in order to eliminate the contracture of the psoas muscle. For a period of 2 weeks the hips are slowly abducted to cancel out the contrac-ture of the adductors (Fig. 9.32). Next, using the Ortolani manoeuvre the hip is reduced under anesthesia and immobilized in 90–100° flexion and 45–60° abduction in a spica cast. If noninvasive reduction of the hip under anesthesia is not successful, a tenotomy of the adductor longus and psoas muscles is performed via a medial incision. If reduction is still unsuccessful (this occurs in less than 5 % of cases), in children younger than 12 months, open reduction takes place through the same incision (Ludloff approach) and in children aged 12 months or older an anterolateral approach is taken (Flowchart 9.1).

**Fig. 9.31** Spica cast with the hips immobilized in 90–100° flexion and 45–60° abduction

In the open reduction through an anterolateral approach the joint capsule is opened in a T-shape on the front side through an anterolateral approach and excess fat tissue in the acetabular fossa and the lengthened and enlarged ligament of the head of the femur (teres ligament) are removed. The




**Flowchart 9.1** Treatment followed by the authors for reduction of a hip dislocation. + reduction, — no reduction obtained, * for treatment (of residual) acetabular dysplasia, see Table 9.6

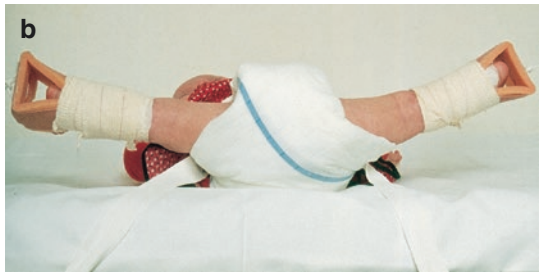




**Fig. 9.32** (**a**) In children aged 6–18 months with a hip dislocation in which the abduction is less than 30°, the adduction contracture is first treated with a balanced suspensory traction. During this traction the hips are flexed 90° in order to eliminate the contracture of the iliopsoas muscle. (**b**) Next, for a period of 2 weeks the hips are slowly abducted to cancel out the contracture of the adductors. (**c**) When the hips are fully abducted, on the dislocated (*left*) side one often sees that the leg is in external rotation and that internal rotation isn't really possible. This is caused by the contracture of the iliopsoas muscle

vascular contribution through the teres ligament is minimal. The transverse acetabular ligament is released. The infolded acetabular labrum is everted and left intact. The hip is reduced and the capsule is reefed. Open reduction will be followed by spica cast immobilization for 3 months and abduction orthosis treatment until the anteroposterior X-ray of the pelvis show a normalized acetabular angle or the child has reached the age of 2 years. Acetabular dysplasia will restore in nearly all cases if reduction takes place before the age of 12 months, and in 50 % of cases if reduction is done between the ages of 1 and 2 years. One generally waits until the age of 4 years before an indication for pelvic osteotomy is made. The enhanced valgus/anteversion position of the proximal femur tends to correct spontaneously.

**Subluxation/dislocation of the hip in children aged 18 months to 3 years** In these cases an adductor longus and iliopsoas tenotomy and an open reduction are performed. If necessary, a proximal femoral varus external rotation osteotomy is done because of an enhanced valgus and anteversion position of the proximal femur. If there is a high degree of dislocation the femur is shortened by as much as the femoral head rises above the edge of the acetabulum. Finally, a Pemberton or Salter pelvic osteotomy is performed (Figs. 9.23 and 9.24). In children older than 2½ years with a bilateral dislocation it is prudent to operate on one hip at a time (Flowchart 9.1).

**Subluxation/dislocation of the hip in children older than ages 3–5** In children older than the 3–5-year-old range this almost always involves a bilateral hip dislocation. In such cases it is extremely difficult to get the femoral head to fit into the acetabulum and keep it there. One should consider whether the means are actually worse than the malady. In most cases it is better to accept the situation as it is, especially in bilateral dislocations. A complete dislocation does not cause any pain, but the child will have a waddling gait.

**Complications** A complication of the treatment is ischemic femoral head necrosis (Figs. 9.33, 9.34, 9.35, and 9.36). This complication never occurs in

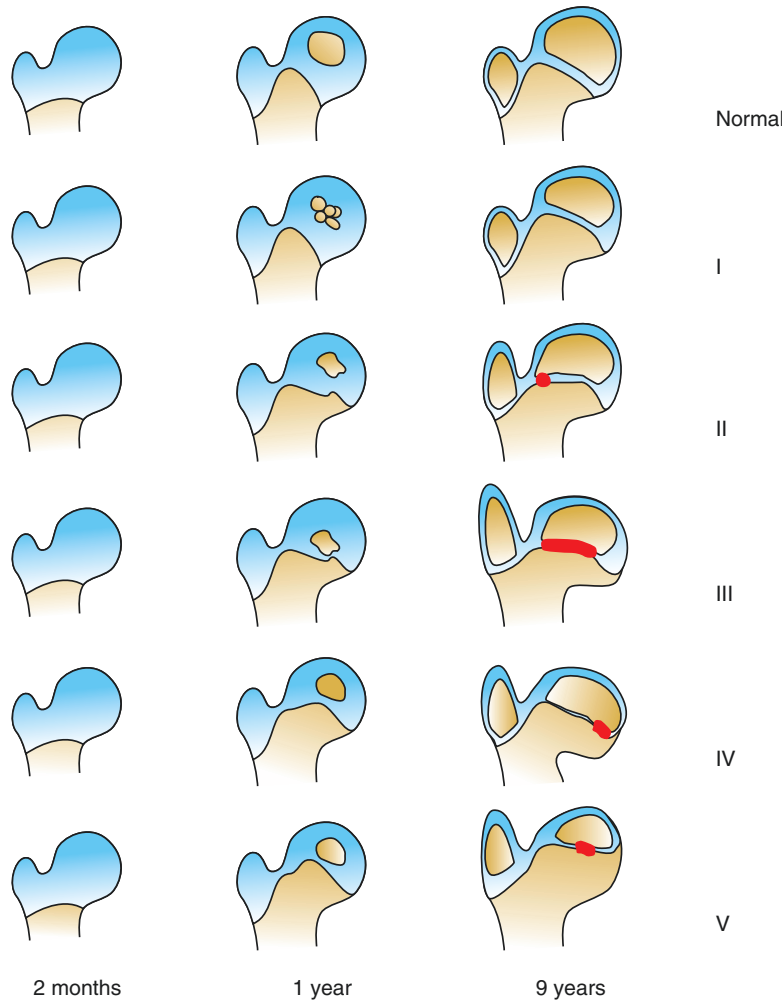



**Fig. 9.33** Classification of ischemic femoral head necrosis according to Buchholz and Ogden 1978, I-IV. Type V was added in 1980 by Kalamchi and McEwan (see literature). *Type I*: Temporary ischemic femoral head necrosis. As a consequence of the circulatory disorder of the femoral head an irregular ossification of the epiphysis develops which ends up restoring itself fully. *Type II*: Lateral ischemic femoral head necrosis. A fusion occurs on the lateral side of the growth plate of the femoral head, causing a valgus position of the femoral head with respect to the femoral neck; the growth in length of the femoral head is curbed, but the upper edge of the femoral head remains cranial to the apex of the greater trochanter. *Type III*: Full ischemic femoral head necrosis. There are severe changes in the epiphysis and metaphysis and of the irregular edges of the growth plate. The ossification center of the epiphysis does not appear before 1 year after reduction, and if a core is present it will not increase in size the first year after reduction. A severe shortening of the femoral neck occurs with a varus position of the femoral head, as well as a considerable overgrowth of the greater trochanter, whose apex lies significantly higher than the upper edge of the femoral head. *Type IV*: Medial ischemic femoral head necrosis. An osseous bridge develops in the epiphysis on the medial side of the femoral head, causing a varus position. There is relative overgrowth of the greater trochanter, but its apex does not rise up above the upper edge of the femoral head. *Type V*: Central ischemic femoral head necrosis. There is an irregular central part in the epiphysis that leads to a reduced growth of the femoral head and a relative overgrowth of the greater trochanter, but the greater trochanter remains at the same level as the upper edge of the femoral head

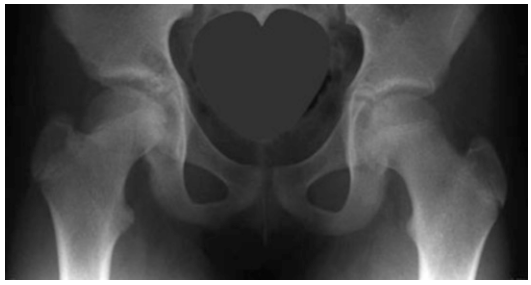


**Fig. 9.34** Ischemic femoral head necrosis type II of the right hip

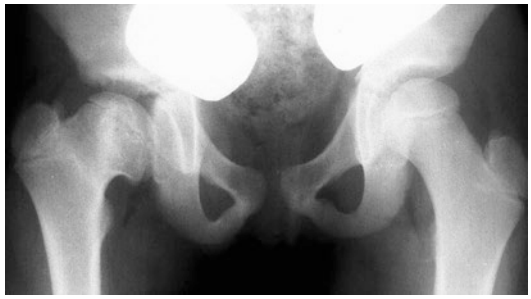


**Fig. 9.35** Ischemic femoral head necrosis type IV of the right hip

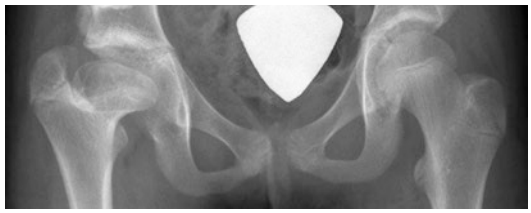


**Fig. 9.36** Ischemic femoral head necrosis type III of the right hip

an untreated hip dislocation. Treatment using a Visser orthosis or Pavlik harness is responsible for a low percentage of ischemic femoral head necrosis. Forced abduction should be avoided because otherwise chances increase for ischemic femoral head necrosis. Traction treatment reduces the chances of ischemic femoral head necrosis, operative treatment increases them. If after reduction the hips are immobilized in a spica cast in more than 70° abduction, risks of ischemic femoral head necrosis are very high, so this should always be avoided. Percentages up to 70% may occur.

Ischemic femoral head necrosis occurs with forced abduction due to compression of the medial circumflex femoral artery or damage caused by an operation. A proper classification should be identifiable on X-rays whithin 2 years of treatment of developmental dysplasia of the hip. A large percentage of patients will not require any treatment during adolescence and young childhood. In a small percentage of patients, femoral head deformity and acetabular dysplasia can be treated with either femoral osteotomy or appropriate acetabuloplasty or both. Up to the age of 8 overgrowth of the greater trochanter can be treated by an epiphysiodesis and in adulthood with a transfer of the greater trochanter distally for improving abductor strenght. Severe leg lenght inequality can be corrected by appropriate techniques (see Chap. 15).

In a follow-up of 12 years, 17% of children treated for a hip dislocation with a perfect result at age 3 still develop acetabular dysplasia (for treatment, see pp. 85–87).

## The Legs Can Not Be Approximated or with Pelvic Obliquity

Complaint: the parent cannot bring the newborn child's legs close together.

Assessment: one or both legs are in abduction. If, in a unilateral anomaly, an attempt is made to bring the legs together, a pelvic obliquity occurs as well as a compensatory scoliosis with the convex side on the side of the leg with the abduction contracture (Fig. 9.37). In a bilateral condition the legs cannot be approximated.

Diagnosis: **congenital abduction contracture of the hip**

Explanatory note: the contracture can be unilateral or bilateral. This condition is caused by an abnormal position in the uterus. Accompanying postural irregularities can be torticollis, adduction contracture on the contralateral side, and foot anomalies such as talipes calcaneovalgus.

Supplementary assessment: anteroposterior X-rays of the pelvis show no abnormality.

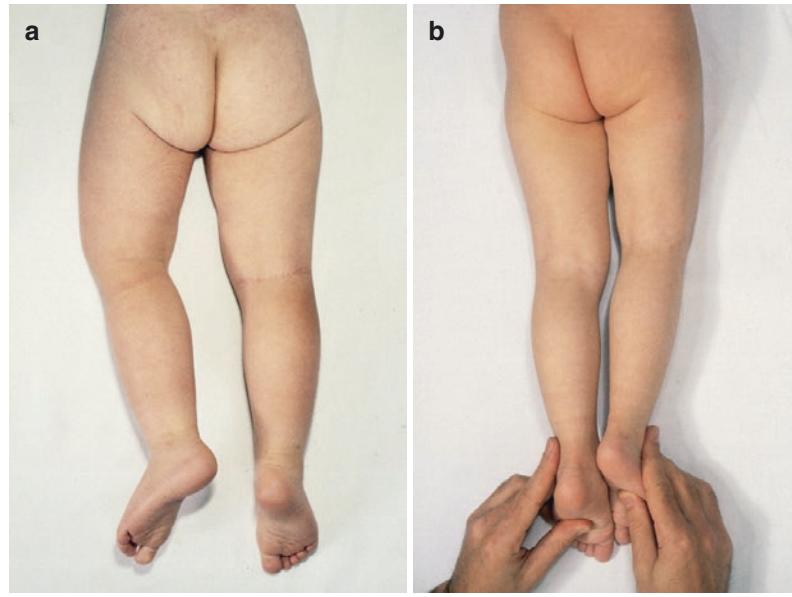



**Fig. 9.37** (**a**) Abduction contracture of the left hip. (**b**) If an attempt is made to bring the legs together in a unilateral abduction contracture, a pelvic obliquity will occur and a compensatory scoliosis with the convex side towards the leg with the abduction contracture, in this case the left side

Primary care treatment: treatment should be started as promptly as possible, preferably the first 2 weeks after birth. The physiotherapist must teach the mother stretching exercises, in order to adapt the hip passively. The stretching exercises have to be done with 20 repetitions, six times a day. If these stretching exercises are started early, a contracture will disappear within 8 weeks.

When to refer: when stretching exercises do not help eliminate the adduction contracture of the hip.

Secondary care treatment: **congenital abduction contraction of the hip.** A spica cast that immobilizes the involved hip in adduction, extension and internal rotation for a period of 4 weeks.

## Waddling Gait

Complaint: the child waddles like a duck.

Assessment: the air space between both legs just under the perineum is widened (thigh gap). The Trendelenburg test is positive (Fig. 9.17).

Differential diagnosis:
**coxa vara**
congenital coxa vara
acquired coxa vara
**bilateral hip dislocation**

Explanatory note: **coxa vara.** In coxa vara there is a reduced angle between the femoral head and the femoral shaft (<120°). Because of this varus position, abduction of the hips is reduced and adduction enhanced. The anomaly affects 1 in 25,000 children. Boys and girls are affected equally, as are the right and left sides. An apparent limb shortening (usually less than 3 cm) or painless limp may be present in unilateral cases. A waddling gait is more characteristic in bilateral cases. One-third of cases presents with bilateral involvement. There are two types of coxa vara.

*Congenital coxa vara* This type is rare and is present at birth. It is often accompanied by other congenital anomalies such as a shortened femur (femoral hypoplasia), proximal focal femoral deficiency or cleidocranial dysostosis. This type tends to be recognized early because of the accompanying anomalies.

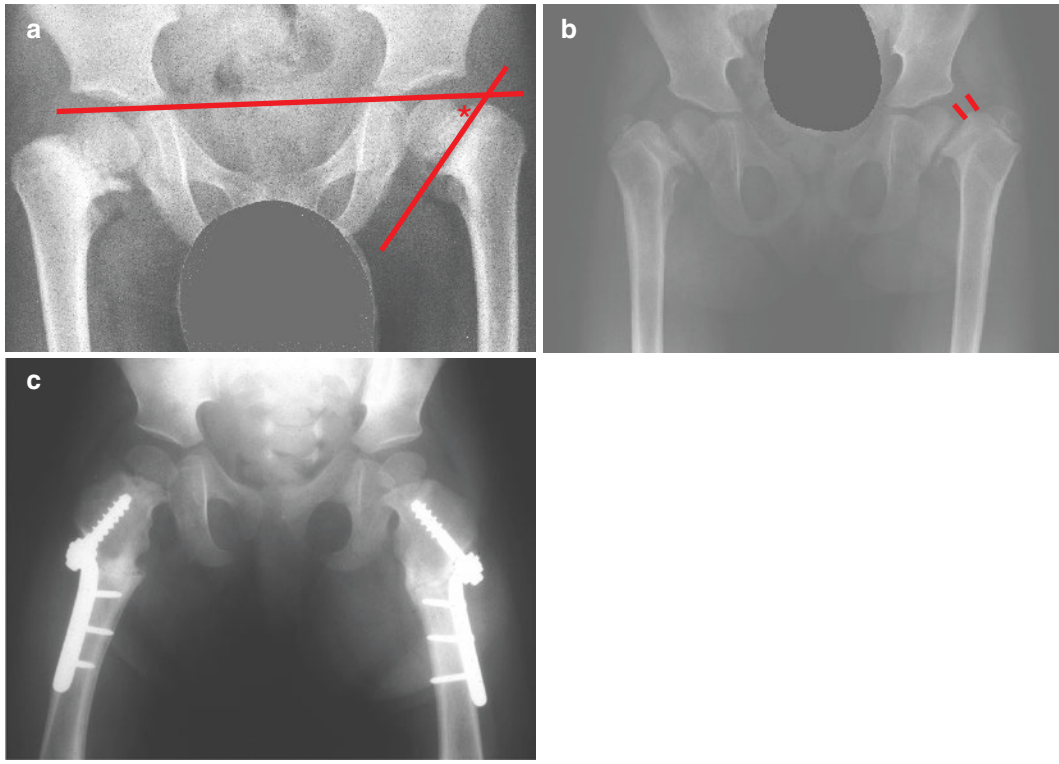



**Fig. 9.38** Bilateral coxa vara. (**a**) In coxa vara there is a reduced angle between the femoral shaft and the femoral neck (varus position). The angle between the Hilgenreiner line and the epiphyseal plate is called the Hilgenreiner epiphyseal angle (*). If this angle exceeds 60° the varus deformity will be progressive. (**b**) With a Hilgenreiner epiphyseal angle larger than 60° the epiphysis shifts distally with respect to the metaphysis, thus increasing the varus position of the proximal femur. (**c**) Proximal femoral valgus osteotomy in coxa vara

*Acquired coxa vara* This type is usually recognized between the ages of 3 and 5 years. As the child grows and becomes heavier, the epiphysis slowly glides downwards and the angle between the femoral shaft and the femoral neck, the caput collum diaphysis (CCD) angle, decreases, so that the highest point of the trochanter major ends up lying proximal to the femoral head.

In 50 % of cases there is a positive Trendelenburg test if the highest point of the greater trochanter lies equally high as the upper edge of the femoral head. In nearly 100 % of cases there is a positive Trendelenburg test (Fig. 9.17) if the proximal part of the greater trochanter lies higher than the upper edge of the femoral head.

**Bilateral hip dislocation** For explanatory note, supplementary assessment, primary care treatment, when to refer and secondary care treatment, see Chap. 9.

Supplementary assessment: Anteroposterior X-ray of the pelvis. In this image the angle between the Hilgenreiner line and the epiphyseal growth plate of the femoral head is measured (Fig. 9.38).

Primary care treatment: None.

When to refer: A child with a waddling gait should be referred for further assessment and possible treatment.

Secondary care treatment: **congenital and acquired coxa vara.** A proximal femoral valgus osteotomy is performed if the angle between the femoral head and the femoral shaft is less than 110°, if the Hilgenreiner epiphyseal angle is larger 60°, and if the varus deformity is progressive (Fig. 9.38). If the Hilgenreiner epiphyseal angle is smaller than 45°, the varus deformity will restore itself spontaneously. In the end there will be a normal angle between

the femoral head and the femoral shaft. If the Hilgenreiner epiphyseal angle is between 45 and 60°, an anteroposterior X-ray of the pelvis should be taken every 6 months. A wait-and-see policy can be adopted if there is no progression in the varus deformity, if there is progression, a proximal femoral valgus osteotomy should be performed. A varus deformity of the hip may recur after this operation. For this reason, the child should be regularly checked until growth is completed.

## Snapping Hip

Complaint: there is a snapping of the hip that the patient can feel and sometimes hear. The patient sometimes has the feeling that the hip is temporarily "stuck" (lock phenomenon).

Assessment: the patient can usually demonstrate the snapping itself, but not always.

Differential diagnosis:

**snapping of the iliotibial tract**
**snapping of iliopsoas tendon**
**recurrent hip subluxation/dislocation**
voluntary hip subluxation/dislocation
habitual hip subluxation/dislocation
**osteochondritis dissecans of the femoral head**
**acetabular labral tear**

Explanatory note: **snapping of the iliotibial tract.** When walking, the iliotibial tract is seen and felt to snap over the greater trochanter (Fig. 9.39). It mostly affects children at puberty. The snapping tends to be painless and children sometimes experience it as if the hip is moving out of its socket. The snapping of the iliotibial tract can best be observed in a walking child while the examiner places his hand on the greater trochanter region. In a non-weight-bearing assessment, snapping of the iliotibial tract is difficult to incite.

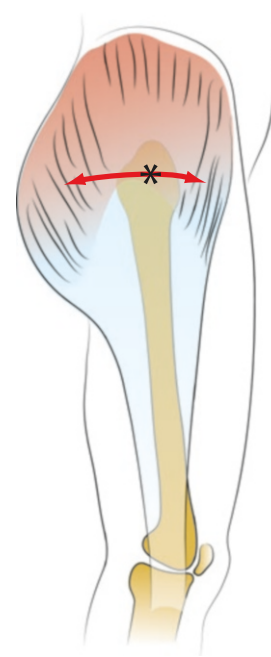


**Fig. 9.39** Snapping of the iliotibial tract over the greater trochanter (*) (Redrawn from: Dandy DJ. Essential Orthopaedics and Trauma. 2nd ed. Edinburch/London/Madrid/Melbourne/New York/Tokyo: Churchill Livingstone; 1993. p. 391)

**Snapping of the iliopsoas tendon** In a weight-bearing leg, as the hip goes from flexion into extension the iliopsoas muscle snaps over the anterior side of the joint capsule and the iliopectineal (or iliopubic) eminence (Fig. 9.40). There is also an irritated bursa between the iliopsoas tendon and the joint capsule if the snapping causes pain. Pressure pain on the anterior side of the hip joint may be present.

**Recurrent hip subluxation/dislocation** The only symptom given tends to be a snapping hip. The subluxation/dislocation is not painful. No adequate trauma has occured.

*Voluntary hip subluxation/dislocation* This type is due to hypermobility disorders such as in Down[2] and Ehlers-Danlos syndromes[2]. This dislocation can occur posteriorly as well as anteriorly.

*Habitual hip subluxation/dislocation* This is a subluxation/dislocation in normal children with no associated ligamentous laxity. The dislocation is most commonly posterior. The anomaly is six times more frequent in females than in males.

[2]See Appendix.

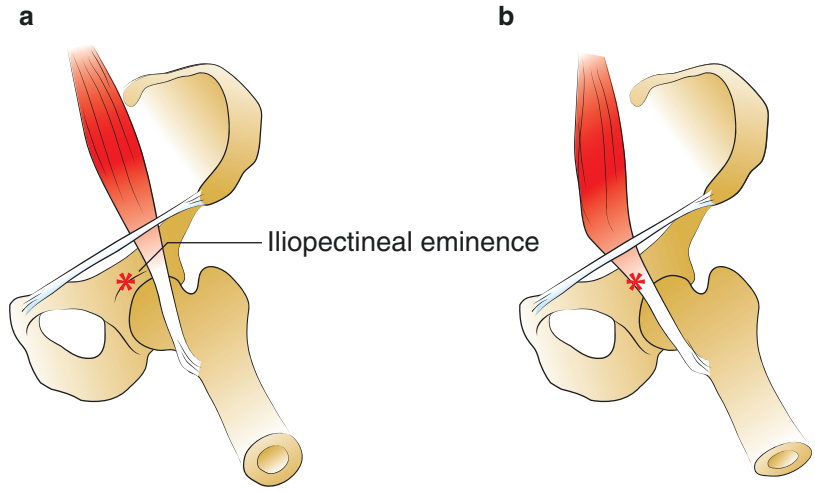



**Fig. 9.40** Snapping of the iliopsoas tendon. In the weight-bearing leg, as the hip goes from flexion into extension the iliopsoas muscle snaps over the anterior side of the joint capsule and the iliopectineal (or iliopubic) eminence. (**a**) Flexion. (**b**) Extension. The psoas muscle shifts medially in extension, snapping over the anterior side of the joint capsule and the iliopectineal (or iliopubic) eminence (*). This eminence is a curvature of the pelvis in the proximal part of the pubic bone on the medial upper side of the acetabulum

The onset of symptoms was noted before the age of 9 years and most commonly between the age of 2 and 4 years.

**Osteochondritis dissecans of the femoral head** This is a condition in which an area of necrotic subchondral bone occurs as a result of loss of vascularisation. The cause is unknown. Necrotic subchondral bone lesions can occur in various joints (elbow, hip, knee and ankle, and the first metatarsophalangeal joint). Osteochondritis dissecans is most common in the knee and elbow. The hip anomaly is mostly localized in the superolateral part of the femoral head. It is relatively common in sickle cell anemia, multiple epiphyseal dysplasia[2], Gaucher disease, and in 3 % of patients with Legg-Calvé-Perthes disease (Fig. 9.63). Symptoms vary greatly: some individuals have none and others complain about snapping of the hip, locking symptoms, and a feeling of sagging through the hip.

**Acetabular labral tear** This can occur spontaneously during puberty, but an accident is often involved. Tears of the acetabular labrum may also appear in acetabular dysplasia, Legg-Calvé-Perthes disease, and after a slipped capital femoral epiphysis.

Patients complain about locking symptoms and snapping of the hip. Ninety percent of the cases diagnosed with acetabular labral tears have had complaints of pain in the groin. This can be an indication for an anterior labral tear, whereas buttock pain is more consistent with posterior tears and less common. These symptoms can increase when the patient bears weight or performs twisting movements in the hip. Pain may also occur while climbing stairs.

The tests for an acetabular labral tear are as follow:

**Anterior acetabular labral tear test** The patient lies in supine position and brings the hip in to full hipflexion, abduction and external rotation. The limb is than passively moved toward hipextension as the leg is simultaneously adducted and internally rotated. Patients with a anterior labral tear will experience sharp catching pain with or without an audible snapping.

**Posterior acetabular labral tear test** The patient lies in prone position with the knee extended and the hip extended, abducted and externally rotated. The examinor passively extends, adducts and internally rotates the hip. Sharp catching pain with

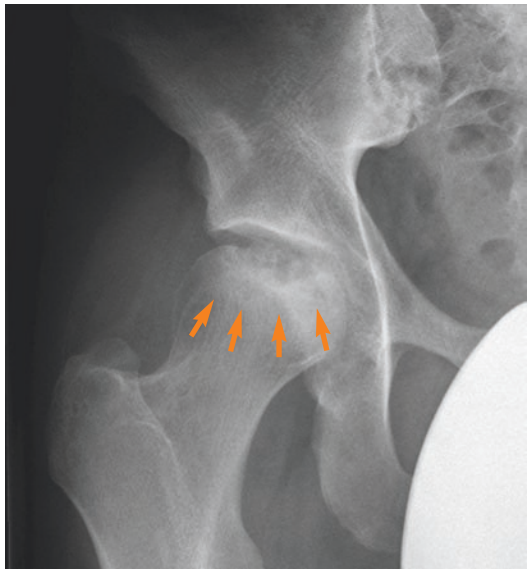


**Fig. 9.41** Osteochondritis dissecans of the right hip

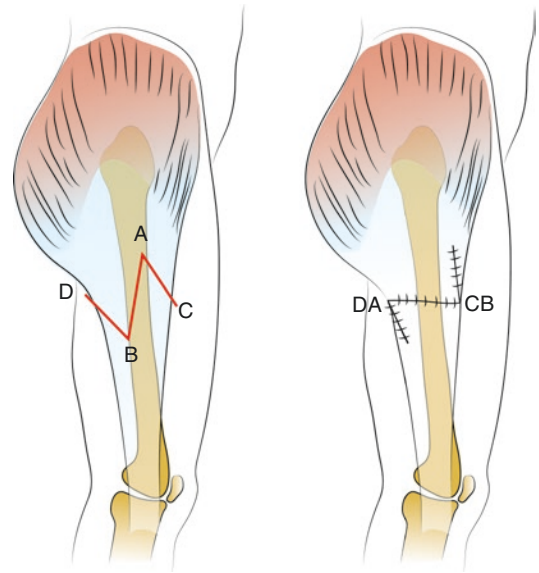



**Fig. 9.42** Lengthening of the iliotibial tract by means of a Z-plasty (Redrawn from: Brighall CG, Stainsby GD. The snapping hip treatment by Z-plasty. J Bone Joint Surg. 1991;73 B:253–54)

or without snapping will be an indication for a posterior labral tear. The pain disappears for a short while after injecting the hip joint with a local anesthetic.

Supplementary assessment: no further assessment is needed for iliotibial tract snapping. Snapping of the iliopsoas tendon can best be depicted on ultrasound. If there is an irritated bursa on this examination one can also get an impression about the size of this bursa. On an anteroposterior X-ray an osteochondritis dissecans (Fig. 9.41) and a non-reduced voluntary or habitual hip dislocation can be recognized. If a tear of the acetabular labrum is suspected, a MRA should be requested.

Primary care treatment: a wait-and-see policy can be adopted if the snapping of the iliotibial tract is not painful. Some children can avoid it by changing their gait pattern. If there is a painful snap as a result of a snapping iliotibial tract, NSAID's can be prescribed or the area around the iliotibial tract can be injected with corticosteroids.

If snapping of the iliopsoas tendon is painful, NSAID's can be prescribed at first. An injection into an irritated bursa between the iliopsoas tendon and the anterior side of the joint capsule is technically difficult. This treatment must be left to secondary care. In habitual hip subluxation/dislocation the parents are encouraged to persuade the child not to dislocate the hip. Spontaneous cuorrection mostly occurs after about 2 years after the initial subluxation/dislocation.

When to refer: when a snapping iliotibial tract or iliopsoas tendon does not react well to conservative treatment. Referral is indicated for a voluntary hip dislocations, suspected osteochondritis dissecans and tears of the acetabular labrum.

Secondary care treatment: **snapping of the iliotibial tract.** After surgically treating a snapping hip as a result of a snapping iliotibial tract, no guarantee can be given that the patient will be symptom-free after the operation. The operation involves elongation of the iliotibial tract with a Z-plasty (Fig. 9.42).

**Snapping of the iliopsoas tendon** Guided by an ultrasound or X-ray illumination, an injection

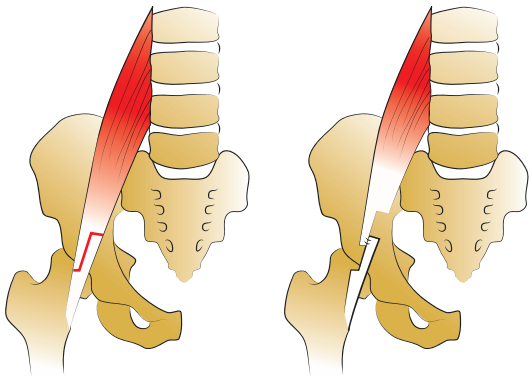


**Fig. 9.43** Z-shaped lengthening of the iliopsoas tendon

with corticosteroids can be given into the irritated bursa between the iliopsoas tendon and the front side of the joint capsule. A Z-shaped lengthening of the iliopsoas tendon can be considered if this gives insufficient results (Fig. 9.43).

**Voluntary subluxation/dislocation** For a voluntary hip dislocation, young children can first be treated with a hip abduction orthosis for 6 months. If dislocations still occur after that, the joint capsule can be reefed — in an anterior dislocation on the anterior side and in a posterior dislocation on the posterior side. A proximal femoral external rotation osteotomy can be performed for a recurrent posterior dislocation or a Salter pelvic osteotomy for a recurrent anterior dislocation if there are still dislocations after reefing.

**Osteochondritis dissecans of the femoral head** One can wait and see what happens as long as there is no loose body. Hip loading activities and sports such as volleyball, basketball and soccer are discouraged. In most cases the anomalies disappear spontaneously. In a small percentage of cases there is a loose body that can be removed with an arthroscopy or an arthrotomy of the hip joint.

**Acetabular labral tear** A tear that is mostly located on the anterior side of the hip joint can be rounded off arthroscopically or with an arthrotomy, just as in any existing cartilaginous injury. Suturing of the tear may be considered if the acetabular labrum of the bony part of the acetabulum is torn and there is a sort of "bucket handle" injury.

## Hip Pain

### Vague Pain Around Groin or Pelvis or Upper Leg

Complaint: the pain around the groin, pelvis or thigh keeps changing and occurs mainly after doing sports. There can be night pain; the child is not sick and can fully move the hip.

Assessment: sometimes there is light pressure pain on the pelvis, hip or thigh.

Differential diagnosis:

**bone tumor**
**ischiopubic osteochondritis (Van Neck disease)**

Explanatory note: **bone tumor** (Table 9.7)

The most common bone tumor of the hip is an osteoid osteoma. This is a benign tumor, located in the cortex, intraosseously, or sometimes beneath the periosteum. Usually this tumor occurs between 10 and 25 years of age. In 75 % of cases it affects males. The tumor is accompanied by a deep pain and it cannot be palpated. There is nocturnal pain in half of cases. The tumor reacts well to salicylates.

**Ischiopubic osteochondritis** Around the cartilage junction between the inferior pubic and ischial rami there is a bony thickening. This affects children between ages 4 and 12, with pain in the groin and/or buttocks. In 30 % of cases there is an antalgic gait. The symptoms disappear spontaneously. This anomaly can be confused with an osteomyelitis, but in contrast to osteomyelitis there tends to be no fever and the infection parameters are negative.

**Table 9.7** Bone tumors at the level of the pelvis and femur. The § sign identifies tumors that aren't very common

| Location | Benign bone tumors | Malignant bone tumors |
|---|---|---|
| Pelvis | Eosinophilic granuloma | Ewing sarcoma |
| | Aneurysmal bone cyst | Osteosarcoma § |
| | Osteochondroma | |
| | Fibrous dysplasia | |
| | Endochondroma § | |
| | Osteoid osteoma § | |
| | Solitary bone cyst § | |
| | Chondroblastoma § | |
| | Primary bone lymphoma § | |
| Femoral neck | Aneurysmal bone cyst | |
| | Chondroblastoma | |
| | Fibrous dysplasia | |
| | Osteochondroma | |
| | Osteoid osteoma | |
| | Solitary bone cyst | |
| Femoral shaft | Eosinophilic granuloma | Ewing sarcoma |
| | Osteoid osteoma | Osteosarcoma § |
| Distal part of the femur | Nonossifying fibroma | Osteosarcoma |
| | Osteochondroma | |

Based on Adler CP, Kozlowski K. Primary bone tumors and tumorous conditions in children. London: Springer; 1993

Supplementary assessment: anteroposterior X-rays of the pelvis and lateral view of the hip involved. The classical picture of an osteoid osteoma is a round radiolucent area with a 1–1.5 cm diameter, called a nidus (Latin for nest), surrounded by a zone of sclerosis. If the nidus diameter is larger than 1.5 cm, one speaks of a osteoblastoma. Histopathologically there is no difference between an osteoid osteoma and an osteoblastoma.

In a cortical localization this sclerotic reaction can be so severe that the nidus is masked on X-rays. The intraosseous variety is mostly localized in the femoral neck, on the medial side, but can also appear in other places (Fig. 9.44). As a result of increased activity of the bone in the area around the tumor, a valgus deformity of the femoral neck, an increase in length of the proximal part of the femur or a caput magnum can occur due to growth stimulation. Subperiostal osteoid osteomas are the third and least common variety, and usually appear in the talar bone. A technetium scan shows strongly enhanced activity around the tumor. In an ischiopubic osteochondritis anteroposterior X-rays of the pelvis show a bony thickening around the junction between the inferior pubic and ischial rami (Fig. 9.45). If there is doubt between ischiopubic osteochondritis and osteomyelitis the infection parameters (CRP and ESR) can be determined. If one of the infection parameters is positive, an MRI tends to offer an answer.

Primary care treatment: there is growing evidence that an osteoid osteoma resolves spontaneously over time and can be treated with aspirin or NSAID's. Average time to resolution is 75 months. Range 24–180 months. Van Neck disease will heal spontaneously.

When to refer: when the osteoid osteoma does not respond to aspirin or NSAID's.

Secondary care treatment: **osteoid osteoma.** Radio frequency ablation under CT guidance.

## Hip Pain and Limited Internal Rotation

Complaint: the child complains about pain in the groin, thigh or knee, and the gait is irregular. The pain is not so severe that the child refuses to walk.

Assessment: the presenting complaint is usually pain in the region of the groin, which may be referred to the anteromedial aspect of the thigh and knee; 15 % of

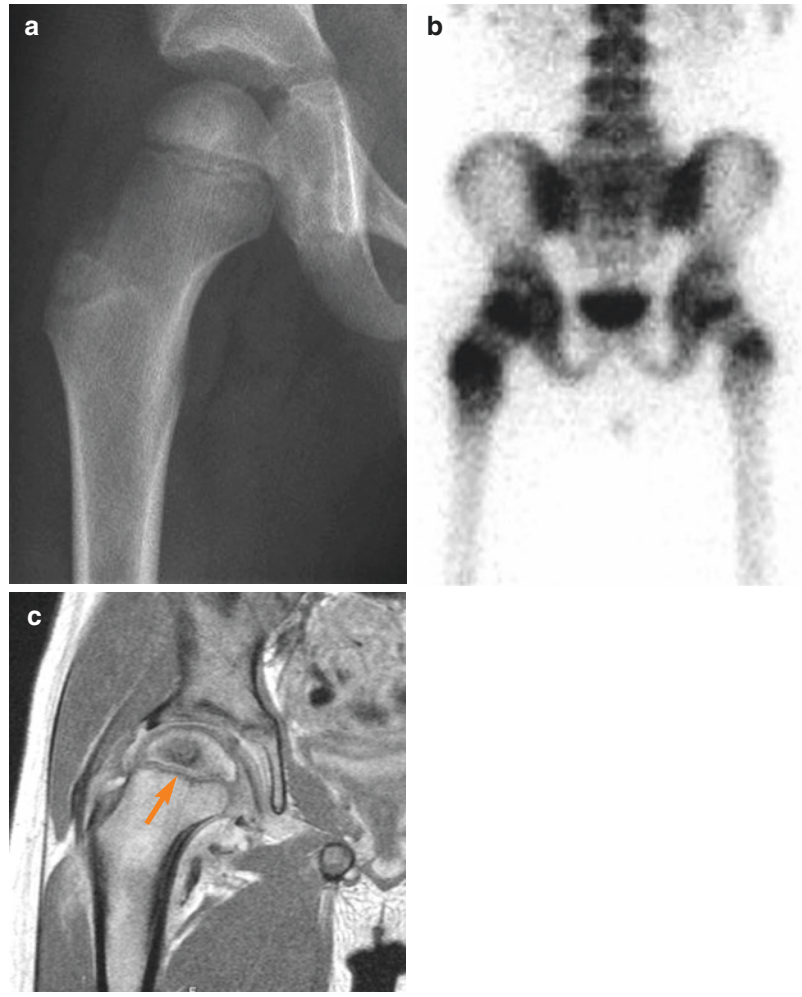



**Fig. 9.44** (**a**) Anteroposterior X-ray of the right hip with an osteoid osteoma in the femoral head. (**b**) A bone scan shows increased activity in the right femoral head epiphysis. (**c**) The osteoid osteoma can be seen better on an MRI than on an X-ray

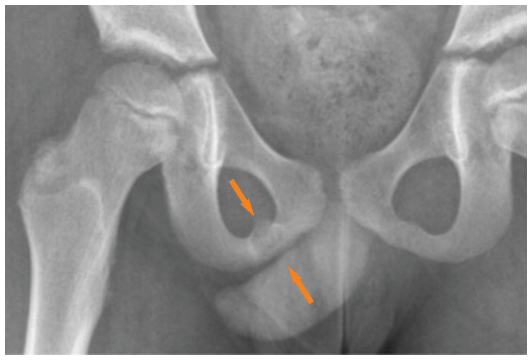


**Fig. 9.45** Right-side ischiopubic osteochondritis also called Van Neck disease (*arrows*)

children complain of pain only in the distal thigh or knee. The child may have an antalgic gait[3] (ant is Latin for against and algos Greek for pain). This can happen in two ways: by making the standing phase as short as possible or by leaning with the upper body over the painful hip during the standing phase of the affected leg[3]. The singular movement that will first become limited is the internal rotation. Limited internal rotation of the hip joint can be most accurately determined with an extended hip. The simplest way to check this is with the child lying prone. The hips are extended and the knees are flexed 90°. The left and right internal rotation of the hip joint can be compared by moving the lower legs outwards (Fig. 9.46). There is a hip problem if there is limited and/or painful internal rotation in the end trajectory. The composite movement in which adduction takes place with a hip

[3]See Addendum

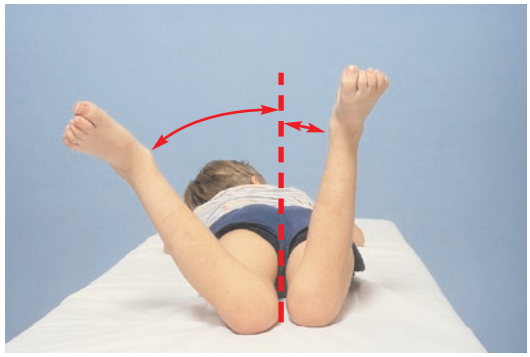


**Fig. 9.46** The internal rotation of the right hip is limited

flexed at 90° becomes limited even earlier than the internal rotation.

Differential diagnosis:

**acute transient synovitis of the hip (coxitis fugax, irritable hip syndrome, observation hip, toxic synovitis, acute transient epiphysitis)**
**Legg-Calvé-Perthes disease (Perthes disease, coxa plana, Waldenström disease)**
**chronic slipped capital femoral epiphysis**

Explanatory note: **acute transient synovitis of the hip.** An acute transient synovitis of the hip causes a painful hip. The symptoms disappear after several days of bed rest. The cause of a coxitis fugax is unknown. Sometimes it develops right after an airway infection or an innocent trauma. Occasionally the condition is recurrent. The clinical picture can also constitute the initial symptoms of Legg-Calvé-Perthes disease. The literature indicates that about 2 % of cases in which a coxitis fugax is suspected are in the initial stages (synovitis phase) of Legg-Calvé-Perthes disease. A transient synovitis of the hip occurs between the second and thirteenth year of life, in 80 % of cases between ages 3 and 6. The ratio between boys and girls varies in the literature from 3:2 to 5:1. In about 5 % of cases there is a bilateral involvement. The child is not sick and the affected leg is not severely painful as is the case in a bacterial infection of the hip joint. The child might have an antalgic gait[4]. Sometimes there is a subfebrile temperature, a slightly elevated WBC count and slightly elevated CRP or ESR levels. X-rays show no abnormalities. The diagnosis is established on an exclusionary basis.

**Legg-Calvé-Perthes disease** In 1910, Legg from the United States, Calvé from France and Perthes from Germany discovered this hip abnormality independently. In 1909 the anomaly had already been noticed by Waldenström in Sweden, but he thought it was tuberculosis. Legg-Calvé-Perthes disease is not to be confused with tuberculosis, although in some situations the radiological images resemble Perthes disease (Table 9.8). In most of these anomalies, on the basis of the clinical picture it can be deduced that there is no Legg-Calvé-Perthes disease. An exception to this is Meyer dysplasia. In Meyer dysplasia the osseous core of the femoral head-epiphysis appears only after age 2 and consists of several parts. This resembles Legg-Calvé-Perthes disease in the fragmentation stage. Meyer dysplasia should be suspected under the age of 4 years, especially if the anomaly is bilateral (Fig. 9.47 and Table 9.9).

Legg-Calvé-Perthes disease affects about 1 in 10,000 children — boys four to five times as often as girls — and is bilateral in 10–12 % of cases. The disease appears between the ages of 2 and 13, in 80 % of cases between ages 4 and 8. In 1.6–20 % of cases it is a familial anomaly. If Legg-Calvé-Perthes disease runs in the family, chances of a new case within the same family are 3 %. For a long time it was thought that Legg-Calvé-Perthes disease was caused by a reduced supply of blood to the femoral head. In more recent years the explanation has gravitated towards a thrombotic venous occlusion of the femoral head, because 75 % of children with Legg-Calvé-Perthes disease have an antithrombin protein-C or S deficiency, a hypofibrinolysis or high lipoprotein A levels, which can cause this affliction. Children with Legg-Calvé-Perthes disease tend to be smaller and heavier than their age peers.

[4] See Chap. 16 and Addendum.

**Table 9.8** Anomalies with radiological findings of the femoral head that resemble Legg-Calvé-Perthes disease

| Category | Disease | Remarks |
|---|---|---|
| Syndrome | Mucopolysaccharidosis[a] | The following applies to these four syndromes:<br>Mostly bilateral<br>Both femoral heads are symmetrically irregular |
| Developmental variant of the epiphysis | Gaucher disease | |
| | Multiple epiphyseal dysplasia[a] | |
| | Spondyloepiphyseal dysplasia[a] | |
| | Meyer dysplasia | See Table 9.9 |
| Circulatory disorder | Developmental dysplasia of the hip | Appears only after treatment, due to forced abduction (obstruction of blood supply) or surgery (damaged blood supply) |
| Hematogenic | Hemophilia | |
| Infectious | Lupus erythematosus | Treated too late |
| | Sickle cell anemia | |
| | Septic arthritis | |
| Metabolic<br>Traumatic | Hyperthyroidism<br>Femoral neck fracture | Most common cause: ischemic femoral head necrosis |
| | Hip dislocation | |
| Tumor | Slipped capital femoral epiphysis<br>Lymphoma | |
| Medicinal | Corticosteroid-induced ischemic femoral head necrosis | Often bilateral, is sometimes observed as late as 3 years after stopping corticosteroids |

[a]See Chap. 16 and Addendum

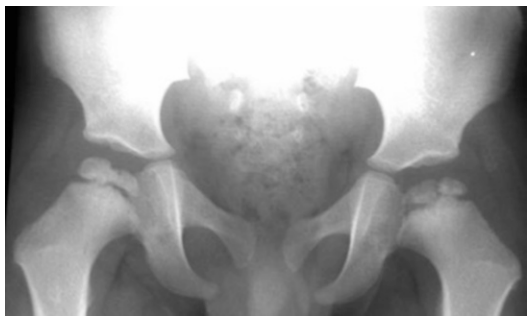


**Fig. 9.47** Meyer dysplasia of both hips

In 9 % of cases the skeletal age runs on average 21 months behind the biological age, so there may be a combination of factors. In any event, the result is necrosis of the femoral head. The disease has four phases (Fig. 9.48). The first phase is usually left out (Fig. 9.49).

I. **Synovitis phase:** This phase lasts several weeks or months. X-rays do not show any irregularities. Technetium scans show a reduced uptake and MRI scans show a reduced signal.
II. **Collapse phase** (in other classifications also known as initial phase): This phase lasts 6–12 months. The epiphysis collapses, as a result of which it loses height. Because of the collapse of the epiphysis there is tissue thickening, giving the epiphysis a more radiopaque appearance on X-rays.
III. **Fragmentation phase:** The fragmentation phase lasts 1–2 years. The necrotic bone is reabsorbed, which is visible on X-rays as a spotty deossification. Deformation of the femoral head may occur during this phase.
IV. **Reconstitution phase:** This phase can take several years and lasts longer the older the child is.

There are several classifications in use that represent the severity of the anomaly on X-rays.

**Salter and Thompson classification** The Salter and Thompson classification is based on the subchondral fracture also known as crescent or Salter sign. This subchondral fracture runs an

**Table 9.9** Differences between Legg-Calvé-Perthes disease and Meyer dysplasia. Meyer dysplasia is probably more frequent, but is often incorrectly diagnosed as Legg-Calvé-Perthes disease or causes no symptoms, in which case it is not identified radiologically and thus not recognized

| | Legg-Calvé-Perthes disease | Meyer dysplasia |
|---|---|---|
| Frequency | 1:10,000 | 1:100,000 |
| Age | 2–13 years (80 % between 4 and 8 years) | 2–4 years |
| Boys | 75–80 % | 90–95 % |
| Bilateral | 10–15 % | 55–60 % |
| Symptoms | Mostly painful | No or very little pain |
| | Movement limitation (internal rotation) | Normal movements from age 2 |
| Appearance of the femoral head epiphysis | 2–6 months (normal) | |
| Appearance of the femoral head epiphysis at the moment of appearance | One piece | Several parts |
| Development of the femoral head epiphysis | Phased trajectory: collapse phase 6–12 months, fragmentation phase 1–2 years, reconstitution phase several years | Progressive improvement in 2-year time span |
| Result | Minimal to severe deformation, 50 % has arthrosis at age 60 | Mostly no or minimal changes, no increased chance of arthrosis |

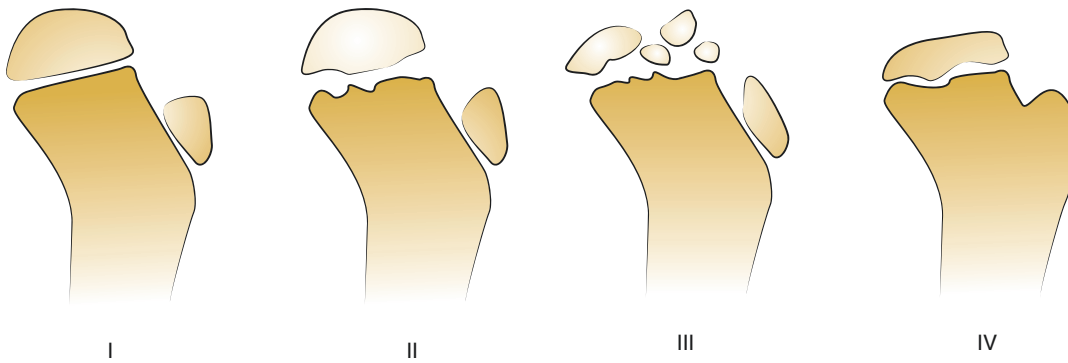



**Fig. 9.48** Legg-Calvé-Perthes disease undergoes four phases: *I* Synovitis. *II* Collapse phase. *III* Fragmentation phase. *IV* Reconstitution phase

anterior-to-posterior course. If the fracture comprises less than half of the width of the epiphysis it is a type A, if it comprises more than half it is type B (Fig. 9.50). A subchondral fracture appears about 3–4 months after the onset of the disease. One problem, however, is that it is visible in only 30 % of X-rays in children with Legg-Calvé-Perthes disease (Fig. 9.51).

**Cattarall classification** (Fig. 9.51). Another classification is that of Cattarall, in which the severity of the anomaly is subdivided into four groups.

- Group 1: Only the anterior part of the femoral head is affected. There is no collapse of the femoral head.
- Group 2: The anterior half of the hip is affected. A collapse of the femoral head is improbable but does occur. Anteroposterior X-rays of the pelvis usually shows a central necrotic part with normal areas on both sides (Fig. 9.52).
- Group 3: Only the posterior part of the hip is still intact. The epiphysis has mostly collapsed and protrudes past the lateral edge of the

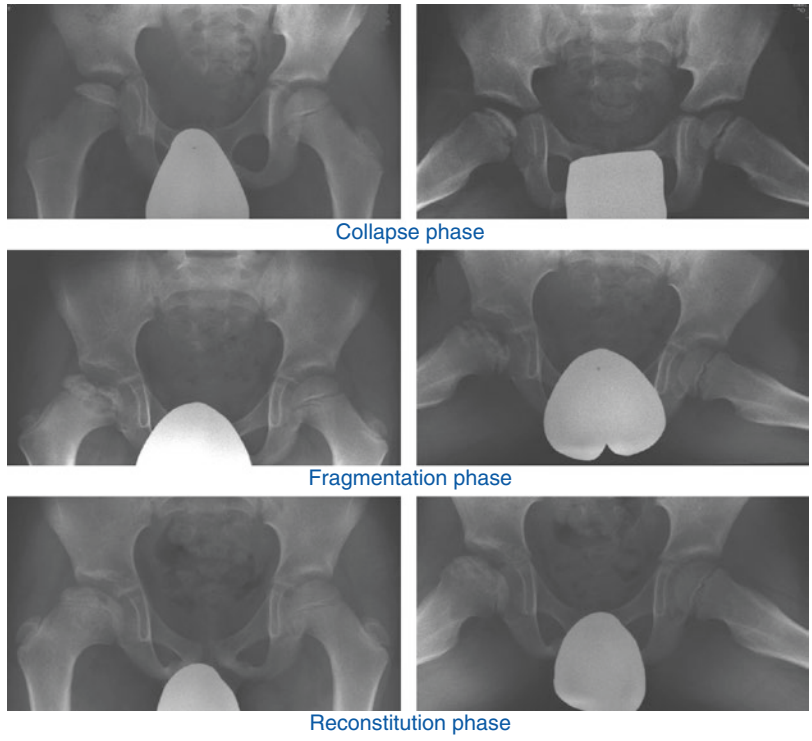



**Fig. 9.49** Legg-Calvé-Perthes disease of the right hip. The left side shows an anteroposterior image of the pelvis, the right side a frog-leg lateral view. In most classifications the first phase (synovitis phase) is left out

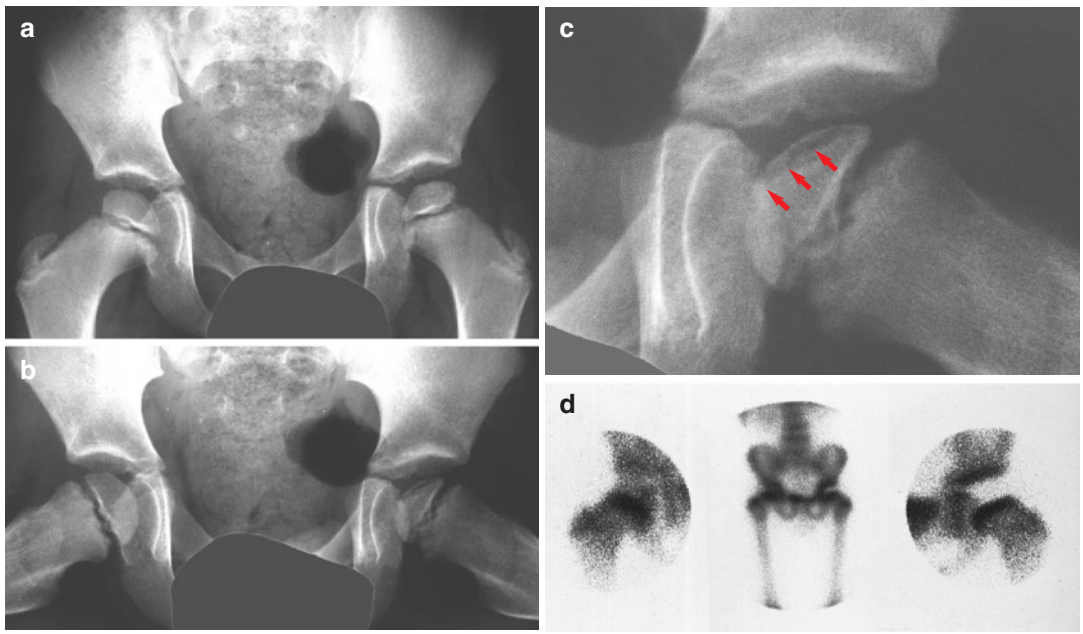



**Fig. 9.50** (**a**, **b**) Anteroposterior and frog-leg lateral view of the pelvis. There is a subchondral fracture of the left femoral head, which is only visible on the frog-leg lateral view (**b**). (**c**) Magnified frog-leg lateral view of the left hip. The subchondral fracture comprises more than half of the width of the epiphysis. This makes it a Salter and Thompson type B. (**d**) The bone scan shows reduced activity of part of the femoral head as a result of a disorder in vascularisation

acetabulum. This is called lateralization of the epiphysis. On X-rays a small normal area is still visible on the posteromedial side. There may be metaphyseal cysts.
- Group 4: The entire epiphysis is necrotic. There is a severe collapse of the femoral head with posterolateral protrusion, which gives the femoral head a mushroom shape. There are metaphyseal cysts.

Radiologically speaking, especially at an early stage in the disease, these four groups are difficult to distinguish from each other. Cattarall also introduced the concept of "head at risk signs", observable on anteroposterior X-rays of the pelvis. The presence of head-at-risk signs increases the chances of a poor result. These risk signs are: calcification and overgrowth of the lateral side of the epiphysis, metaphyseal cysts, a more horizontal trajectory of the growth plates with respect to the normal contralateral side, V-shaped radiolucency on the lateral aspect of the physis known as the Gage sign, and an increase in the distance between the acetabular teardrop figure and the medial part of the metaphysis (Fig. 9.53).

**Herring classification** (Fig. 9.51). A more practical and reproducible classification is that of Herring. This classification is based on anteroposterior radiological imaging of the hip. The epiphysis is divided into three pillars: the lateral pillar comprises 15–30 % of the width of the epiphysis, the central pillar about 50 % and the medial pillar 20–35 %. In group A the lateral pillar is normal, in group B there is a collapse of the lateral pillar of less than 50 %, and in group C there is a collapse of the lateral pillar of more than 50 %.

**Prognostic factors** Age at the beginning of the condition is also a prognostic factor. Children younger than 6 years tend to have a good prognosis, even if more than half of the femoral head is involved. The femoral head has more time to remodel during growth. However, there can be poor end results in this younger-than-6 age group too. The final result is seldom good in children in whom the disease begins at age 8 or later. Although the disease is four to five times more common among boys than girls, the severe forms in which more than half of the femoral head is involved are more common in girls. A severe form of Legg-Calvé-Perthes disease is only twice as common in boys than in girls.

If less than half of the femoral head is affected, the prognosis is better than when more than half is affected. The presence of head-at-risk signs on anteroposterior X-rays of the pelvis increases the risk of a poor result.

To summarize, we can say that in the following cases there is an increased risk of a less favorable result:

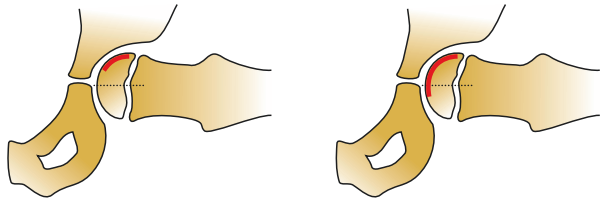
- The child is older than age 6.
- The femoral head is extensively affected (Salter type B, Cattarall groups 3 and 4, and Herring group C).
- Head-at-risk signs.

**Chronic slipped capital femoral epiphysis** A child with chronic slipped capital femoral epiphysis can still walk, but the gait is antalgic[5] (see Chap. 16 and addendum) and internal rotation is limited (Fig. 9.54). If there is acute or acute-on-chronic slippage, the child doesn't walk.

A slipped capital femoral epiphysis is a slippage of the femoral head at the level of the epiphyseal growth plate. The epiphysis slips with respect to the femoral neck, in general medially and posteriorly. Very seldom there is a lateral and anterior slippage. The anomaly can occur as part of several endocrine disorders (Table 9.10), hence suspecting a hormonal cause should be obvious. In a large number of cases there is obesity. Especially in boys with obesity, the genitals, especially the penis, are relatively too small and adiposogenital dystrophy (Fröhlich syndrome), which is extremely rare, may be mistakenly suspected (Fig. 9.55).

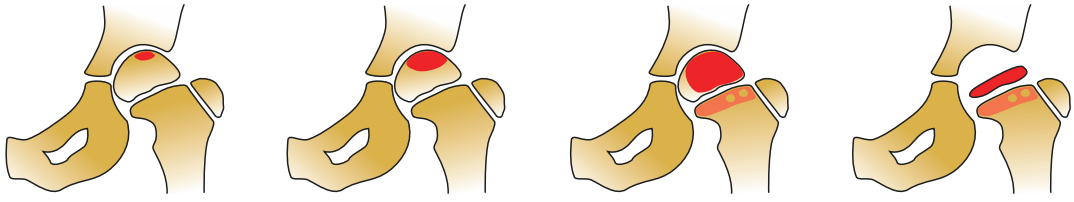
Epiphysiolysis of the femoral head affects 2 in a population of 100,000. In boys the condition is

[5] See Addendum.

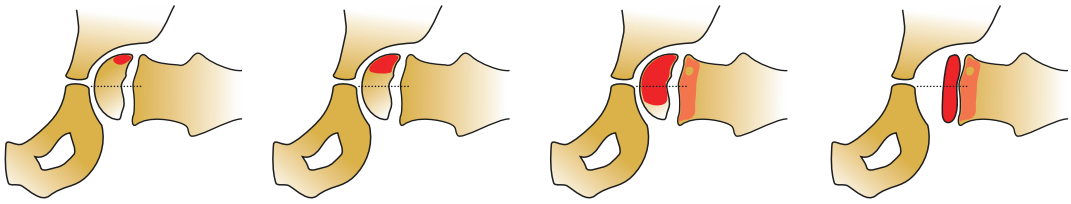


**A** < 50% **B** > 50%

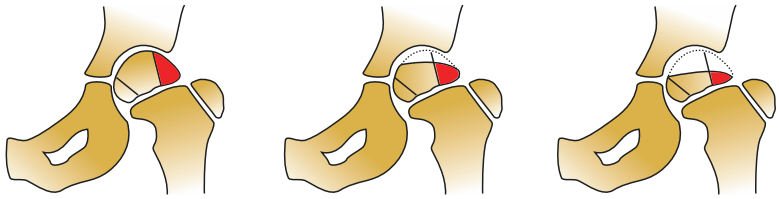
Salter-Thompson



**1** 0-25% **2** 25-50% **3** 50+% **4** 100%



Catterell



**A** lateral pillar 100% **B** lateral pillar < 50% **C** lateral pillar > 50%

Herring

**Fig. 9.51** Salter and Thompson classification: This classification is made on a frog-leg lateral X-ray of the hip joint. *Type A*: The subchondral fracture comprises less than half of the width of the epiphysis. *Type B*: The subchondral fracture comprises more than half of the width of the epiphysis. Cattarall Classification: This classification is made on an anteroposterior as well as an frog-leg lateral X-ray of the hip joint. *Group 1*: Only the anterior side of the femoral head is involved (25 %). *Group 2*: The anterior part of the femoral head is affected more than 25 % but less than 50 %. *Group 3*: More than 50 % of the femoral head is involved, only the posterior part of the femoral head is still intact. In the proximal part of the metaphysis there may be cysts. *Group 4*: The entire epiphysis is necrotic. There is a severe collapse of the femoral head with lateral and posterior protrusion, giving the hip the shape of a mushroom. The metaphysis has cysts. Herring classification: This classification is done based on an anteroposterior X-ray of the hip. In *group A* the lateral pillar is normal, in *group B* there is a collapse of less than 50 % of the lateral pillar, in *group C* there is a collapse of more than 50 % of the lateral pillar (Redrawn from: Staheli RT. Fundamentals of pediatric orthopaedics. 4th ed. Philadelphia/Baltimore/New York/London/Buenos Aires/Hongkong/Sydney/Tokyo: Lippincot Williams & Wilkins. 2008)

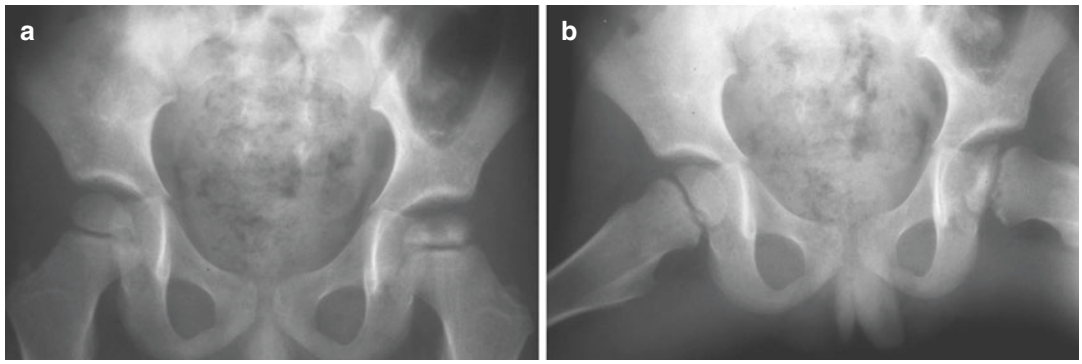



**Fig. 9.52** (**a**) Anteroposterior and (**b**) frog-leg lateral X-ray of the pelvis. According to the Cattarall classification there is group 2 Legg-Calvé-Perthes disease of the left hip. The anteroposterior pelvic image shows a central necrotic part with normal areas on both sides

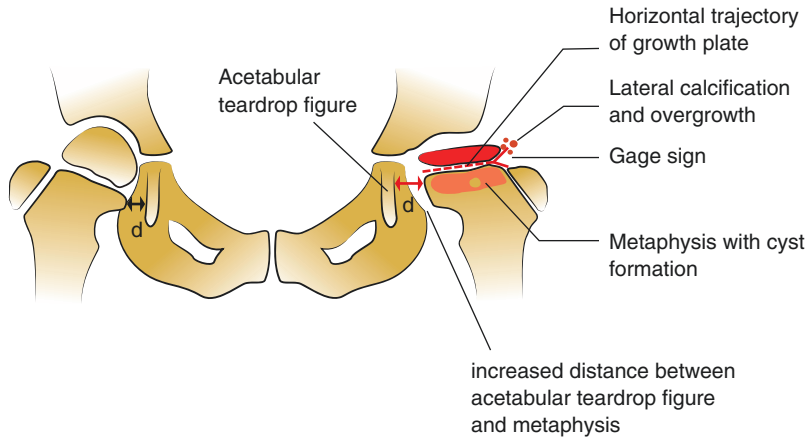



**Fig. 9.53** Graphic depiction of an anteroposterior X-ray of the pelvis with head-at-risk signs in Legg-Calvé-Perthes disease. These risk signs are: calcification and overgrowth on the lateral side of the epiphysis, metaphyseal cyst(s), a more horizontal trajectory of the growth disc, a V-shaped radiolucency in the lateral part of the epiphysis and adjoining metaphyseal portion (*Gage sign*), and an increase in the distance between the acetabular teardrop figure and the medial part of the metaphysis (Redrawn from: Schulitz KP, Dustmann HO. Morbus Perthes. Ätiopathogenese. Differentialdiagnose, Therapie und Prognose. Berlin/Heidelberg/New York/London/Paris/Tokyo/Hong Kong/Barcelona/Budapest: Springer Verlag; 1991)

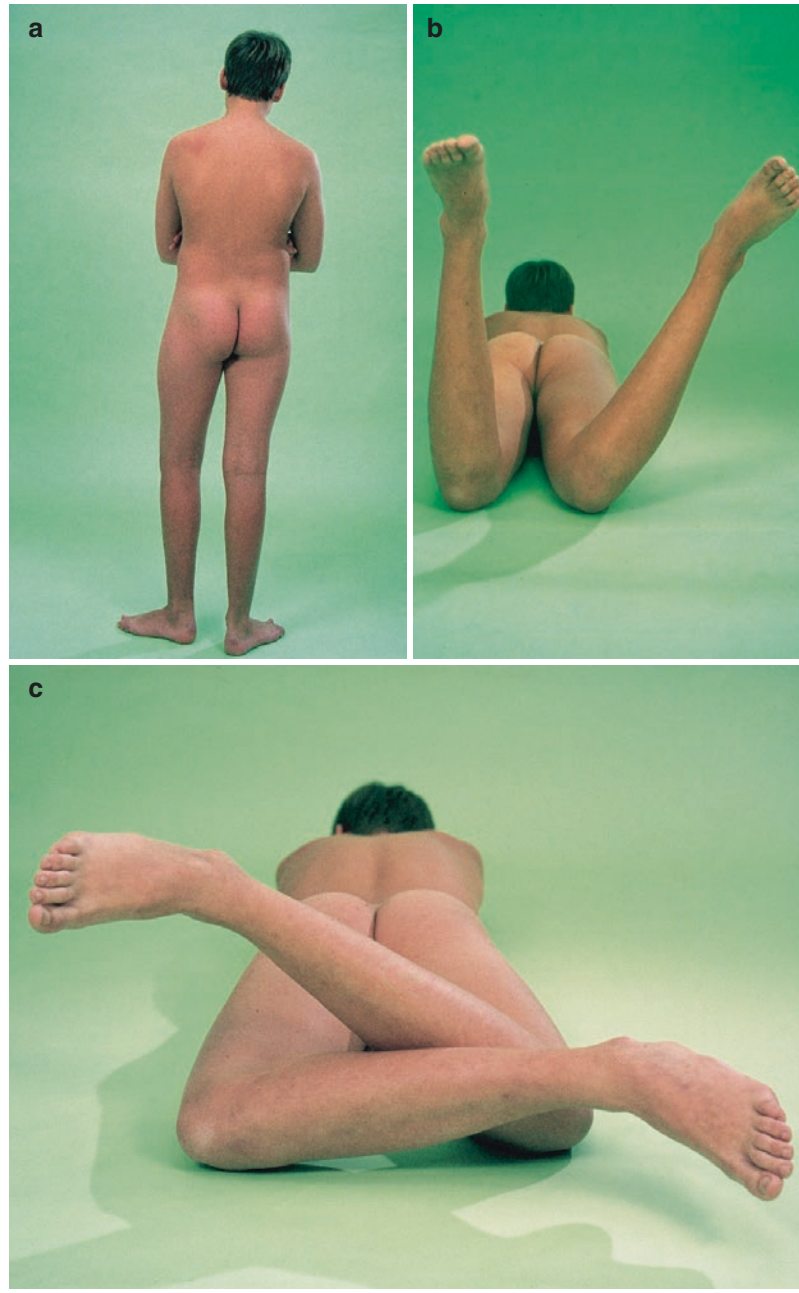



**Fig. 9.54** Chronic slipped capital femoral epiphysis of the left hip. The patient can still walk, but there is an antalgic limp (see addendum). (**a**) The left leg is in external rotation. (**b**) Internal rotation of the left hip is limited. (**c**) External rotation of the left hip has increased

twice as common as in girls, affecting the left hip three times more often than the right hip. The anomaly appears from the sixth year of life until the end of growth, but in girls mostly between the ages of 11 and 13 at an average of 12 years, and in boys mostly between the ages of 13 and 15 at an average of 14. In girls it rarely develops after menstruation starts. Especially if the epiphysiolysis occurs before the age of 10, an endocrine disorder should be considered. In 20–25 % of cases the anomaly occurs bilaterally. In half of the bilateral cases the slippage of the femoral head appears more or less simultaneously. In 80 % the second hip slips within 18 months, in the other half of bilateral cases. Young patients and patients with endocrine disorders have a much greater risk of bilateral slippage. Instead of chronic slippage there can be acute or acute-on-chronic slippage.

**Classification** A classification can be made based on the duration of symptoms, weight-bearing ability, radiologic and ultrasound assessment.

**Duration of symptoms** In acute slippage there is a sudden onset of symptoms that have lasted less than 3 weeks. In chronic slippage the symptoms last longer than 3 weeks and develop gradually. In an acute-on-chronic slippage there is an acute increase of already existing, gradually developing symptoms that have lasted longer than 3 weeks. The most common type is chronic slippage (85 % of cases). These patients have few symptoms, and all walk with an antalgic gait[4]. In 5 % of cases there is acute slippage and in 10 % there is acute-on-chronic slippage. Mostly a fall or stumble precedes this acute shifting, and the hip joint is very painful, as a result of which the child can no longer walk or stand on the affected leg. This classification strongly depends on the recollections of the child and the parents.

**Table 9.10** Endocrine disorders in which a slipped capital femoral epiphysis occurs relatively frequently

| Endocrine disorder | Malfunction |
|---|---|
| Craniopharyngioma | Hypophysis (brain tumor) |
| Gigantism | Hypophysis (hypophysis tumor) |
| Hypopituitarism | Hypophysis |
| Congenital hypothyroidism or cretinism | Thyroid gland |
| Acquired hypothyroidism or juvenile mixoedema | Thyroid gland |
| Hypoparathyroidism | Parathyroid gland |
| Klinefelter syndrome* | Testicles |
| Adiposogenital dystrophy | Testicles (hypothalamus tumor) |

*See Appendix

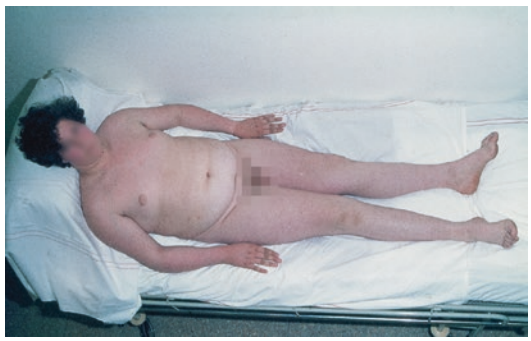


**Fig. 9.55** There is obesity in many cases of a slipped capital femoral epiphysis. Particularly in boys with obesity the genitals (especially the penis) are relatively too small, which often leads to a mistaken diagnosis of the extremely rare adiposogenital dystrophy. This patient had an acute slipped capital femoral epiphysis of the right hip

**Weight-bearing ability** A subclassification into stable and unstable slippage is a more practical one. With unstable slippage the patient has so much pain that it is impossible to stand on the affected leg. Walking, even with crutches, is impossible. With a stable slippage the patient can stand on the affected leg, and walking with or without crutches is possible. An acute and acute-on-chronic slippage are unstable, a chronic is stable.

**Radiological and ultrasound assessment** The classic classification into acute, chronic and acute-on-chronic is also possible using X-ray images and ultrasound. On this basis, an acute slip is defined as a slip where ultrasound assessment shows a hydrops in the joint and X-rays show no remodelling. A chronic slip is defined as a slip without hydrops on the ultrasound but with signs of remodelling or subperiostal new bone formation around the epiphysis-metaphysis junction on the X-rays. Acute-on-chronic slippage shows hydrops in the joint on the ultrasound as well as signs of remodelling or subperiostal new bone formation around the epiphysis-metaphysis junction on the X-rays (Table 9.11).

**Table 9.11** Classification into acute, chronic and acute-on-chronic slipped capital femoral epiphysis

| Classification of slipped capital femoral epiphysis | Symptoms duration | Weightbearing | X-rays | Ultrasound |
|---|---|---|---|---|
| Acute slip | <3 weeks | No weight-bearing (unstable) | No remodelling | Hydrops |
| Chronic slip | >3 weeks | Walks without crutches (stable) | Remodelling | No hydrops |
| Acuteonchronic slip | >3 weeks | No weight-bearing (unstable) | Remodelling | Hydrops |

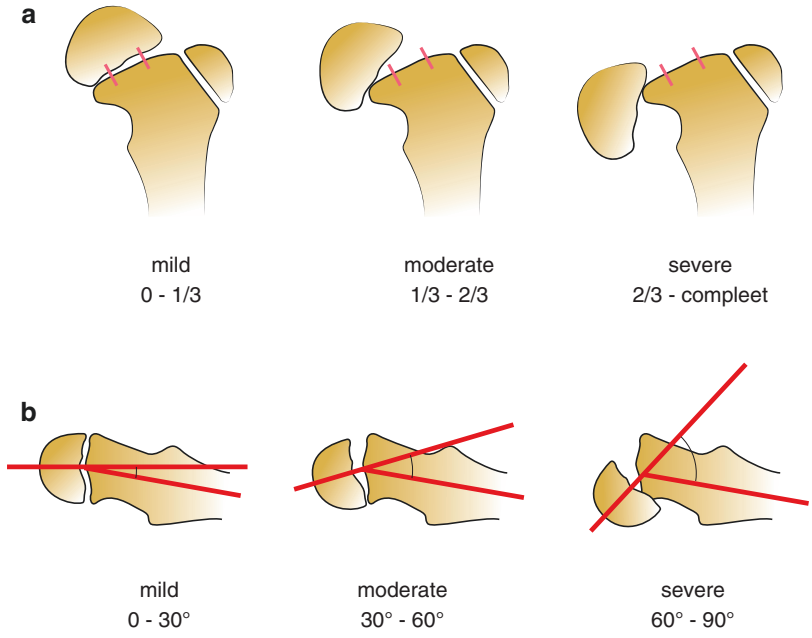



**Fig. 9.56** The degree of severity of the slippage in a slipped capital femoral epiphysis can be established radiologically. (**a**) If an anteroposterior X-ray shows that the epiphysis has slipped less than one-third with respect to the metaphysis one speaks of a mild slip, one- to two-thirds is a moderate slip, and if the epiphysis has glided more than two-thirds there is a severe slip. (**b**) The angle between metaphysis and epiphysis is determined on a frog-leg lateral X-ray. In mild slippage the angle is less than 30°, moderate slippage is identified with an angle between 30° and 60°, and if the angle exceeds 60° the slippage is considered severe

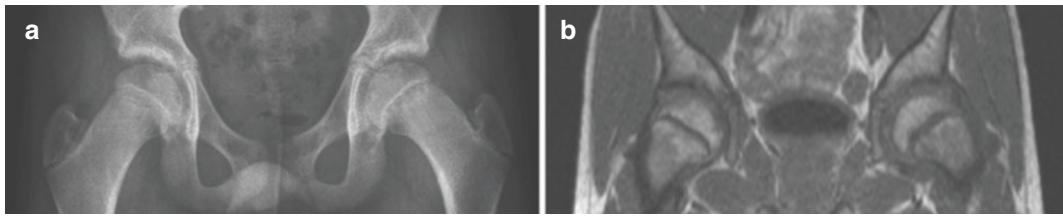



**Fig. 9.57** In the early phase of a slipped femoral head epiphysis there is a widening of the growth plate, without slippage ("preslip"). (**a**) On the anteroposterior X-ray of the pelvis a widening is shown of the growth plate of the left hip. (**b**) Visibility is better on an MRI image

**Degree of slippage** The degree of slippage can be subclassified into minimal, moderate and severe as seen on an anteroposterior or a frog-leg lateral X-ray of the pelvis (Fig. 9.56). In the early phase of chronic slippage there may be only a widening of the growth plate without a slip. This is known as "pre-slip" (Fig. 9.57).

Supplementary assessment: Anteroposterior and frog-leg lateral X-ray of the pelvis. In an acute transient synovitis of the hip no irregularities are observable on conventional X-rays.

To determine the diagnosis of acute transient synovitis, a diagnosis per exclusionem, an ultrasound of the hip is not necessary. If there is hydrops in the hip joint the internal rotation of the hip joint is limited which can be confirmed with an ultrasound, but not more than that either. One should realize however that the condition may not be an acute transient synovitis of the hip but the synovitis stage of Legg-Calvé-Perthes disease.

If the symptoms and movement limitation of the hip persist after a week, it would be

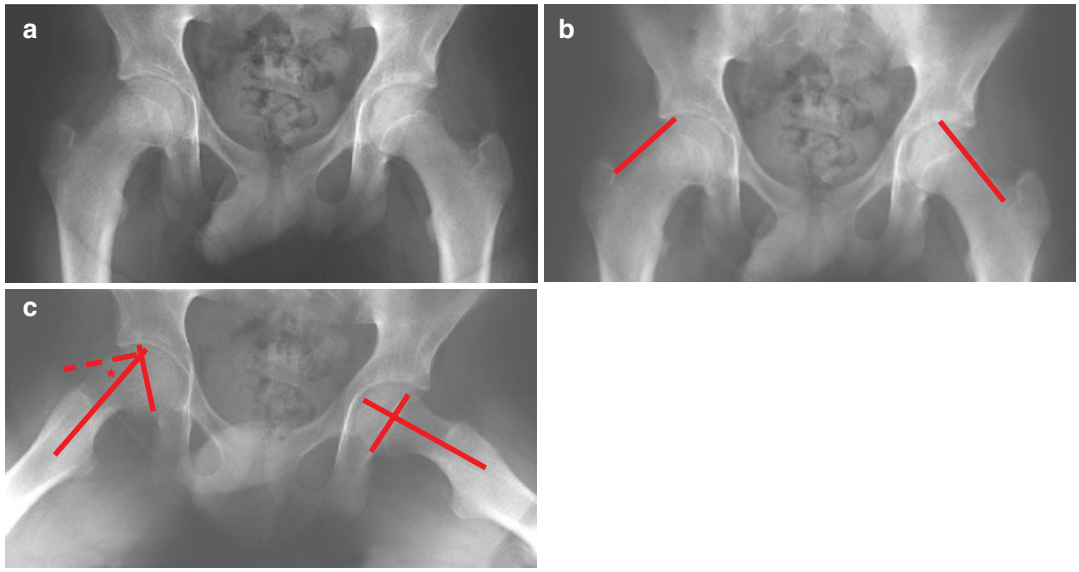



**Fig. 9.58** (**a**) Anteroposterior X-ray of the pelvis with a rightside slipped capital femoral epiphysis. This is difficult to recognize on an anteroposterior image when the slippage is mild. (**b**) To detect slippage on an anteroposterior X-ray of the pelvis, the Klein line can be drawn along the upper edge of the femoral neck. If this line does not go through the femoral head (*Trethowan sign*), as is the case in the right hip, then a slipped capital femoral epiphysis is present. On the left side the Klein line does go through the femoral head so there is no slippage. (**c**) Minimal slip of the epiphysis can be seen better on a frog-leg lateral view of the pelvis than on an anteroposterior image.* Degree of slippage of the right-hip epiphysis

wise to perform additional radiological assessment of the hip again after 2 months, to see if there is a case of Legg-Calvé-Perthes disease. Just like a slipped capital femoral epiphysis, Legg-Calvé-Perthes disease is sometimes missed on anteroposterior X-rays of the pelvis. A mild form of slipped capital femoral epiphysis and a mild form of Legg-Calvé-Perthes disease can be more easily seen on a pelvic frog-leg lateral X-ray than on anteroposterior X-rays of the pelvis (Fig. 9.58). To track a slipped capital femoral epiphysis on an anteroposterior X-ray of the pelvis, the Klein line along the upper lateral sides of the femoral neck can be helpful. If this line doesn't go through the femoral head, the so-called Trethowan sign, then there is a slipped capital femoral epiphysis (Fig. 9.58).

Primary care treatment: One can prescribe 3–4 days of bed rest in children older than 2 years and younger than 13 without fever and with a painful internal rotation limitation of the hip joint. If the pain disappears after 3–7 days, the internal rotation limitation has disappeared and internal rotation in the end trajectory is no longer painful, then it was a coxitis fugax and further treatment is not necessary. The chances are great that one is dealing with synovitis stage of Legg-Calvé-Perthes disease if after a week the internal rotation is still painful and limited.

When to refer: If there is Legg-Calvé-Perthes disease or a chronic slipped capital femoral epiphysis.

Secondary care treatment: **acute transient synovitis of the hip.** None.

**Legg-Calvé-Perthes disease** The treatment of Legg-Calvé-Perthes disease is very controversial. In the past, treatment has varied from operation for every case to no treatment at all. According to Herring et al. (2004), in Legg-Calvé-Perthes disease there is no difference in results between non-treatment and treatment with an abduction orthesis (Fig. 9.59) or physiotherapy, and no difference in terms of the final

**Fig. 9.59** The long-term benefits of casting and bracing, e.g., with a Scottish Rite, have been called into question

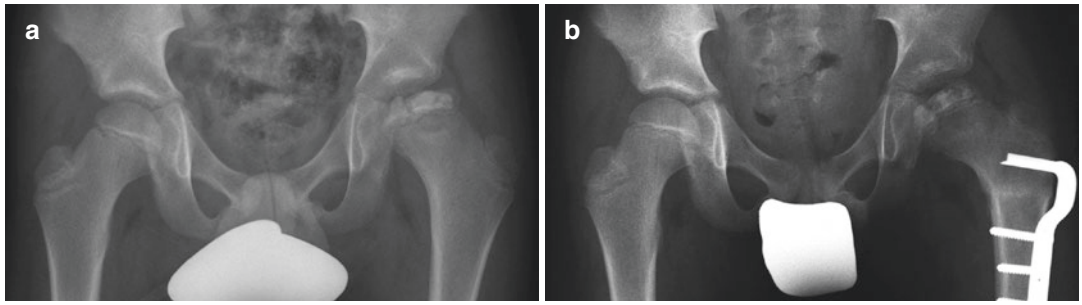



**Fig. 9.60** (**a**) Legg-Calvé-Perthes disease of the left hip, Herring group B. (**b**) Proximal femoral varus external rotation osteotomy. Children are eligible for operative treatment of Legg-Calvé-Perthes disease only if they got the disease at age 8 or older and if they can be classified according to Herring group B, or if there is doubt as to whether they can be classified under Herring group B or C

result between operative and nonoperative treatment in children younger than age 8. As regards the final result there is no difference between nonoperative and operative treatment in children older than 8 in Herring groups A and C. In group C, girls aged 8 or older at the start of the disease do considerably worse than boys. Children eligible for operative treatment are only those in whom the disease started at age 8 or later and who can be classified in Herring group B, and children in which there is doubt as to whether they should be classified under Herring group B or C. In such cases a choice is made for treatment aimed at enclosing the femoral head entirely in the hip socket known as containment treatment. This can be accomplished with a proximal femoral varus external rotation osteotomy (Fig. 9.60) or a Salter pelvic osteotomy. There is no difference in the final result between these two types of operative treatment. A shelf arthroplasty can also be performed to prevent lateral overgrowth of the capital epiphysis.

In the fragmentation phase the acetabular rim may make an indentation in the soft femoral head, if the disease has spread over more than half of the femoral head. This causes what is known as a hinge abduction, in which the femoral head does not turn into the socket when in abduction, but hinges around the indentation (Fig. 9.61). A proximal

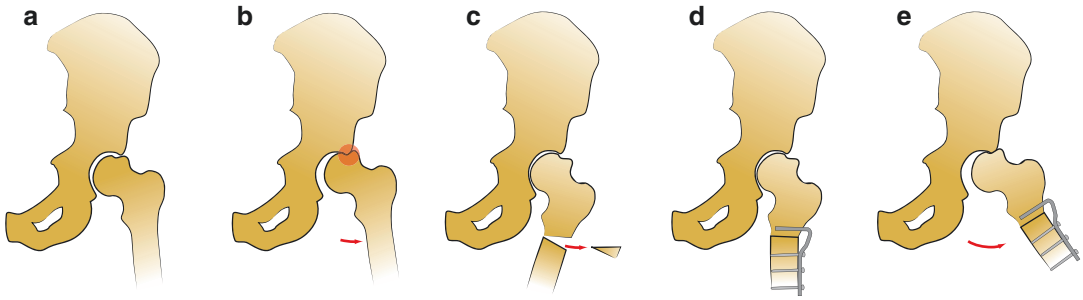



**Fig. 9.61** (**a**) If in Legg-Calvé-Perthes disease more than half of the femoral head is involved, the acetabular rim can indent the soft femoral head, forming a knob on the lateral side of the femoral head because the acetabulum does not cover the femoral head completely. (**b**) There is a hinge abduction, which does not allow the femoral head to turn into the acetabulum when in abduction but instead hinges around the indentation. (**c**, **d**) In case of a hinge abduction a proximal femoral valgus osteotomy can be considered. (**b**) This increases the abduction possibility in the hip joint

**Table 9.12** Stulberg classification for final result of Legg-Calvé-Perthes disease

| |
|---|
| Type I: there is a normal femoral head and acetabulum |
| Type II: the femoral head is spherical within 2 mm of a circle on both an anteroposterior and a frog-leg lateral X-ray. Normal acetabulum and congruent joint |
| Type III: there is an aspheric elliptic-shaped femoral head with a normal acetabulum and a congruent joint |
| Type IV: there is an aspheric elliptic-shaped femoral head with a flattened acetabulum and a congruent joint |
| Type V: there is an aspheric femoral head with a normal acetabulum and an incongruent joint |

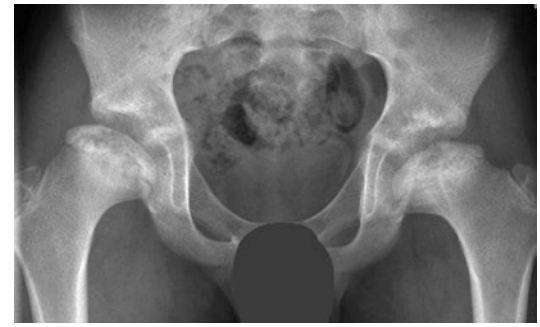


**Fig. 9.62** Anteroposterior X-ray of the pelvis after Legg-Calvé-Perthes disease. Both sides Stulberg III

femoral valgus osteotomy can be considered for a hinge abduction. This increases the abduction possibility in the hip joint (Fig. 9.61).

Up to the age of 8 it can be useful to perform an epiphysiodesis of the greater trochanter using a screw in the case of an elevated position of the greater trochanter in order to prevent a Trendelenburg gait.

To determine the final result several classifications are available. The most common is that of Stulberg (Table 9.12). Stulberg types I and II have a good prognosis, types III and IV have chances of mild-to moderate degenerative changes. In hips that fall under type V the degenerative changes develop at a young age, accompanied by pain (Figs. 9.62 and 9.63).

**Chronic slipped capital femoral epiphysis** When treating a slipped femoral head epiphysis, the motto—don't make it even worse—applies.

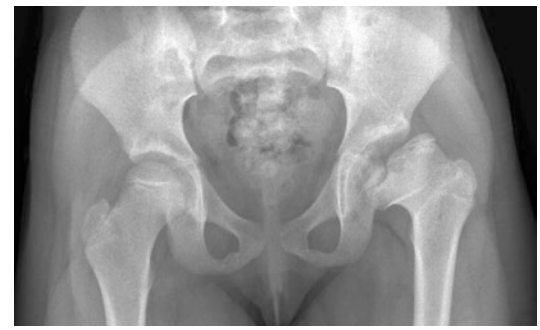


**Fig. 9.63** Anteroposterior X-ray of the pelvis after Legg-Calvé-Perthes disease of left hip. Stulberg V. There is also an osteochondritis dissecans of the affected left femoral head

Before the treatment is started, one should be aware of the following priorities:

- avoid ischemic femoral head necrosis
- avoid chondrolysis

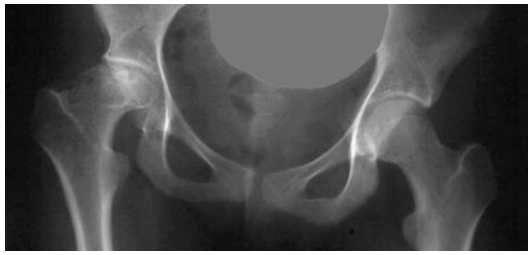


**Fig. 9.64** Ischemic femoral head necrosis of the right femoral head after a slipped capital femoral epiphysis

**Table 9.13** Risk factors for ischemic femoral head necrosis

| |
|---|
| Acute (unstable) slip |
| Severe slip |
| Forced reduction of an acute slip |
| Attempting to reduce a chronic slip |
| Femoral neck osteotomy |

**Table 9.14** Risk factors for a chondrolysis

| |
|---|
| Severe slip |
| Slippage has been present for a long time without treatment |
| Cast immobilization |
| Penetration of a screw or metal pins into the joint |

- prevent further slippage
- finally, correct the deformity where possible

Ischemic femoral head necrosis is a severe complication resulting from a circulatory disorder (Fig. 9.64). It is accompanied by severe pain, stiffness and early degenerative changes in the joint (Table 9.13).

An equally severe complication is chondrolysis, in which the cartilage disappears. These patients also have severe pain, stiffness and contractures of the hip (Table 9.14). There isn't a debate anymore about which type of primary treatment is necessary for chronic slippage: the epiphysis is fixed with a centrally placed screw (Fig. 9.65). If the chronic slip is not fixed there is a 10 % risk of further slippage. An advanced chronic slip must never be reduced, as this increases the risk of ischemic femoral head necrosis.

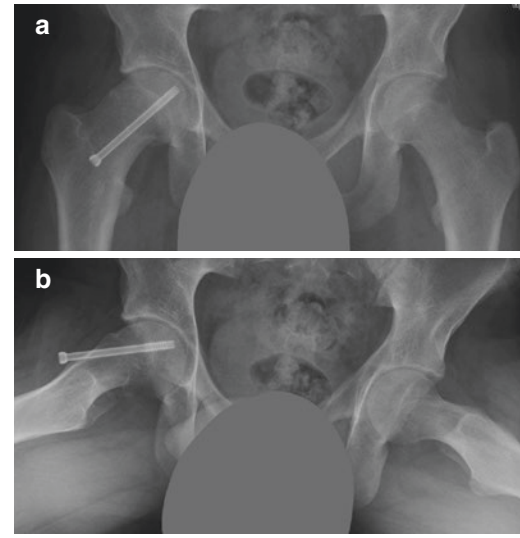



**Fig. 9.65** Fixation of a chronic slip of the right hip using a screw. The screw should be centrally located on the anteroposterior (**a**) as well as on the frog-leg lateral X-ray (**b**). The screw has to be inserted into the anterior side of the femoral neck because the epiphysis has glided posteriorly

The risk of ischemic head necrosis and chondrolysis are both 4 %, for untreated chronic slippage. After fixation of chronic slippage using a screw, there is a 5 % chance of ischemic femoral head necrosis and a 5 % chance of chondrolysis. The latter especially after pin or screw penetration into the hip joint at the operation. Only in 2–5 % of cases is a secondary proximal femoral osteotomy necessary after primary treatment of the epiphysiolysis due to contractures of the hip. A femoral neck osteotomy has the most chance of producing an ischemic head necrosis and a lateral femoral neck or intertrochanteric or subtrochanteric osteotomy the least (Fig. 9.66).

Even in those cases where there is no chondrolysis and/or ischemic femoral head necrosis, degenerative changes in the hip joint occur in 20–30 % of cases after 30 years.

### Femoroacetabular Impingement

Femoroacetabular impingement may also develop. In this case there is usually pain in the groin area especially with turning, twisting and squatting. There are three types: pincer, cam and combined impingement.

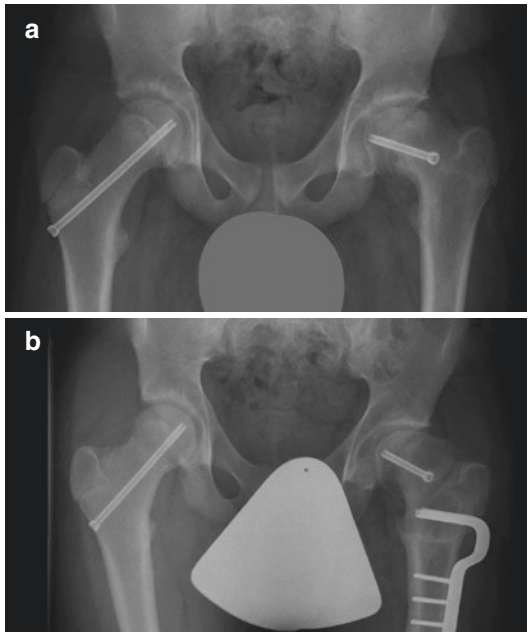



**Fig. 9.66** (**a**) Situation after reduction and fixation of a rightsided acute slip and left-sided chronic slip. (**b**) An intertrochanteric valgus and flexion osteotomy of the left femur was performed to relieve painful flexion and abduction of the left hip caused by a protruding anterior edge of the metaphysis

**Pincer** This type of impingement is a result of direct contact between the acetabular ring and the femoral head-neck junction, because extra bone extends out over the normal rim of the acetabulum. The labrum can be crushed under the prominent rim of the acetabulum.

**Cam** In cam impingement (abbreviation for camshaft which the shape of the femoral head and neck resembles) the femoral head is not round and cannot rotate smoothly inside the acetabulum. A protuberance forms on the border of the femoral head that grinds the cartilage inside the acetabulum.

**Combined** Combined impingement just means that both the pincer and the cam types are present. Patients with coxa magna in Legg-Calvé-Perthes disease and in a slipped capital femoral epiphysis are especially prone to development of a cam or combined type.

If non-steroidal anti-inflammatory medications do not help arthroscopic trimming of the bony rim of the acetabulum and the protuberance on the femoral head may be an option.

## Hip Pain and Movement Limitation in All Directions

Complaint: An adolescent complains about movement limitation in the hip.

Assessment: Mobility of the hip joint is limited in all directions.

Differential diagnosis:

**acetabular protrusion**
**(Otto pelvis, arthrokatadysis)**
**chondrolysis**
acquired type
idiopathic type

Explanatory note: **acetabular protrusion.** In this condition there is an abnormally deep acetabulum with the femoral head abnormally deep in the pelvis. The medial boundary of the acetabulum is paper-thin. The anomaly is also known as Otto pelvis, after Otto (1824), the Pole who discovered this anomaly. This is a rare anomaly that sometimes runs in families and is relatively often seen in Marfan syndrome[6]. The incidence is 1 in 10,000, affecting girls much more often than boys. In early adolescence a movement limitation of the hip joint develops in all directions, without pain in the foreground. At a later age degenerative changes develop that are accompanied by pain.

**Chondrolysis** A chondrolysis is characterized by a progressive destruction of the cartilage, causing a narrowing of the joint space accompanied by a movement limitation of the hip in all directions. There is an acquired and idiopathic type.

*Acquired type* This condition is seen most frequently after a slipped capital femoral epiphysis, but also occurs after infection, trauma or prolonged spica cast immobilization.

[6]See appendix

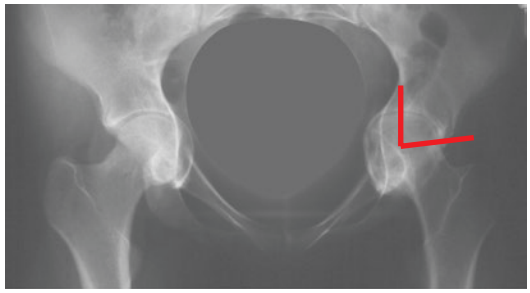


**Fig. 9.67** Acetabular protrusion (Otto pelvis or Otto-Chrobach disease). The CE angle on both sides exceeds 60°

*Idiopathic type* In this type there is probably an autoimmune reaction at play, that remains limited to the hip joint. This condition is rare. It affects girls between ages 9 and 18, with an average age of 12, and boys between ages 13 and 20, with an average age of 15. The anomaly is bilateral in less than 10 % of cases. There is pain in the anteromedial side of the hip joint. The pain is accompanied by a progressive movement limitation of the hip joint in all directions, and there is an antalgic gait. There may also be contractures, the most common being flexion, abduction and external rotation. The girls:boys ratio is 5:1. A chondrolysis presents with an acute phase of 6–12 months, followed by a chronic phase that can persist for 3–5 years. In the chronic phase there are two possibilities: in 50 % of cases there is a reduction of pain and an improvement in mobility. Radiologically there is an improvement of the cartilage interval, as the joint gap widens. This is known as the benign form. In the recalcitrant form there is a progressive loss of the cartilage interval, which can lead to a very painful hip or a painless ankylosis of the hip joint.

Supplementary assessment: anteroposterior X-rays of the pelvis. One speaks of an acetabular protrusion when the CE angle is larger than 60° on an anteroposterior X-ray of the hip. If the CE angle is between 40 and 60° one speaks of a deep acetabulum. CE is an abbreviation of C for "center" and E for "lateral edge of the acetabular roof" (Fig. 9.67). In a normal hip joint the cartilage interval is 4 mm or greater. There is a chondrolysis when the cartilage interval is 3 mm or less and in the course of time is accompanied by local osteoporosis and sclerosis of the borders of the hip joint (Fig. 9.68).

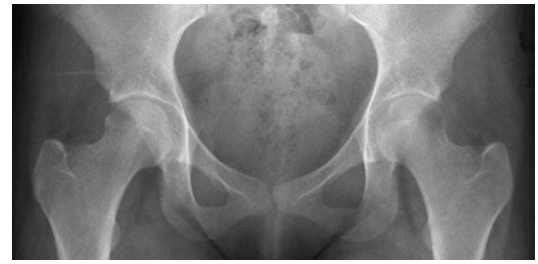


**Fig. 9.68** Idiopathic chondrolysis of the left hip

Primary care treatment: hip-loading activities and sports like volleyball, basketball and soccer are discouraged.

When to refer: as soon as the diagnosis of acetabular protrusion or chondrolysis are established.

Secondary care treatment: **acetabular protrusion.** An epiphysiodesis of the triradiate or y-cartilage on the inner side of the pelvis should be considered to prevent progression of the anomaly, between the ages of 8 and 10. The y-cartilage is a common growth plate of the iliac, ischial and pubic bones. For the rest one should wait until the pain due to degenerative changes in the joint increases to such a degree that the patient becomes eligible for a total hip joint replacement arthroplasty.

**Chondrolysis** Analgesics, partially loading the hip, temporary traction and/or bed rest to get through the acute phase. After about 3 years it can be established whether it is a benign or recalcitrant form. The benign type necessitates no further treatment. It may be possible to treat contractures of a painless ankylosis for the recalcitrant type. If the recalcitrant type does not lead to a painless ankylosis, a capsulotomy can be considered to reduce the pain, but this is not a common operation. A hip joint replacement arthroplasty is usually performed if no other options are available.

## Hip Pain After a Trivial Trauma

Complaint: after a fall or stumble the child cannot walk or stand on the affected leg.

Assessment: all movements are painful and extremely limited.

Differential diagnosis:

**fractured unicameral bone cyst (juvenile-, solitary-, simple bone cyst)**
**acute and acute-on-chronic slipped capital femoral epiphysis**

Explanatory note: **fractured unicameral bone cyst.** A unicameral bone cyst is a cavity filled with fluid that is covered with a thin, fibrous membrane. The initial location of the cyst is metaphyseal, but due to growth it ends up lying in the diaphysis starting in the twelfth year of life. In 90 % of cases the anomaly is found in patients younger than age 20 (70 % are boys). In 60 % of cases the cyst is localized in the proximal part of the humerus, and in 30 % of cases in the proximal part of the femur. In more than 80 % of cases a spontaneous fracture is the first symptom of the anomaly.

**Acute and acute-on-chronic slipped capital femoral epiphysis** The child is no longer able to walk or stand on the affected leg in contrast to the case of chronic slippage. For further explanation, see also pp. 156–158, 161–163.

Supplementary assessment: an anteroposterior and lateral X-ray of the hip joint for a fractured solitary bone cyst is sufficient (Fig. 9.69). A slipped femoral head epiphysis can often be seen clearly on a frog-leg lateral X-ray of the pelvis. If the epiphysis has slipped far, then flexion and abduction of the hip will be very limited and taking a frog-leg lateral X-ray of the hip is impossible. In that case a CT-scan is the best method to reliably determine the degree of slippage (Fig. 9.70).

Primary care treatment: none.

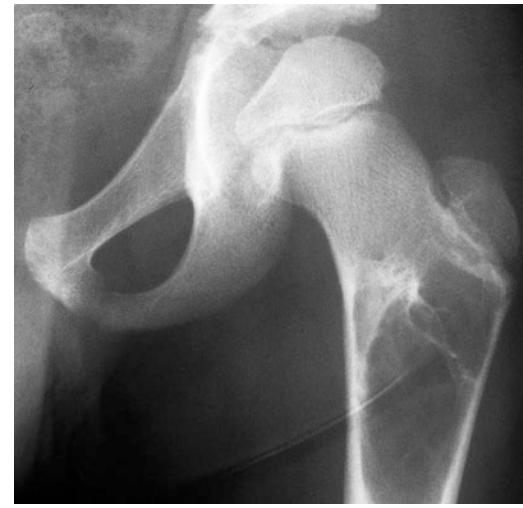


**Fig. 9.69** Solitary bone cyst in the proximal part of the left femur

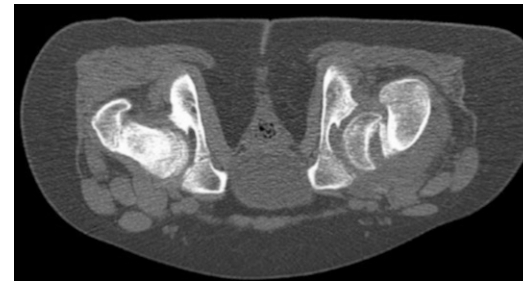


**Fig. 9.70** If in a slipped capital femoral epiphysis the epiphysis has slipped far, then flexion and abduction of the involved hip are extremely limited and it is not possible to take a lateral X-ray of the hip to get a reliable impression of the degree of slippage; a CT-scan would be the best method. The capital femoral head epiphysis of the left hip shows severe slippage

When to refer: in the case of an unicameral bone cyst, and in case of an acute or an acute-on-chronic slipped capital femoral epiphysis.

Secondary care treatment: **fractured unicameral bone cyst.** One can opt for immobilization in a spica cast if the fracture is not dislocated. After fracture healing in 25 % of cases the bone cyst disappeares. If the bone cyst has not disappeared after fracture healing, or the cyst is found by chance, it can be injected with corticosteroids. This is

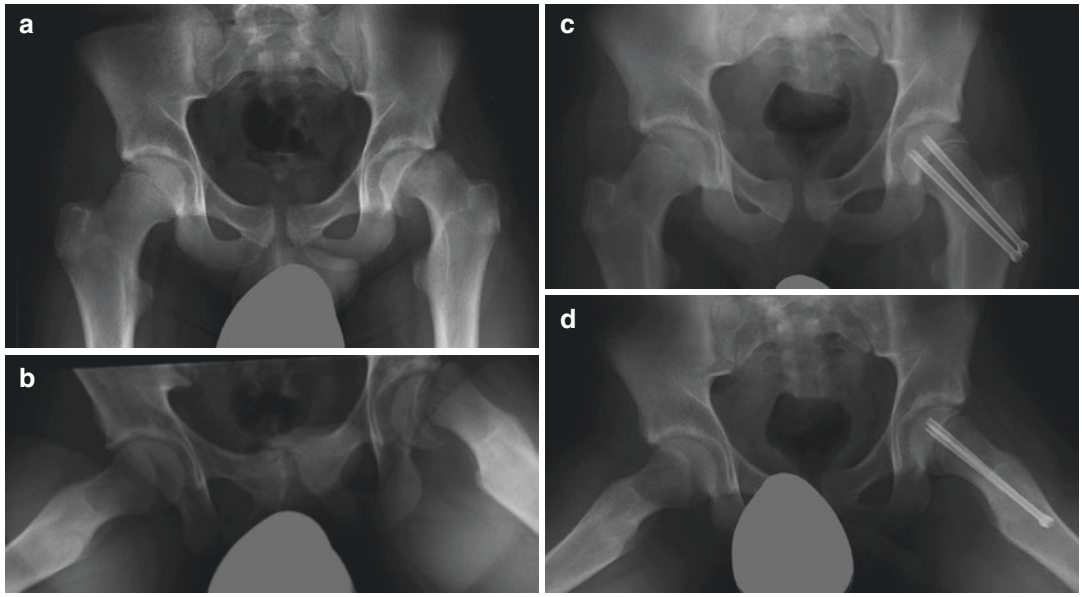



**Fig. 9.71** (**a**) Anteroposterior pelvic X-ray, and (**b**) frog-leg lateral X-ray of an acute slip of the left hip. (**c**) Anteroposterior pelvic X-ray, and (**d**) frog-leg lateral X-ray of the pelvis after reduction and fixation of an acute slip using two screws

followed by full recovery in 40 % of cases, partial recovery in 50 %, and no recovery in 10 %. The local injection with corticosteroids may be repeated three or four times as needed, with periods of several months between. Irresponsive cases can be treated with curettage and grafting with autogeneous, bone bank or artificial bone. After curettage and bone grafting there is full recovery in 50 % of bone cysts, incomplete recovery in 20 % and recurrence in 30 %. After a fracture of a cyst an alternative, certainly in cases of dislocation is operative stabilization of the fracture with flexible intramedullary rods or an angled plate with screws, after curettage and filling the bone cyst using autologous bone, homologous bone from the bone bank or artificial bone.

**Acute and acute-on-chronic slipped capital femoral epiphysis** When the slip is unstable, such as is the case in an acute slip or acute-on-chronic slip, in 60 % of cases there is severe slippage, in 30 % moderate and in 10 % minimal. In a mild slippage fixation with a centrally inserted screw is carried out without attempting reduction. In cases of an unstable slip with moderate slippage one can opt for screw fixation or attempt reduction followed by screw fixation. An attempt is made to reduce and fixate with one or two screws in case of severe slippage (Fig. 9.71). Reduction of an unstable epiphysiolysis should be performed carefully and be preceded by decompression of the hip, to prevent an ischemic femoral head necrosis as much as possible. Here one can choose for puncture of the joint or an open capsulotomy (Table 9.15). If reduction is performed between 24 h and 7 days after the symptoms begin, chances of an ischemic femoral head necrosis are greater than when reduction is done within 24 h or after 7 days. Also, chances of an ischemic head necrosis after reduction increase with the degree of severity of the slippage. Reduction is often not always sufficient. An average improvement of 60 % is obtained in acute slippage and in acute-on-chronic slippage. In 30 % of cases femoral head necrosis occurs and in 30 % of cases a chondrolysis, if the epiphysis is reduced for an acute or an acute-on-chronic slip. After 6–12 months gradual recovery of the cartilage interval occurs spontaneously with improved mobility of the hip (benign form) in half of the

**Table 9.15** Treatment of slipped capital femoral epiphysis

| Slip | Minimal (including pre-slip) | Moderate | Severe |
|---|---|---|---|
| Acute | Fixation in situ | Fixation in situ or decompression, reduction and fixation | Decompression, reduction and fixation |
| Acute-on-chronic | Fixation in situ | Fixation in situ or decompression, reduction and fixation | Decompression, reduction and fixation |
| Chronic | Fixation in situ | Fixation in situ | Fixation in situ |

chondrolysis cases. In the recalcitrant form (the other half of cases) a progressive loss of cartilage interval with premature arthritis develops.

Whereas a slipped femoral head epiphysis affects boys twice as often as girls, chances of a chondrolysis in girls are two to five times greater than in boys.

## Extremely Painful Immobile Hip

Complaint: the pain is so severe that the child cannot move or stand on the leg.

Differential diagnosis:

**acute osteomyelitis**
**septic arthritis**
**recurrent posttraumatic hip dislocation**

Explanatory note: **acute osteomyelitis or septic arthritis.** In children younger than 18 months with a very painful hip there is usually an acute osteomyelitis or septic arthritis. The affected extremity is extremely painful and can not be examined. The hip lies mostly in flexion, abduction and external rotation. The child is usually not ill and has no fever. WBC count and CRP are normal or slightly elevated.

In older children septic arthritis of the hip is less common. These children will also be unable to move the affected leg and will certainly not be walking. These older children tend to be ill and have a fever, as well as an elevated WBC count, CRP and ESR (see Chap. 18).

**Recurrent posttraumatic hip dislocation** A recurrent posttraumatic hip dislocation can occur after a subluxation/dislocation as a result of an extensive trauma, which stretches the posterior side of the capsule. This dislocation is always orientated posteriorly and is painful. Recurrences are more common in children than adults. The affected children tend to be younger than 10 years of age.

Supplementary assessment: (see Chap. 18).

A recurrent pathological hip dislocation can be seen clearly on an anteroposterior X-ray of the pelvis. Additionally, a CT arthrogram or a MRA is indicated to either rule out or show a loose body or a loosened glenoid labrum.

Primary care treatment: none.

When to refer: acute osteomyelitis or septic arthritis of the hip and a recurrent posttraumatic hip dislocation should be referred as quickly as possible.

Secondary care treatment: **acute osteomyelitis** (see Chap. 18).

**Septic arthritis** (see Chap. 18)

**Recurrent posttraumatic hip dislocation** The hip is reduced under anaesthetic and immobilized for 6 weeks with a spica cast with the hips in 45° flexion, neutral rotation and 20° abduction. If up to three recurrences occur after this, then it is best to reef the posterior side of the joint capsule operatively. If there is a loose body or a torn, obstructing acetabular labrum, which inhibits congruent repositioning, the loose body is removed and the acetabular labrum is everted and fixated. After this intervention, spica cast immobilization follows for 6 weeks.

**Differential Diagnosis : Hip Movement Limitations in Babies and Infants**

| **Hip movement limitation away from the midline of the body** | |
|---|---|
| | **Developmental dysplasia of the hip** |
| The hip can be easely dislocated and reduced in a first days of life. | Dislocatable hip (neonatal hip instability) |
| Increased risk of developmental dysplasia of the hip, physical assessment shows no anomalies | Acetabular displasia |
| Abduction limitation of the hip(s), ages 3–4 months, fysical assessment shows no anomalies. | Subluxation/dislocation of the hip |
| **The legs cannot be approximated or with pelvic obliquity** | |
| Adduction limitation of the hip(s). | **Congenital abduction contracture of the hip(s)** |

**Differential Diagnosis: Waddling Gait**

| | **Coxa vara** |
|---|---|
| Thigh gap, limited abduction and enhanced abduction. | Congenital coxa vara, |
| | Acquired coxa vara |
| Thigh gap, enhanced lumbar lordosis. | **Bilateral hip dislocation** |

**Differential Diagnosis: Snapping Hip**

| | |
|---|---|
| Snapping sensations around the greater trochanter. | **Snapping of the iliotibial tract.** |
| Snapping sensations around the groin. | **Snapping of the iliopsoas tendon** |
| Repeated dislocation of the hip. | **Recurrent hip subluxation/dislocation** |
| Associated with hypermobility disorders. | Voluntary hip subluxation/dislocation |
| Not associated with hypermobility disorders and non traumatic. | Habitual hip subluxation/dislocation |
| Locking symptoms. | **Osteochondritis dissecans** |
| Pain when descending stairs. | **Acetabular labral tear** |

**Differential Diagnosis: Hip Pain**

| **Vague pain around the groin, pelvis or upper leg** | |
|---|---|
| Changeable pain, especially after sports. | **Bone tumor** |
| Pain in the groin. | **Ischiopubic osteochondritis** |
| | **(Van Neck disease)** |
| **Hip pain and limited internal rotation** | |
| Transient hip pain in children aged 2–13 years. | **Acute transient synovitis of the hip** |
| | (coxitis fugax, irritable hip syndrome, observation hip, toxic synovitis, acute transient epiphysitis) |
| Persistent hip pain in children aged 2–13 years. | **Legg-Calvé-Perthes disease** |
| | **(Perthes disease, Waldenström disease, coxa plana)** |
| Persistent hip pain in children aged 10–16 years. | **Chronic slipped capital femoral epiphysis** |
| **Hip pain and movement limitations in all directions** | |
| Movement limitations of the hip in all directions. CE angle > 60°. | **Acetabular protrusion (Otto pelvis, arthrokatadysis),** |
| Progressive destruction of the cartilage. | **Chondrolysis** |
| After a slipped capital femoral epiphysis, infection, trauma, prolonged spica cast immobilization. | Acquired type |
| Cause unknown. | Idiopathic type |
| **Hip pain after a trivial trauma** | |
| Sudden pain in the hip. The patient can no longer stand. | **Fractured unicameral bone cyst** |
| | **(juvenile-, solitary-, simple bone cyst)** |
| | **Acute and acute-on-chronic slipped capital femoral epiphysis** |
| **Extremely painful immobile hip** | |
| The hip is extremely painful and the leg cannot be moved. | **Acute osteomyelitis,** |
| | **Septic arthritis** |
| Previous traumatic hip dislocation. | **Recurrent posttraumatic hip** |
| | **subluxation/dislocation** |

# Knee

# 10

## Knee Misalignment

### Bowleg

Complaint: unilateral or bilateral bowleg(s).

Assessment: the lower leg is in adduction with respect to the upper leg. Many researchers use the distance between the medial femoral condyles as a measure for genua vara. There has been no longitudinal research on this measurement. It is better to measure the angle between the upper and the lower leg.

Differential diagnosis:

**physiological genua vara**
**idiopathic excessive genua vara**
**Blount disease (tibia vara, osteochondrosis deformans tibiae)**
infantile type
adolescent type
**rickets (infantile osteomalacia)**
**skeletal dysplasia**
**focal fibrocartilaginous dysplasia**
**dysplasia epiphysealis hemimelica (Trevor disease)**
**infected or fractured growth plate**

Explanatory note: **physiological genua vara.** There is a varus position in the knees up to 15° physiologically from birth until the age of 1 year. This physiological development is bilateral and appears to increase when the child stands and walks (Fig. 10.1). The varus position is localized in the distal part of the femur as well as in the proximal part of the tibia. There is often a lateral bowing of the tibiae. In many cases the bowlegs are optically accentuated by an enhanced external rotation in the hip joints and an internal torsion of the lower legs. In most cases bowlegs undergo spontaneous, slow correction and the legs straighten up when the child is 1½–2 years old, (Fig. 10.2). In isolated cases the spontaneous correction is completed only after ages 3–4. The best method to see whether the bowlegs will correct spontaneously is by placing the child lying in a supine position on the examination table so that the medial malleoli are approximated. It is almost certain that the bowleg posture will correct spontaneously if the knees straighten when you apply pressure on the outer sides.

**Idiopathic excessive genua vara** This deformity is bilateral and most common in Asians and may be familial. Operative treatment is seldom required. It is not clear from the literature whether and to what degree there is an increased chance of degenerative arthritis of the medial compartment.

**Blount disease** Blount disease is a rare anomaly. It is a metaphyseal developmental disorder on the medial side in the proximal part of the tibia. The disease can cause unilateral or bilateral


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_10

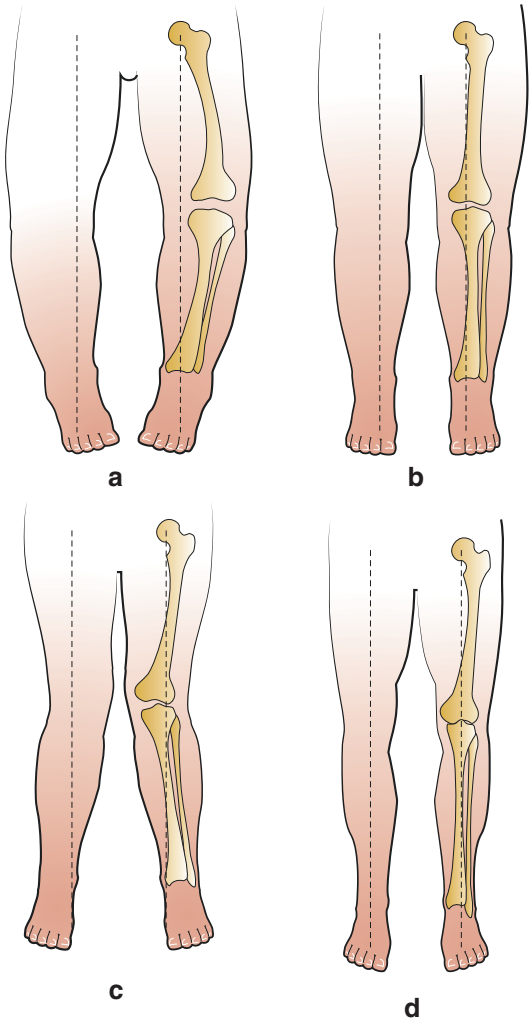



**Fig. 10.1** (**a**) A physiological genua vara may occur up to 15° from the birth until the age of 1 year. (**b**) Once the child reaches the age of 1½–2 years, the genua vara has generally corrected slowly. The legs are now straight. (**c**) During the second and third years of life knock knees develop. (**d**) The valgus deviation corrects itself gradually, so that by the age of 7 a normal femorotibial valgus angle has developed in 95 % of cases

bowlegs at the age of 1½–3 years (infantile type), as well as starting at 8 years of age (adolescent type) (Table 10.1). A juvenile type is also recognized that occurs between the ages of 4 and 8, but it is probably an infantile type that has been diagnosed too late. That type is always bilateral. The disease affects girls as much as boys, and is most common among children of African origin.

*Infantile type* Radiological assessment is indicated when both legs are not straight at the age of 1½–2 years, or when bowlegs develop after that age. The infantile type is bilateral in 60 % of cases (Fig. 10.3). A unilateral bowleg is always abnormal. The infantile type often presents with internal torsion of the tibia. Six stages can be distinguished radiologically (Figs. 10.4 and 10.5).

*Adolescent type* The adolescent type is generally less pronounced than the infantile type. The varus position in adolescence never exceeds 20°. In 20 % of cases this type is bilateral (Fig. 10.6). In the unilateral adolescent type a difference in leg length of 1–3 cm may be seen (Fig. 10.7).

**Rickets** Bowlegs can also be caused by a lack of vitamin D. This is known as rickets, infantile osteomalacia or the English disease. Children don't generally have a vitamin D deficiency in developed countries and the disease is rare. It tends to be caused by an hereditary enzymatic defect (pseudovitamin D-deficiency I), a congenital resistance (pseudovitamin D-deficiency II) or a renal phosphate loss in familial phosphate diabetes (X-chromosome dominant hereditary renal tubular disorder).

In addition to bowlegs, physical assessment shows skull thinning (craniotabes) as well as broad wrists, knees and ankles. As a result of the broadening of the junction between the ribs and the rib cartilage, a "rosary" forms on the anterior side of the ribs at the level of the sternum. There may also be a pigeon breast and antecurvatum deformity of the lower legs. X-rays show widened growth plates and the metaphyses appear frayed (Fig. 10.8).

**Skeletal dysplasia** Bowlegs can be part of generalized skeletal anomalies such as achondroplasia and multiple epiphyseal dysplasia[1]. A shorter body height (Fig. 10.9) is seen in skeletal dysplasia.

[1] See Appendix.

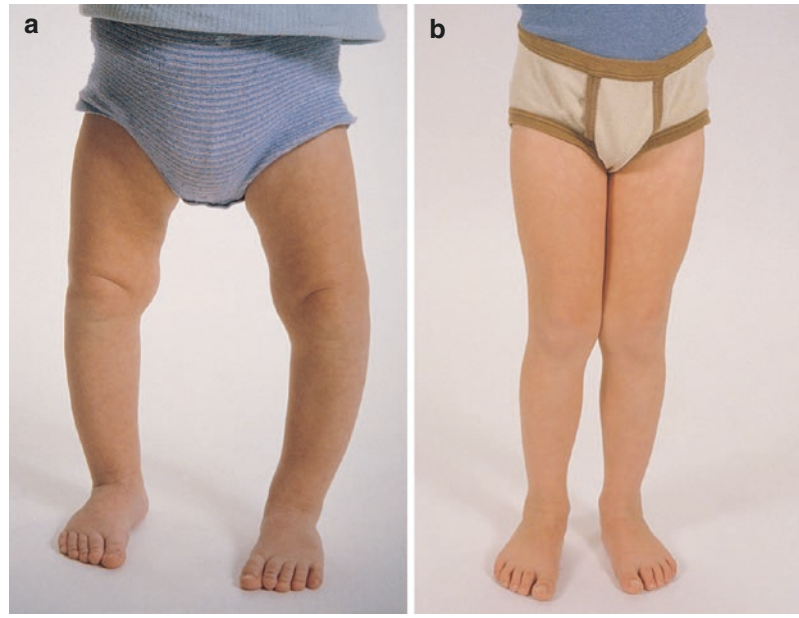



**Fig. 10.2** (**a**) One-year-old boy. Physiological genua vara. (**b**) The same boy as in Fig. 10.2a, now 2 years old. The bowlegs have spontaneously corrected. There is even a slight light valgus deviation of both knees

**Table 10.1** Differences between infantile and adolescent type of Blount disease

| | Infantile type | Adolescent type |
|---|---|---|
| Varus > 20° | Nearly always | Never |
| Internal torsion of the tibia | Frequent | Seldom |
| Recurrence chance after treatment | Frequent | Seldom |

**Focal fibrocartilaginous dysplasia** In focal fibrocartilaginous dysplasia there is a unilateral genu varum resulting from a local abnormality on the medial side in the proximal part of the tibia (Fig. 10.10). MRI assessment shows an elliptical cortical bone defect. Microscopical assessment shows fibrous tissue with small areas of cartilage. This abnormal tissue stunts growth on the medial side of the tibia, but on the other side it keeps growing and a bowleg develops.

**Dysplasia epiphysealis hemimelica** This is an osteochondral overgrowth of an epiphysis or articular cartilage. It is a rare, non-painful condition with an incidence of 1 in 1,000,000. The condition manifests itself mostly between the ages of 2 and 14 years. It is three times more common in boys than in girls. There is a hereditary factor. The anomaly is intra-articular and remains limited to half of the extremity. The medial side is affected twice as often as the lateral part, and in two-thirds of the patients the condition is present in several epiphyses simultaneously. The most common localizations are the distal part of the femur, the distal part of the tibia and talus. A varus deformity will develop if the anomaly is localized on the lateral side. A valgus deformity will develop if the anomaly is localized on the medial side (Fig. 10.22). There is also movement limitation and a swelling of the knee, in case of a bowleg on the lateral side. Radiological assessment shows a lobe-shaped swelling on the lateral side of the distal femoral epiphysis, accompanied by multiple calcifications. Histologically speaking, the anomaly cannot be distinguished from an extra-articular osteochondroma. Malignant degeneration has never been described.

**Infected or fractured growth plate** A bowleg can also be caused by an infection or a fracture, in which the growth plate is damaged on the medial side just above or just below the knee. Part of the cartilage of the growth plate is replaced by bony tissue, creating a bony connection between the epiphysis and metaphysis. If the bony connection

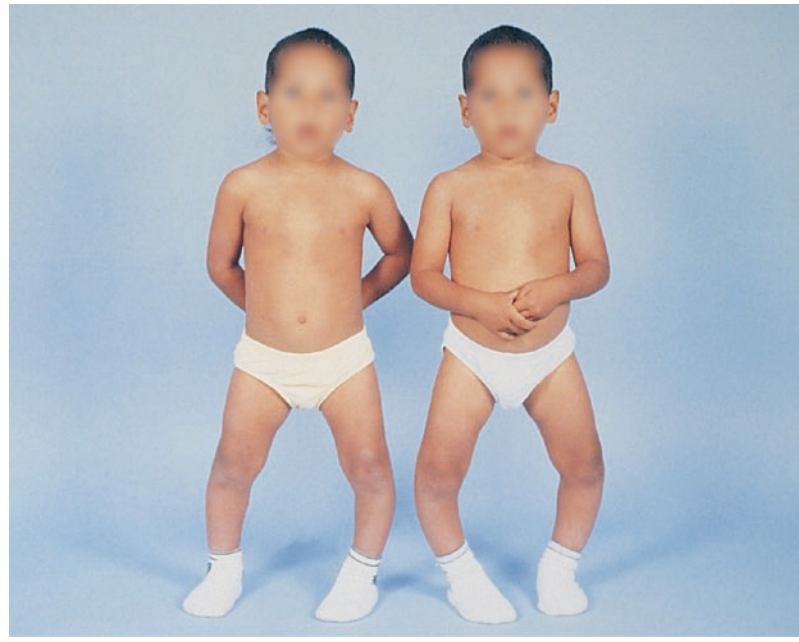


**Fig. 10.3** Bilateral bowlegs as a result of an infantile type of Blount disease in 3-year-old identical twins

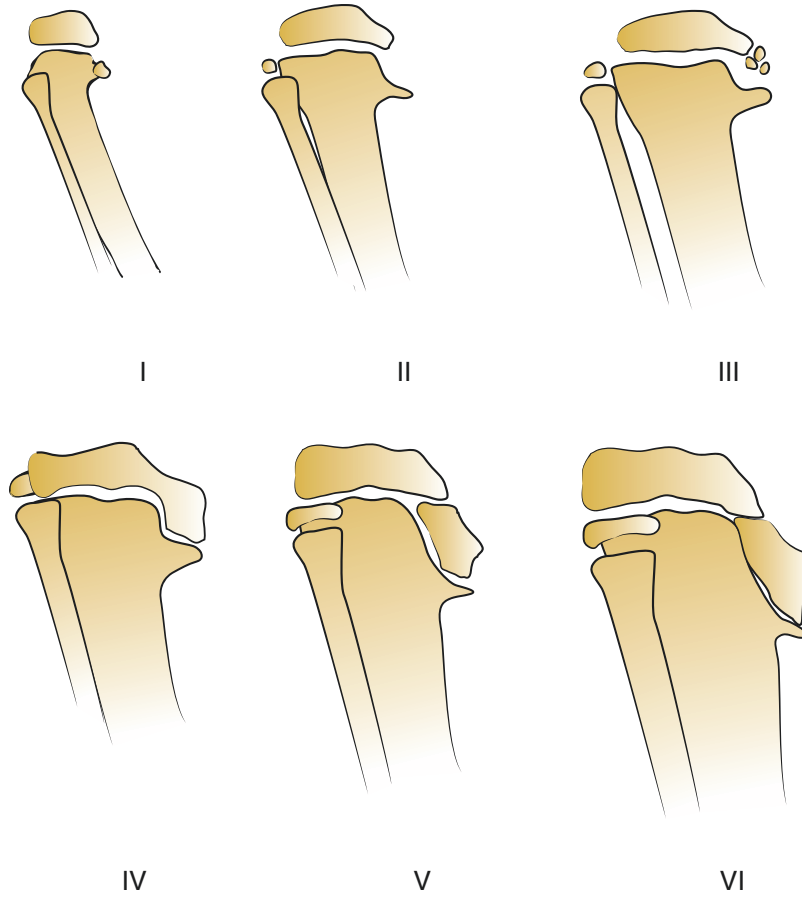



**Fig. 10.4** Langenskiöld and Riska classification for infantile Blount disease (Redrawn from: Langenskiöld A, Riska EB. Tibia vara (osteochondrosis deformans tibiae): a survey of seventy-one cases. J Bone Joint Surg Am. 1964;46:1405–20). *I* beak-shaped or irregular ossification of the medial side of the proximal tibial metaphysis. *II* and *III* progressive sagging of the medial side of the proximal tibial metaphysis. *IV* also sagging of the medial part of the proximal tibial epiphysis. *V* sagging of the medial part of the proximal tibial metaphysis and proximal tibial epiphysis is halted, and the medial and lateral parts of the proximal tibial epiphysis are now separated. *VI* a bony connection has appeared between the medial side of the proximal tibial metaphysis and epiphysis

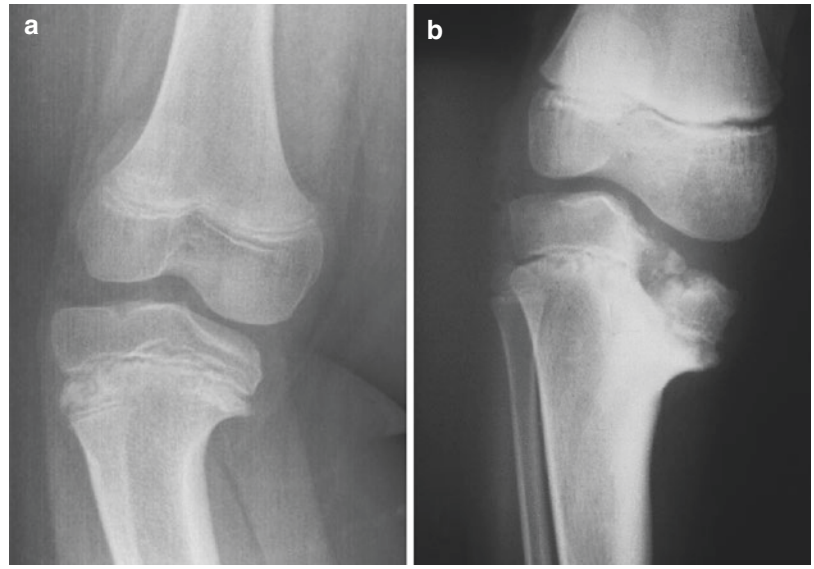



**Fig. 10.5** (**a**) Blount disease (infantile type) stage III. (**b**) Blount disease (infantile type) stage VI

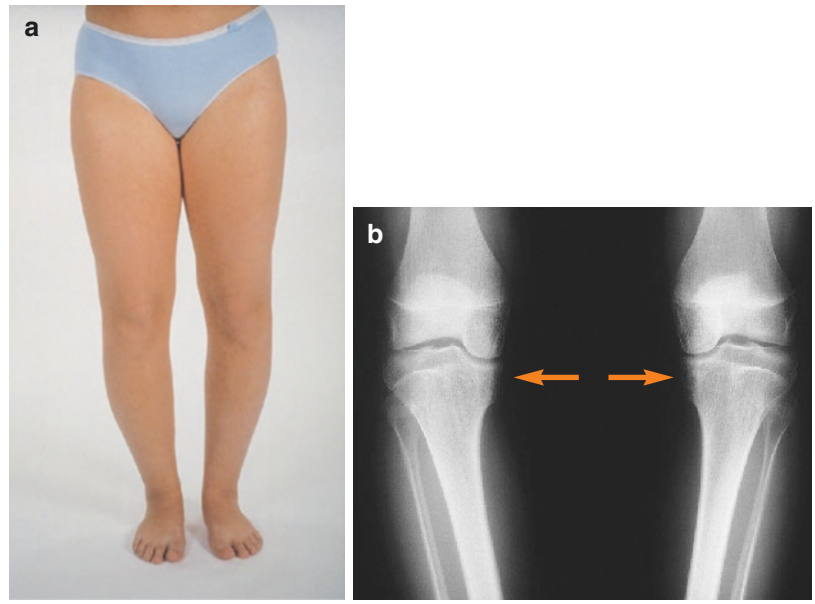



**Fig. 10.6** (**a**) Bilateral Blount disease (adolescent type). This 15-year-old girl developed genua vara within a half year due to a bilateral Blount disease. (**b**) Anteroposterior X-ray of the knees of the same girl as in 10.6a. On the medial side a bony connection has appeared between the proximal tibial metaphysis and epiphysis (*arrow*). The lateral part of the proximal tibial growth plate is intact and keeps growing causing a bowleg

is centrally located, then only leg shortening occurs. If the bony bridge is medially located, then the medial side of the corresponding growth plate stops length growth and the lateral side keeps growing so a bowleg develops (Fig. 10.11).

Supplementary assessment: standing anteroposterior and lateral X-rays of the legs are requested in the case of unilateral or bilateral bowlegs that have not straightened out or become worse by the age of 2 years, as well as when one or two bowlegs develop in adolescence. In the infantile type the metadiaphsyeal angle in the proximal part of the tibia should be measured when in doubt as to whether one is dealing with physiological bowlegs or Blount disease (stage 1) (Fig. 10.12). The chances of developing Blount disease are less than 2% (Fig. 10.13) if this angle is 11° or less. There is a strongly increased risk if the

**Fig. 10.7** An unilateral genu varum is always pathological. This 14-year-old girl developed a leftsided bowleg within half a year due to Blount disease (adolescent type)

angle is between 11° and 16°. Blount disease is confirmed if the angle is greater than 16° (Fig. 10.14). At the level of the growth plate in the proximal part of the tibia there is a bony connection between the medial side of the epiphysis and the metaphysis in an adolescent type of Blount disease. The lateral part of the growth plate is intact. There is a loss of height on the medial side of the proximal tibial epiphysis (Fig. 10.6). A MRI is also requested for focal fibrocartilaginous dysplasia and dysplasia epiphysealis hemimelica.

It is recommended to carry out blood tests for calcium, phosphate, parathyroidhormone, 25 hydroxy vitamin D and 1,25 dihydroxy vitamin D if rickets is suspected. A supplementary CT-scan is requested for growth plate damage caused by an infection or a fracture.

Primary care treatment: the parents should be reassured in cases of physiological genua vara.

When to refer: if genua vara persists or develops after the age of 18 months. A unilateral bowleg is always abnormal and should always be referred. The child should first be referred to a pediatrician in a case with rickets.

Secondary care treatment: **idiopathic excessive genua vara.** One should consider a hemi-epiphysiodesis on the lateral side.

**Blount disease (infantile type)** There are chances of spontaneous recovery in stages I and II of Blount disease The chances of recovery are greater (90%) if stages I and II are treated with antivarus splints (Fig. 10.15). It seems to make no difference whether splint treatment is applied for the entire day or only at night. We advise prescribing antivarus splints only for night use for children younger than 3. A proximal tibial valgus osteotomy (just distal to the tibial tuberosity) with a distal fibulotomy is indicated if there has been no recovery within 1 year or if the child is older than 3. The same applies in a child with Blount disease stage III. A similar intervention should be carried out before the age of 4 in stages I, II and III. The chances of recurrence are higher if the operation is postponed until after that age, A valgus position of 5°–10° should be aimed for. For stages I, II and III a lateral hemi-epiphysiodesis of the growth plate of the proximal part of the tibia could be an alternative. This prevents progression of the anomaly, but correction will not be achieved. What's more, it will stagnate the growth from this growth plate. In stages IV and V children are at least 6 years of age. A growth plate distraction of the medial part of the proximal tibial growth plate and also a proximal tibial valgus osteotomy just distal of the tibial tuberosity with a fibulotomy, may be considered, in which the proximal tibial growth plate on the medial side has not yet closed.

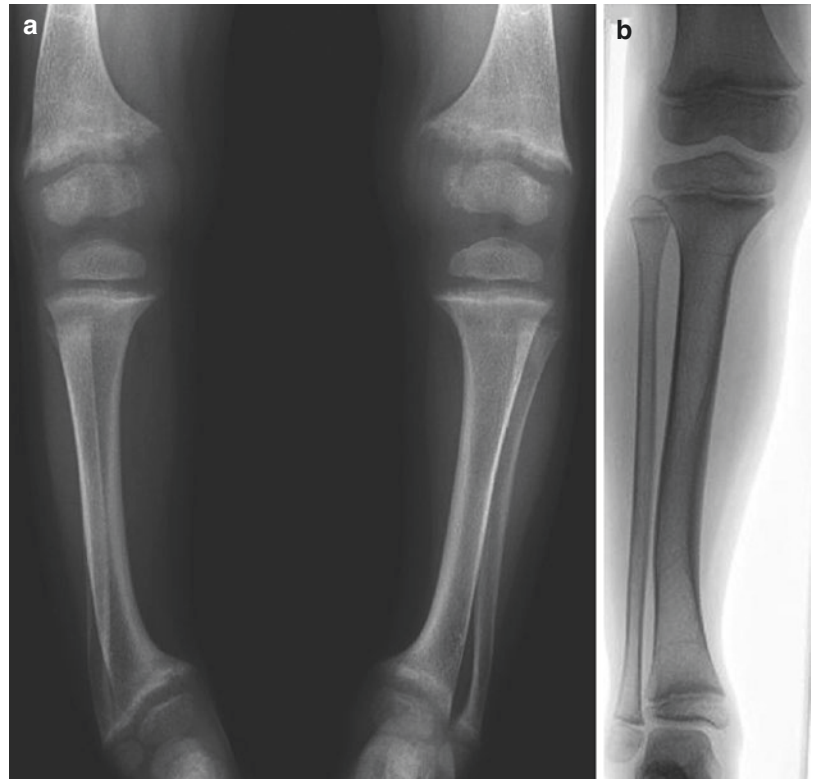



**Fig. 10.8** (**a**) Bowlegs as a result of rickets (pseudovitamin D-deficiency). In this patient the distal femoral and distal tibial growth plates appear frayed. (**b**) One year after treatment of the pseudovitamin D-deficiency the appearance of the growth plates is normal and the varus deviation has spontaneously corrected

In stage VI the child is older than 9 and there is a bony connection between the epiphysis and the metaphysis on the medial side of the proximal tibial growth plate. The position of the proximal part of the tibia is corrected by a corrective osteotomy of the medial tibial plateau and a proximal tibial valgus osteotomy with a distal fibulotomy (Fig. 10.16). The lateral part of the proximal tibial growth plate of the affected tibia is destroyed to prevent recurrence. An epiphysiodesis of the proximal tibial growth plate on the non-affected side may be performed in addition to prevent a difference in leg length.

**Blount disease (adolescent type)** The adolescent type of this anomaly is not as pronounced as in the infantile type (Table 10.1). A definitive hemi-epiphysiodesis in which the lateral part of the proximal tibial growth plate is destroyed is usually sufficient. A proximal tibial valgus osteotomy can be performed in combination with a distal fibulotomy for a severe type. If necessary, an epiphysiodesis of the contralateral proximal tibial growth plate should also be carried out in order to prevent progression in the leg length difference depending on the growth expectations.

**Rickets** Rickets should first be treated by a pediatrician. Spontaneous improvement of the deformity may occur after treatment of the underlying cause (Fig. 10.8), as healthy growth plates redirect themselves perpendicular to the weightbearing lines (Hueter-Volkmann law, 1862). A temporary hemi-epiphysiodesis of the lateral side of the growth plates in the distal part of the femur and/or the proximal part of the tibia will be performed using clamps, eight-plates or screws depending on the localization in severe cases where no improvement occurs. The fixation materials are removed as soon as the legs are straight.

**Skeletal dysplasia** Operative intervention is indicated in 50% of achondrodysplasia[2] cases with such a misalignment. A temporary epiphysiodesis of the lateral side of the growth plates in the distal

[2]See Appendix.

part of the femur and the proximal part of the tibia can be performed using clamps or eight-plates if the child is still growing. A correction osteotomy is an option if the child is no longer growing,

**Focal fibrocartilaginous dysplasia** At first the bowleg will increase, but between the ages of 4 and 8 the defect will be spontaneously filled up with bony tissue; the bowleg position will tend to correct spontaneously if there is a varus deformity of less than 30°. A proximal valgus osteotomy of the tibia in combination with a distal fibulotomy may be performed if there is no spontaneous correction between the ages of 4 and 8.

**Dysplasia epiphysealis hemimelica** Wedge excision at the level of the deformity, thus correcting the abnormal position. The chances of recurrence are great.

**Infected or fractured growth plate** This bony bridge can be removed operatively if the bony connection between the epiphysis and metaphysis resulting from growth plate damage due to infection or fracture amounts to less than one-third of the total area of the growth plate. The growth plate usually recovers full function after this, and in a large number of cases the misalignment will correct spontaneously because the


**Fig. 10.9** (**a**) Bowlegs in achondroplasia. The arms and legs are relatively short with respect to the rest of the body. Normally, the hands reach about halfway down the upper legs. (**b**) Patients with achondroplasia (see appendix) have a lumbar hyperlordosis

growth plate tries to direct itself perpendicular to the load line (Hueter-Volkmann law, 1862). In some cases, however, it is not possible to remove the bony bridge, or because the bone bridge consists of more than one-third of the total area of the total growth plate. In such cases it is best to destroy the lateral part of the growth plate and perform a correction osteotomy. The difference in leg length should be subsequently corrected at the appropriate time.

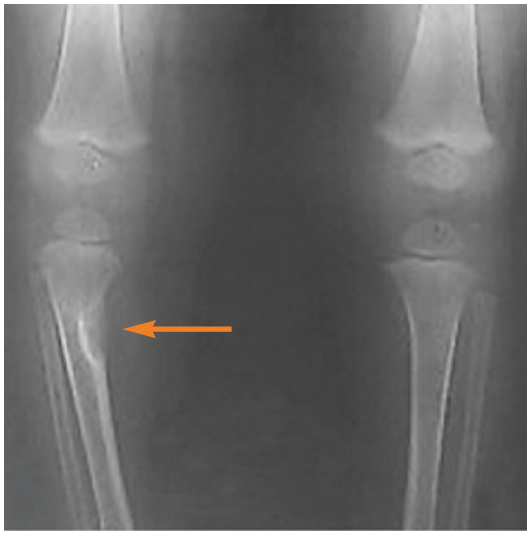


**Fig. 10.10** Right-sided bowleg caused by focal fibrocartilaginous dysplasia (*arrow*)

## Knock Knee

Complaint: unilateral or bilateral knock knee(s).

Assessment: the lower leg is in abduction with respect to the upper leg.

Differential diagnosis:

**physiological genua valga**
**idiopathic excessive genua valga**
**rickets (infantile osteomalacia)**
**skeletal dysplasia**
**multiple osteochondromata (exostoses)**
**dysplasia epiphysealis hemimelica (Trevor disease)**
**infected or fractured growth plate**
**fractured proximal tibial metaphysis**

Explanatory note: the distance between the medial malleoli is usually used to determine the degree of knock knee. There are no publications with a longitudinal assessment of

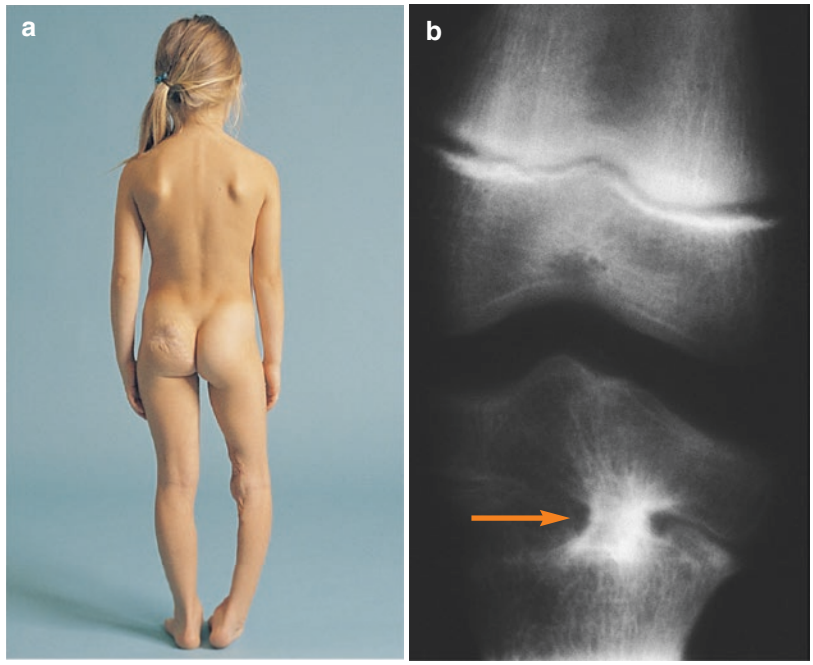



**Fig. 10.11** (**a**) Unilateral right-sided bowleg resulting from a bone fracture in the right proximal tibial growth plate. (**b**) Anteroposterior tomogram of the right knee, same patient as in Fig. 10.11a. Bony connection (*arrow*) between the epiphysis and metaphysis (bone bridge). In this case the medial side of this growth plate stops growing. The lateral side of the growth plate provides continuous growth causing a bowleg

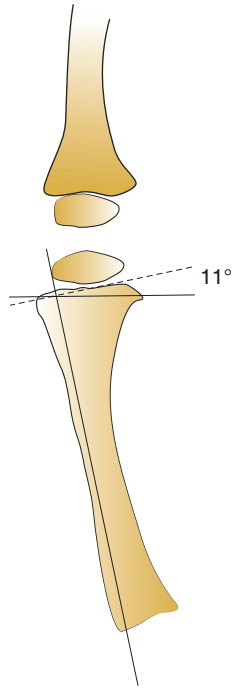



**Fig. 10.12** Metadiaphyseal angle. This is the angle between the longitudinal axis of the tibia and the line that connects the most prominent part of the lateral and medial sides of the proximal tibial metaphysis

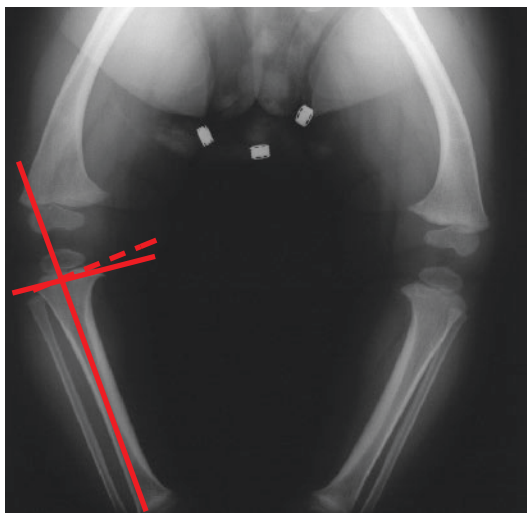


**Fig. 10.13** Anteroposterior X-rays of both legs with a varus position on both sides, both in the distal part of the femur and the proximal part of the tibia. The metadiaphyseal angle is less than 11°, so in this case we probably are dealing with physiological bowlegs. These bowlegs correct spontaneously

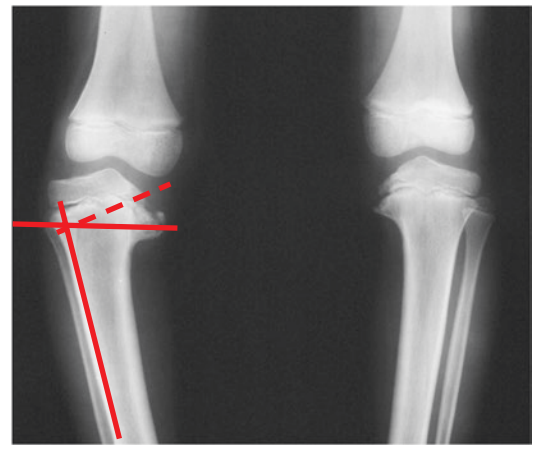


**Fig. 10.14** Anteroposterior X-rays of both legs. In this case the metadiaphyseal angle is 20°, indicating Blount disease

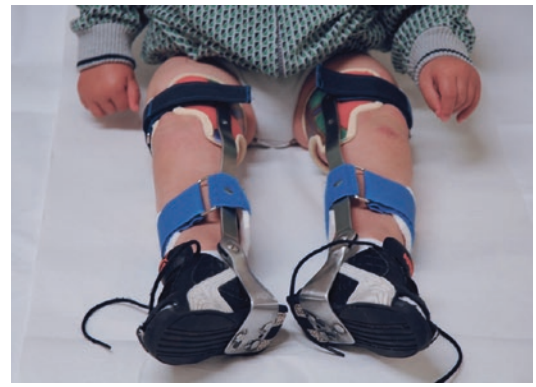


**Fig. 10.15** Antivarus knees splints

this deformity. The distance is also influenced by the leg length. The longer the legs, the larger the intermalleolar distance will be without an increase in the knock knee.

This measurement is even more unreliable in the case of a unilateral knock knee. In a physical assessment it is much better to measure the angle between the thigh and lower leg with a goniometer and to use the femoral tibial angle as a measurement.

**Physiological genua valga** During the second and third years of life knock knees develop slowly with an average annual rate of 12° between the upper and lower leg after the legs with a

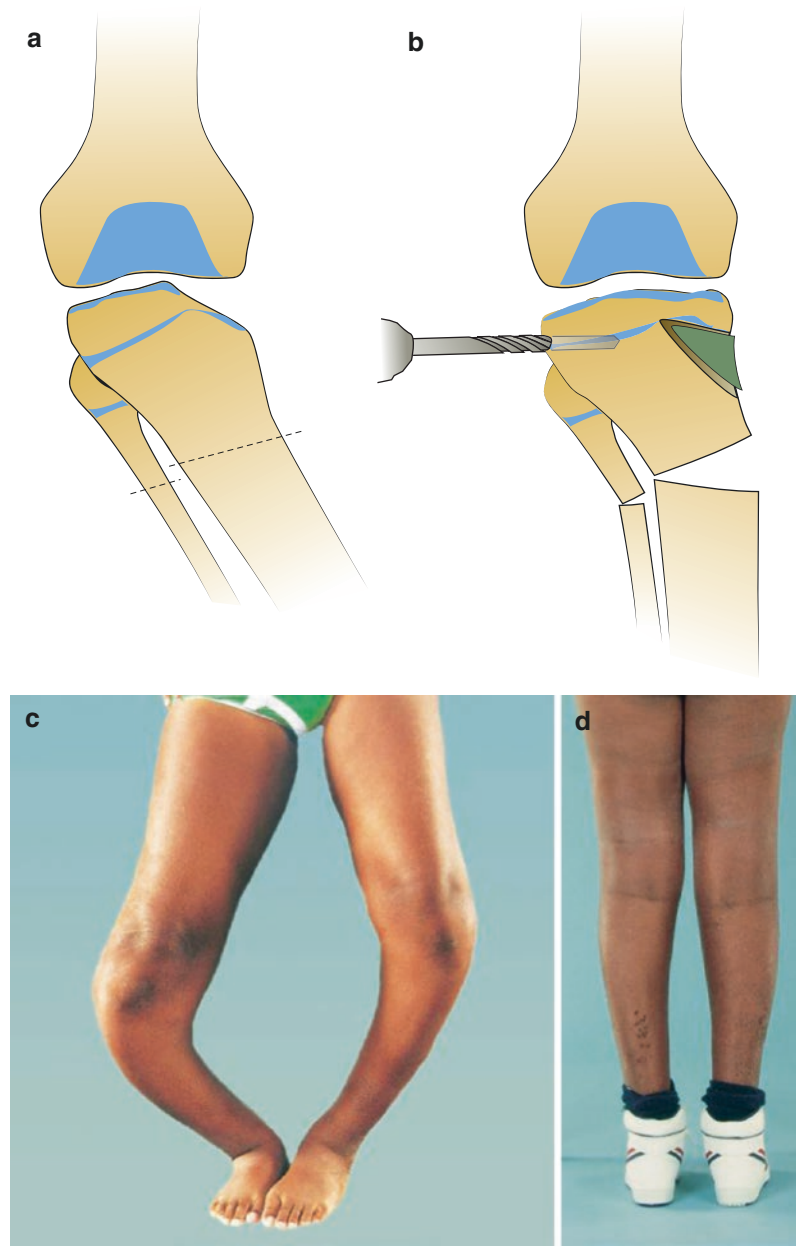



**Fig. 10.16** (**a**) Stage VI of infantile type of Blount disease. (**b**) The proximal medial part of the tibia is corrected by a straightening osteotomy of the medial tibial plateau and a proximal tibial valgus osteotomy together with a distal fibulotomy. The lateral part of the proximal tibial growth plate is destroyed. (**c**) Bowlegs in a Blount disease stage VI. (**d**) The same patient as in Fig. 10.16c after operative correction

physiological genua vara have straightened by the age of 18 months, This physiological development is bilateral. A knock knee is often optically enhanced by a simultaneous development of external rotation in the lower legs and in some cases an enhanced internal rotation in the hip joints. This can also be accentuated in cases with thick thighs and flatfeet. The valgus position corrects itself gradually, so that by the age of 7 years a normal valgus angle of 8° in girls and 7° in boys has developed in more than 95 % of cases (Fig. 10.17).

Parents ask for advice more often with knock knees than with bowlegs. This is because in physiological bowlegs children have been on their feet and walking only for a short time, if at all. In older children, the legs are longer and the "anomaly" is more noticeable.

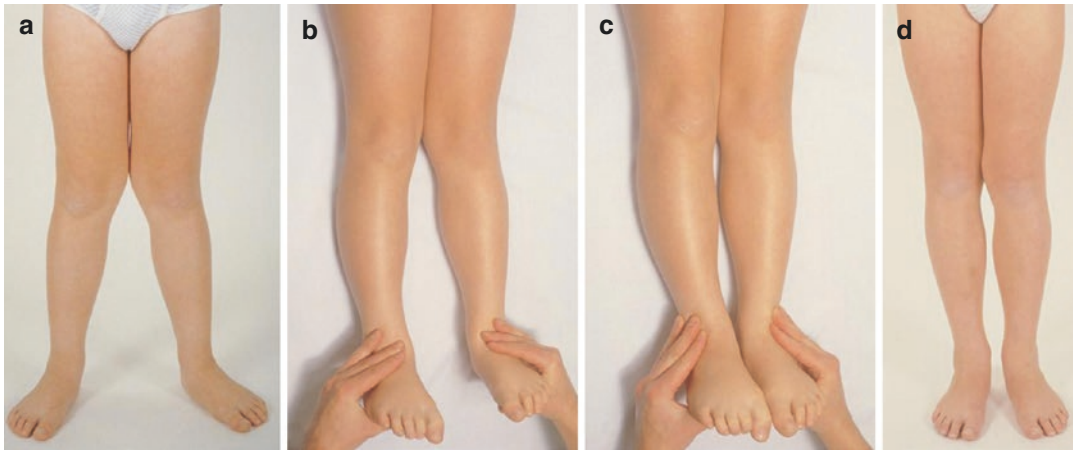



**Fig. 10.17** (**a**) Four-year-old boy with physiological genua valga, accentuated by thick thighs and flatfeet. (**b**) The same boy as in Fig. 10.17a placed on the examination table in such a way that the patellae point forward. The effect of any existing flatfeet is neutralized in this situation, so that the genua valga are already less noticeable. You say to the parents: "that's the way they are now". (**c**) Next, you grab both ankles and press the medial malleoli against each other, then say to the parents: "that's the way they will be in the future". (**d**) The same boy as in Fig. 10.17a, now 8 years old. The knock knees have spontaneously corrected

A good method to show parents that the knock knees will correct spontaneously is to place the child supine on the examination table so that the medial femoral condyles are approximated. Next, encompass both ankles and try to press the medial malleoli against each other. The chances of spontaneous correction are great if you can do that, (Fig. 10.17). The chances are great that the knock knees will not correct spontaneously if this is not possible.

**Idiopathic excessive genua valga** If there is still a genua valga in excess of 10° the chances of spontaneous recovery are minimal after the physiological period, after 7 years of age, (Fig. 10.18)

**Rickets** Rickets can give rise to bowlegs as well as knock knees (see pp. 172).

**Skeletal dysplasia** Knock knees can also develop in addition to bowlegs (Fig. 10.19) in generalized bony anomalies such as achondroplasia[3] and multiple epiphyseal dysplasia,[3] (see pp. 172).

**Multiple osteochondromata** This is a developmental disorder in the periphery of the epiphyseal plates. The benign bony growth develops mostly with a cartilaginous cap. The anomaly increases in size until the patient stops growing. This problem may be solitary but can also appear in

**Fig. 10.18** A 9-year-old girl with idiopathic excessive genua valga

[3]See Appendix

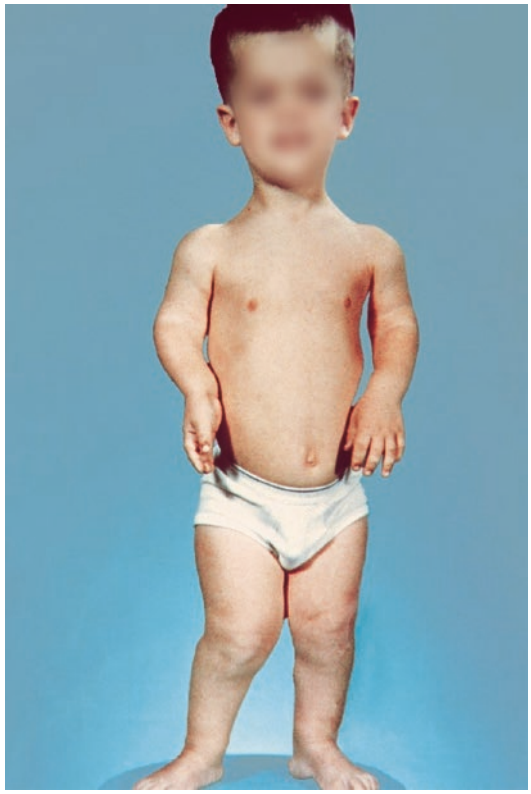


**Fig. 10.19** Knock knees in achondroplasia

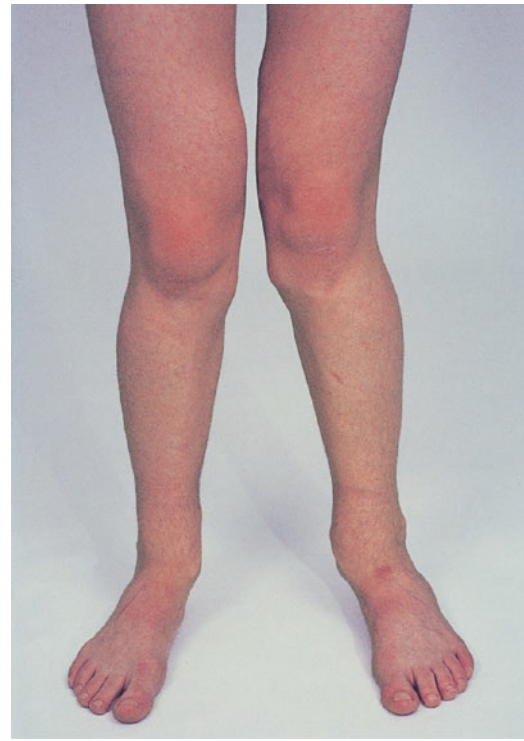


**Fig. 10.20** Genua valga as a result of multiple osteochondromata (exostoses) around the knees

multiple areas. Multiple osteochondromata are involved in a hereditary disease in more than 60 % of cases. Hereditary multiple exostoses (or osteochondromata), are abbreviated as HME or HMO. There is often a knock knee position in multiple osteochondromata (Fig. 10.20). This is caused by a large osteochondroma in the proximal part of the fibula. There is reduced growth in length of the fibula, as a result of which the bone works as a brake, causing a valgus deformity in the proximal part of the lower leg (Fig. 10.21).

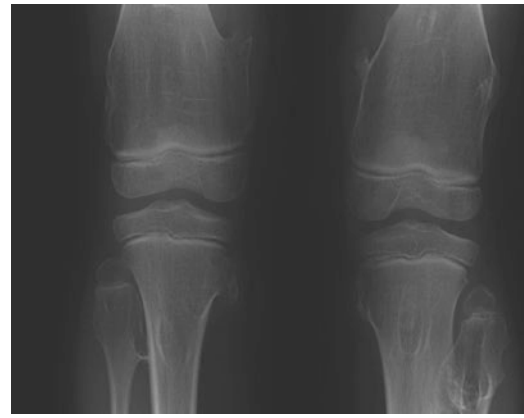


**Fig. 10.21** Osteochondromata in the proximal part of the fibula cause disturbed growth of the fibula; the fibula works as a brake and causes a valgus position of the knee

**Dysplasia epiphysealis hemimelica** This is an osteochondral overgrowth of the epiphysis on the medial side of the femur (Fig. 10.22). Besides the valgus 0 there is a limitation in movement and a swelling on the medial side of the knee (for explanatory note, see pp. 173).

**Infected or fractured growth plate** A unilateral knock knee is always abnormal. Part of the cartilage of the growth plate will be replaced by bone tissue if there is damage to the growth plate caused by an infection or fracture. A knock knee develops if this bony connection appears on the lateral side of the distal femoral or proximal tibial

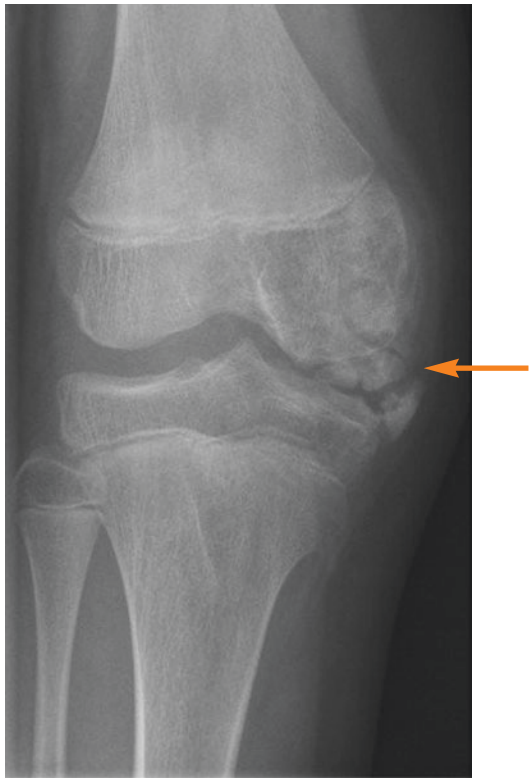


**Fig. 10.22** Hemimelic epiphyseal dysplasia. This is an intra-articular osteochondroma around the medial femoral condyle (*arrow*) that causes a knock knee

growth plate in the distal part of the femur or the proximal part of the tibia.

**Fractured proximal tibial metaphysis** A genu valgum may also be the result of a fracture on the medial side in the proximal tibial metaphysis (Fig. 10.23). A valgus deformity develops in a large number of cases after cast immobilization. This is probably caused by interposition of the periosteum at the level of the fracture, which will not allow closed (noninvasive) anatomical reduction.

Supplementary assessment: anteroposterior standing and lateral X-rays of the legs, and for hemimelic epiphyseal dysplasia a MRI. If rickets is suspected it is recommended to do blood tests to access calcium, phosphate, parathormone, 25 hydroxy vitamin D and 1,25 dihydroxy vitamin D. A supplementary CT-scan is requested in the case of growth plate damage caused by an infection or a fracture.

Primary care treatment: in the case of physiological knock knee you should reassure the parents and tell them that their child should come back if the deformity is still present at the age of 10.

When to refer: if there is a genua valgum with a femorotibial angle of 10° or more after the age of 10. The child should be referred to a pediatrician in the case of rickets.

Secondary care treatment: **idiopathic excessive genua valga.** One may consider a temporary hemi-epiphysiodesis using clamps, an eight-plate or screws on the medial side of the growth plates in the distal part of the femora for 11-year-old girls and 12-year-old boys if there are excessive knock knees with an angle of 10° or more between the upper and lower leg, (Fig. 10.24). The lateral part of the growth plates keep on growing, so that the leg straightens. The osteosynthetic materials are removed as soon as the knees are straight (usually 3–9 months after the initial operation). This operation is often carried out for cosmetic reasons. It is not clear in the literature which valgus position one should accept as an increased risk for degenerative arthritis in the lateral joint compartment. Once the child is fully grown, pathological knock knees can no longer be corrected by a temporary hemi-epiphysiodesis – instead, a distal femoral varus osteotomy should be considered.

**Rickets** Rickets should be initially treated by a pediatrician. Spontaneous improvement of the deformity occurs after treatment of the underlying cause. In this situation the growth plates are directed perpendicularly to the weight-bearing lines (Hueter-Volkmann law, 1862). A temporary hemi-epiphysiodesis on the medial side of the affected epiphysis or a correction osteotomy may be necessary in severe cases where no improvement occurs.

**Skeletal dysplasia** Temporary hemi-epiphysiodesis of the medial side of the growth plate in the distal part of the femur and proximal part of the tibia. A correction osteotomy may be considered

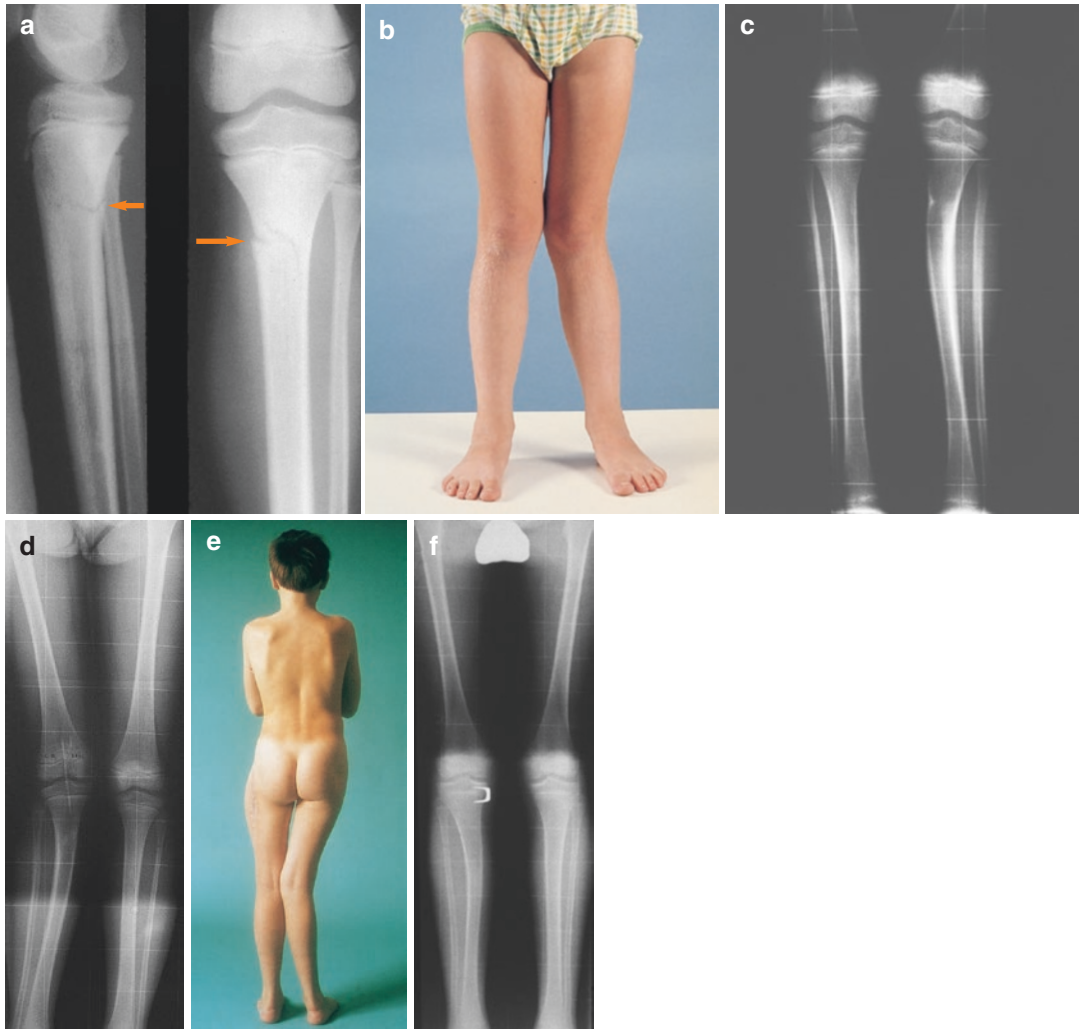



**Fig. 10.23** (**a**) Lateral and anteroposterior X-rays of the left lower leg in a 5-year-old boy with a fracture of the left proximal tibial metaphysis. (**b**) Same patient as in Fig. 10.23a. After healing of the fracture a tibia valgum developed. (**c**) The same patient as in Fig. 10.23a, b. The tibia is now S-shaped. This can only be seen on the X-rays. The leg looks straight on physical assessment. (**d** and **e**) In this case no spontaneous correction has occurred. (**f**) In those cases where no spontaneous correction occurs a temporary hemi-epiphysiodesis on the medial side in the proximal tibial growth plate can be performed

if correction is insufficient once the child is fully grown.

**Multiple osteochondromata** In order to correct or stop the valgus misalignment a temporary hemi-epiphysiodesis on the medial side of the growth plates in the proximal part of both tibiae and a partial resection of the fibula shaft including the periosteum should be performed. The clamps or eight-plate should never be allowed to remain in place for more than 2 years, or a bony bridge will develop in the growth plate. At some point it often becomes necessary to remove the osteochondromata as much as possible and carry out a proximal tibial varus osteotomy at the end of the growth period. In young children there is very high risk of recurrence.

**Dysplasia epiphysealis hemimelica** Wedge excision at the level of the anomaly, thus correcting the

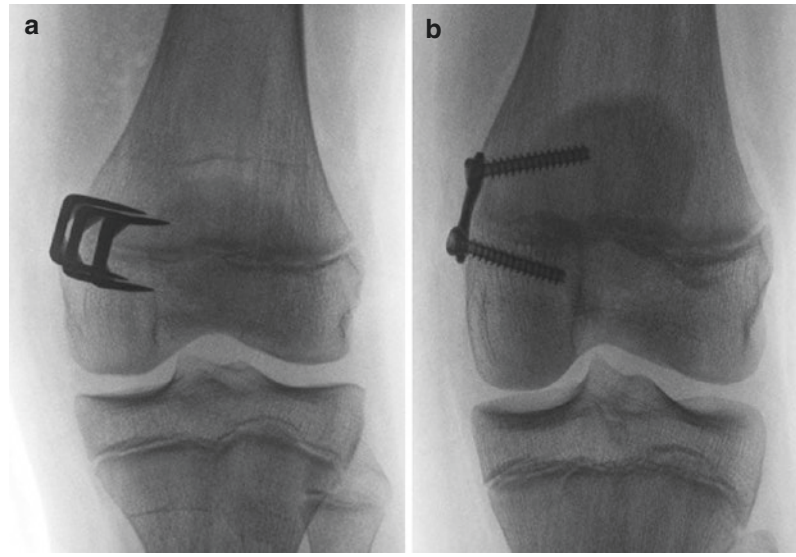



**Fig. 10.24** (**a**) Temporary hemi-epiphysiodesis on the medial side of the distal femoral growth plate of the left knee using clamps, or (**b**) using an eight-plate (*left knee*)

misalignment. However, the chances of recurrence are great even after a wedge excision.

**Infected or fractured growth plate** Operative removal may be considered if the bony bridge covers less than one-third of the total surface of the growth plate. Once the bony bridge is removed, the abnormal position of the knee will be spontaneously corrected during growth, because the growth plates try to direct themselves towards the weight-bearing lines of the leg (Hueter-Volkmann law, 1862). Sometimes it isn't possible to remove the bony bridge, or because the bone bridge comprises more than one-third of the total growth plate. In such cases it is advisable to destroy the rest of the growth plate and perform a correction osteotomy. A difference in leg length should be subsequently corrected at an appropriate time.

**Fractured proximal tibial metaphysis** A wait-and-see policy is adopted for an entire year in the case of a genu valgum caused by a fracture on the medial side in the proximal part of the tibia just distal to the growth plate. A valgus position in the proximal part of the tibia can spontaneous be corrected by the distal tibial growth plate, because it starts directing itself perpendicular to the weight-bearing line (Hueter-Volkmann law, 1862). An S-shaped tibia develops which can only be observed on X-rays (Fig. 10.23). The leg is straight on physical assessment. If this spontaneous correction does not occur, a temporary hemi-epiphysiodesis can be performed by placing an eightplate, clamps or a screw on the medial side of the proximal tibial growth plate in older children Sometimes a decision may be made to do a correction osteotomy, preferably at the end of the growth period. One should realize that in young children there is high risk of recurrence.

## Overstretched Knee Deformity

Complaint: the knee or knees are overstretched.

Assessment: one or both lower legs are in hyperextension with respect to the upper leg(s).

Differential diagnosis:

nonstructural knee hyperextension
- **postural knee hyperextension**
- **hypermobility**

structural knee hyperextension
- **congenital knee hyperextension**
- **congenital knee subluxation**
- **congenital knee dislocation**

tibial tuberosity abnormality
- **acquired recurvatum deformity of the proximal part of the tibia**

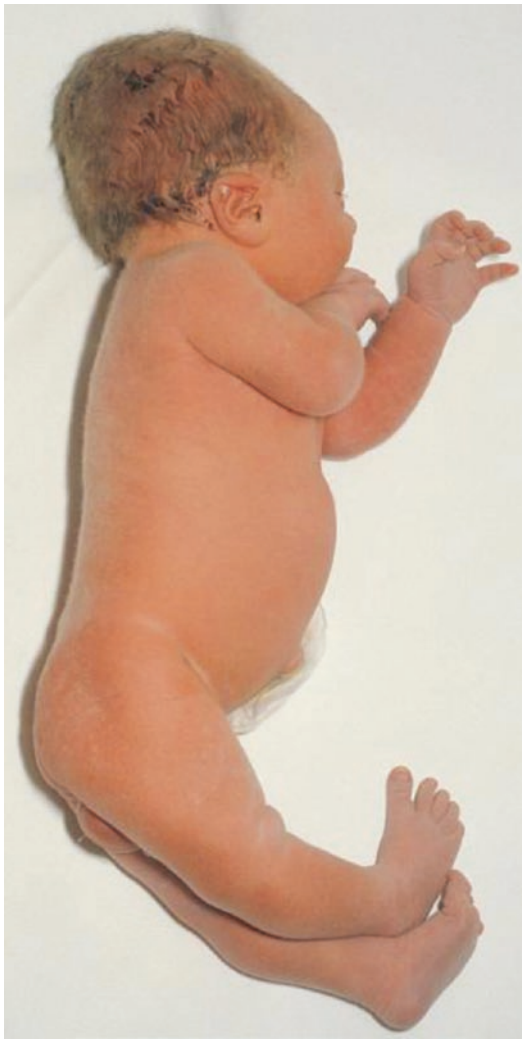


**Fig. 10.25** After a breech presentation the knees may show a hyperextension of 20° (also watch out for developmental dysplasia of one or both hips)

Explanatory note: nonstructural hyperextension. The knee can be fully flexed passively in nonstructural hyperextension

**Postural knee hyperextension** The knees of normal babies have a flexion deformity of 10°–20° up to the age of 2–3 months. The knees may display a hyperextension of 20° after a breech birth (Fig. 10.25). They can be fully flexed on passive assessment. The postural anomaly corrects spontaneously within 3 months. After a breech birth one should also check for developmental dysplasia of the hips.

**Hypermobility** There may also be knees that can hyperextend 10° or more if there is general hypermobility of the joints resulting from joint laxity (Figs. 10.26 and 10.27). The knees can in this case be fully flexed. One can see the hyperextension of the knee when these children stand and walk. Pronounced hypermobile joints are a feature of Ehlers-Danlos syndrome[4].

*Structural hyperextension* In structural hyperextension the knees cannot be completely flexed actively or passively. It can be classified as congenital hyperextension, subluxation or dislocation if the anomaly is present from birth (Fig. 10.28).

**Congenital knee hyperextension** Hyperextension of 15°–20°. The knee can be brought passively into 45°–90° of flexion.

**Congenital knee subluxation** Hyperextension of 25–45°. The knee can be brought passively into a neutral (0°) position.

**Congenital knee dislocation** Hyperextension position of 25–45°. Passive flexion is impossible. A congenital knee dislocation is seen in 1 in 100,000 births. There is a bilateral dislocation in one-third of cases. It is three times more common in girls than in boys. There is also a developmental dysplasia of one or both hips in 70% of children and in 50% of these children they additionally have congenital foot deformities. In a congenital subluxation/dislocation of the knee the proximal part of the tibia is dislocated anteriorly. The term dislocation specifies that the tibial joint surface is not in contact with the femur at all. The term subluxation is used when the proximal tibial joint surface is in partial contact with the distal femur joint surface. There can also be a lateral translation of the tibia with respect to the femur, as well as a valgus position, as a result of a contracture of the iliotibial tract. Semitendinosus and semimembranosus muscles on the medial side of the collateral ligaments may be displaced anteriorly. The quadriceps muscle is shortened, and the suprapatellar recess is fixed onto the femur. The cruciate ligaments may be absent. A congenital knee

[4] See Appendix.

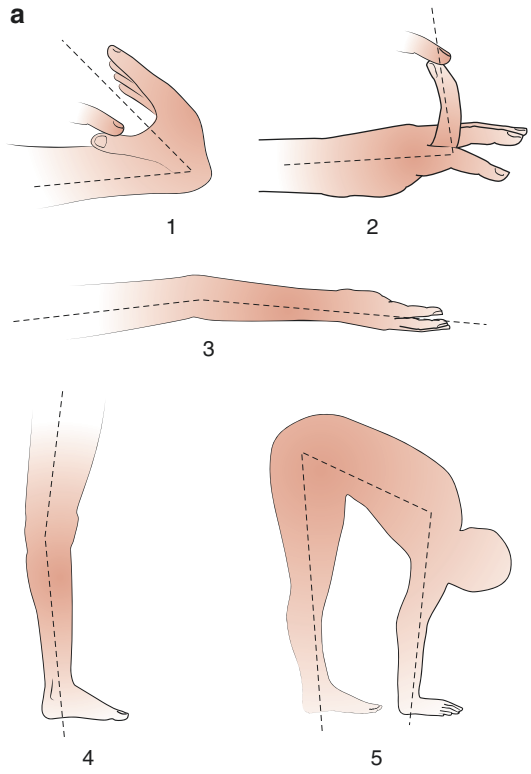



**Fig. 10.26** (**a**) In practice, frequent use is made of the Beighton criteria for hypermobility: *1* passive opposition of the thumb against the anterior side of the forearm. *2* passive hyperextension in the metacarpophalangeal joint of more than 90°. *3* hyperextension in the elbows of more than 10°. *4* hyperextension in the knees of more than 10°. *5* the ability to place one's hand flat on the ground when bending the trunk with extended knees. While testing, the examiner looks at both sides of the body, and one point is allocated per test item, with a maximum of nine attainable points. Hypermobility is present with a score of four points or more. (**b**) Passive position of the thumb against the anterior side of the forearm

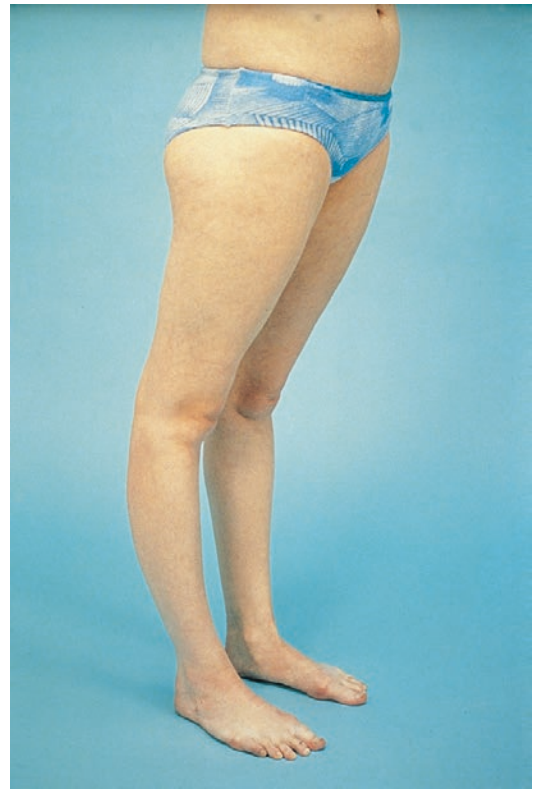


**Fig. 10.27** A 12-year-old girl. The knees can be usually hyperextended if there is hypermobility as a result of congenital ligamental laxity

subluxation/dislocation may be part of the Larsen syndrome[5] (Fig. 10.28) and arthrogryposis multiplex congenita[5]. In Larsen syndrome there may be multiple dislocations of hips, knees and elbows. These children have characteristic facial features, such as a prominent forehead, widely spaced eyes (hypertelorism), and a depressed nasal bridge.

*Tibial tuberosity abnormality* The knee can be fully flexed and the anomaly is not in the joint but just distal to it.

**Acquired recurvatum deformity of the proximal part of the tibia** A premature closure of the anterior side of the proximal tibial growth plate may occur in rare cases, after infection or a fracture of the tibial tuberosity. A recurvation deformity develops due to damage to the anterior side of this growth plate because the apophysis is in continuity

[5]See Appendix.

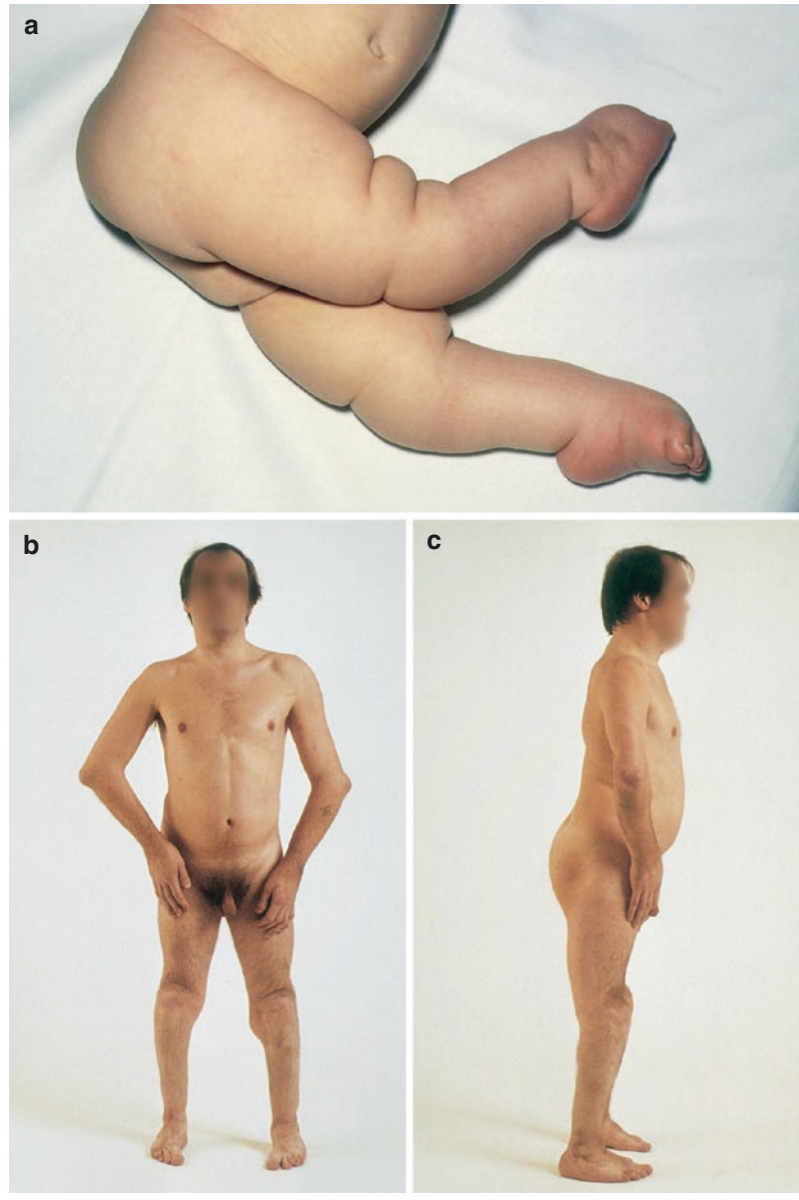



**Fig. 10.28** (**a**) Two-month-old baby with Larsen syndrome, with bilateral knee dislocations and clubfeet. (**b**, **c**) Adult with Larsen syndrome, with untreated bilateral knee and elbow dislocations

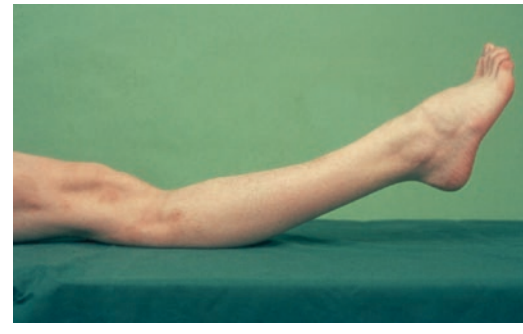


**Fig. 10.29** Recurvatum deformity of the proximal part of the tibia resulting from premature closure of the apophysis of the tibial tuberosity

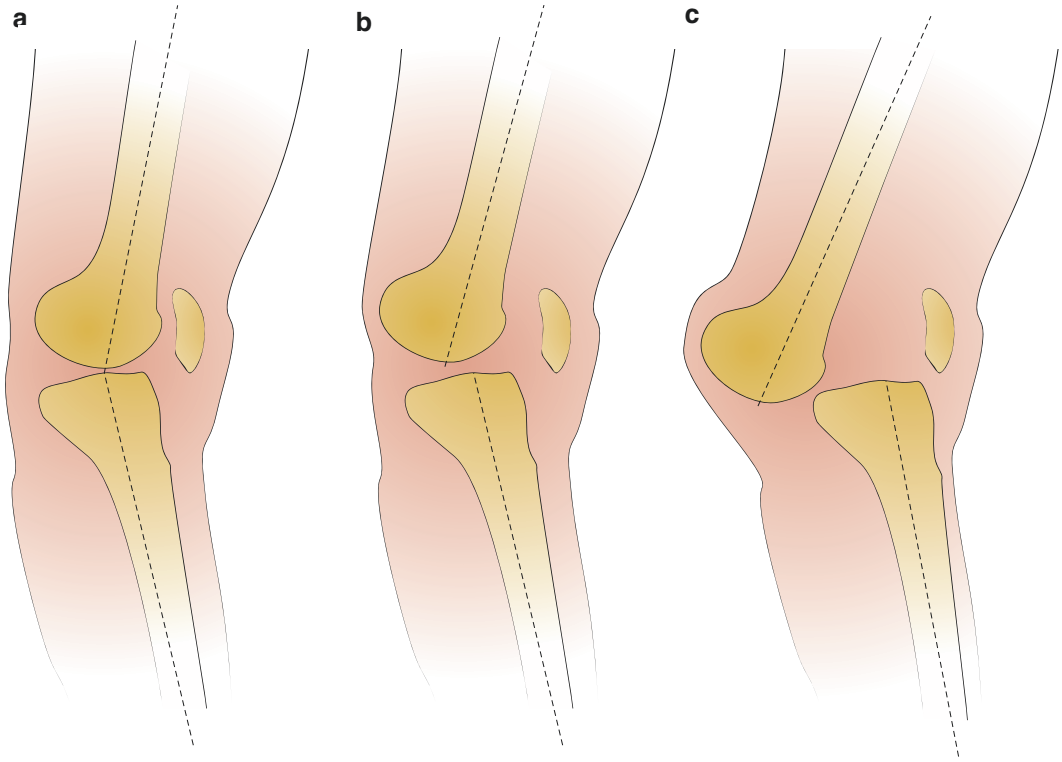



**Fig. 10.30** (**a**) Congenital hyperextension of the knee. (**b**) Congenital subluxation of the knee. (**c**) Congenital dislocation of the knee. In a subluxation/dislocation the proximal part of the tibia is shifted anteriorly (Redrawn from: Curtis BH, Fisher RL. Congenital hyperextension with anterior subluxation of the knee. Surgical treatment and long-term observations. J Bone Joint Surg Am. 1969;51-A:255–69)

with the proximal tibial growth plate, (Fig. 10.29). In extremely rare cases this can also be caused by Osgood-Schlatter disease, which is an apophysitis of the tibial tuberosity (see pp. 231–234).

Supplementary assessment: It is recommended to take lateral X-rays of the knee in newborns if there is hyperextension of 10° or more and if the ipsilateral knee flexion is less than 90°. The X-rays will show an anteriorly displacement of the tibia with respect to the femur if there is a subluxation or dislocation, (Figs. 10.30 and 10.31). X-rays of the knees and CT-scans should be also taken in older children with no previous genu recurvatum but who have developed it in the course of time after a trauma, infection or Osgood-Schlatter disease.

Primary care treatment: children with congenital ligamental laxity and hyperextended knees sometimes complain of pain in the posterior side of the knees. The symptoms usually disappear after introducing a heel elevation under the shoes. As the child grows older it becomes stiffer and the ligamental laxity as well as the hyperextension will tend to disappear.

When to refer: in cases of congenital hyperextension and congenital subluxation/dislocation of the knee in newborns, and when a genu recurvatum deformity occurs at an older age.

Secondary care treatment: **congenital knee hyperextension, congenital knee subluxation and mild congenital knee dislocation.** The practitioner holds the upper leg with one hand and the lower leg with the other. The knee is brought into flexion under traction. Simultaneously, the distal part of the femur is pushed anteriorly. Next, an

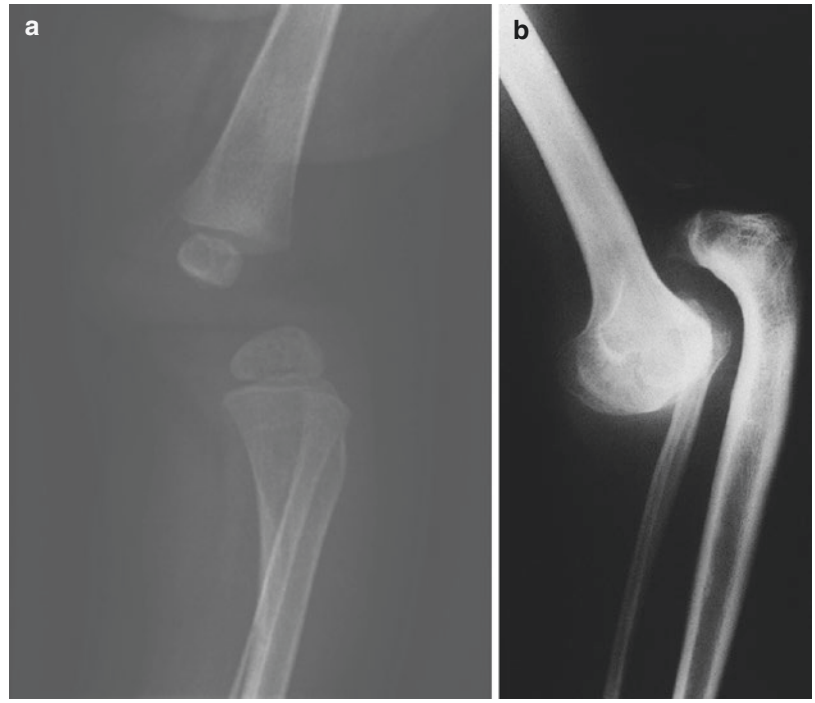



**Fig. 10.31** (**a**) Lateral X-ray of one of the knees of the child from Fig. 10.28a. The tibia is shifted anteriorly with respect to the femur. (**b**) Lateral X-ray of one of the knees of the adult in Fig. 10.28b, c

upper leg cast is applied with the knee in the reduced position. This is repeated until a flexion position of 60° is obtained, after which the knee is treated with a Pavlik harness for 2–3 months (Fig. 9.20).

**Severe congenital knee dislocation** In this case the quadriceps tendon is lengthened in a v-shape, the capsule is divided on the anterior side, and the tensor fascia lata is divided transversely to correct the valgus position (Fig. 10.32). The knee is reduced and immobilized in 30° of flexion for 4–6 weeks in a plaster of Paris cast. After which, a long leg orthosis is worn for an entire year in order to prevent hyperextension.

**Acquired recurvatum deformity of the proximal part of the tibia** A proximal tibial osteotomy is recommended at the end of growth in cases with a recurvation deformity of the proximal part of the tibia. Note that the tibia is usually angled at about 9° posteriorally.

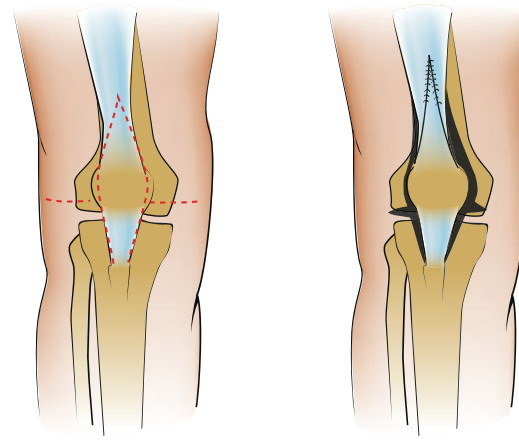


**Fig. 10.32** Operative treatment of a congenital knee dislocation. The quadriceps tendon is lengthened in a V-shape, the capsule is transversely divided on the anterior side and the tensor fascia lata is released to correct the valgus position (Redrawn from: Curtis BH, Fisher RL. Congenital hyperextension with anterior subluxation of the knee. Surgical treatment and longterm observations. J Bone Joint Surg Am. 1969;51-A:255–69)

## Bent Knee Deformity

Complaint: the baby keeps the knee flexed, and it cannot be extended passively.

Assessment: the knee is flexed and is in valgus, the lower leg is in external rotation, or the knee is only flexed.

Differential diagnosis:

**congenital patellar dislocation**
**congenital absent patella**

Explanatory note: **congenital patellar dislocation.** A congenital dislocation of the patellofemoral joint is rare. The dislocation is present at birth but is often diagnosed only at an older age. The patella is laterally dislocated and cannot be reduced manually. The patella is underdeveloped and there is no femoral groove. This anomaly can be unilateral or bilateral, as well as familial. The knee is flexed and is in valgus and the lower leg is in external torsion.

The flexed knee is not neatly rounded but flat, and the femoral condyles are prominent (Fig. 10.33). The patella is not at the front on palpation. The flexion contracture and the valgus position of the knee increase as the child grows. The patella does not ossify before the age of 3, which is why the dislocation isn't visible on X-rays before that time.

**Congenital absent patella** This is a rare condition, one of 10,000 live birth's, that tends to be bilateral and often occurs in combination with other congenital anomalies such as developmental dysplasia of the hip, clubfoot and nail-patella syndrome (onycho-osteodysplasia)[6]. The extension mechanism of the knee remains intact and the knee can be extended actively if the quadriceps femoris muscle is normal. An active extension deficit in the knee will be present if there is reduced quadriceps muscle strength in addition to an absent patella.

Supplementary assessment: X-rays of the knee before the age of 3 are not necessary because the patella is not yet ossified. Anteroposterior as well as lateral X-rays of the knee as well as an additional X-ray of the patellofemoral joint can be requested after the age of 3. The patella is always dislocated laterally in congenital patellar dislocations (Fig. 10.33).

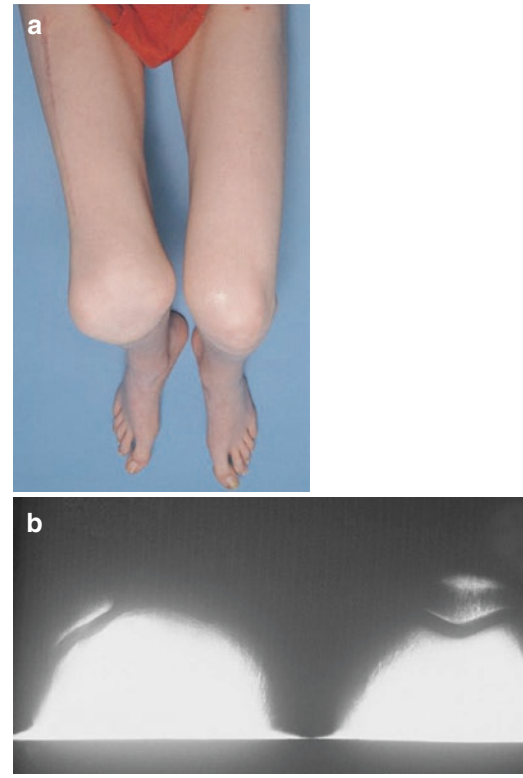



**Fig. 10.33** (**a**) Congenital dislocation of the right patella. (**b**) Axial X-ray of both patellofemoral joints. The right patella is laterally dislocated. An absent femoral groove on the right side is noticeable

Primary care treatment: none.

When to refer: if a flexion contracture of the knee is present 3 months after birth.

Secondary care treatment: **congenital patellar dislocation.** Galeazzi procedure: open repositioning of the patella. A tenodesis of the patella is performed so that it will no longer deviate laterally using the tendon of the semitendinous muscle. The lateral retinaculum is divided lengthwise. The insertion of the vastus

[6] See Appendix.

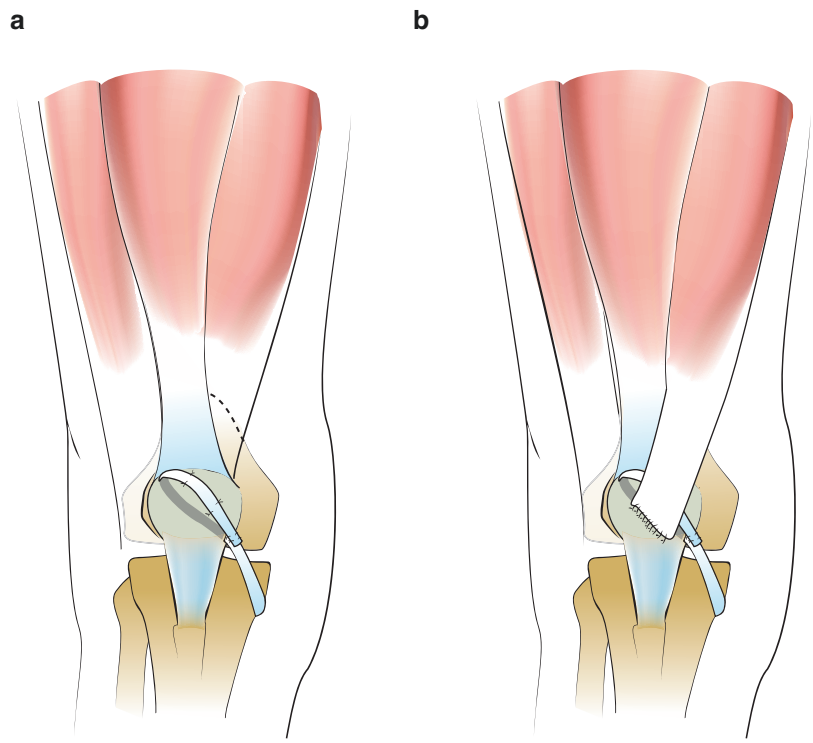



**Fig. 10.34** (**a**) Galeazzi procedure for congenital patellar dislocation. A tenodesis of the patella is performed using the tendon of the semitendinous muscle. (**b**) Modification of this procedure: the lateral retinaculum is divided lengthwise. The insertion of the vastus medialis muscle is released, and fixed laterally and distally onto the patella. The medial capsule and the medial retinaculum are reefed and attached

medialis of the quadriceps femoris muscle is released and repositioned laterally and distally onto the patella, the medial capsule and the medial retinaculum are reefed (Fig. 10.34). Roux-Goldthwait procedure: the lateral half of the patellar ligament is also transposed medially (Fig. 10.35) if there is a Q-angle of more than 20°. Occasionally the quadriceps tendon and/or the hamstrings have to be lengthened.

**Congenital absent patella** Part of the hamstring insertion can be attached to the quadriceps muscle to benefit extension if there is weakness of the quadriceps femoris muscle accompanied by an active extension deficit of the knee.

## Repeated Kneecap Subluxation/ Dislocation

Complaint: symptoms vary widely. Sometimes the only symptom that the patient has is that the kneecap is regularly displaced laterally when flexing and veers back in extension. A repeated subluxation of the kneecap is usual in this case. In the case of a full dislocation the knee will be very painful and the patient will usually fall. The knee is held in flexion. The pain disappears after the knee is passively extended as the kneecap springs back into position.

Assessment: a dislocated patella is palpable on the lateral side of the knee. However, we usually have a situation in which the patella has returned to its normal position. To get an impression about the instability of the patellofemoral joint the knee is flexed slightly by the examiner and the patient is asked to relax the quadriceps muscle. The examiner can then cause abnormal lateralization by pressing the thumbs against the medial side of the patella.

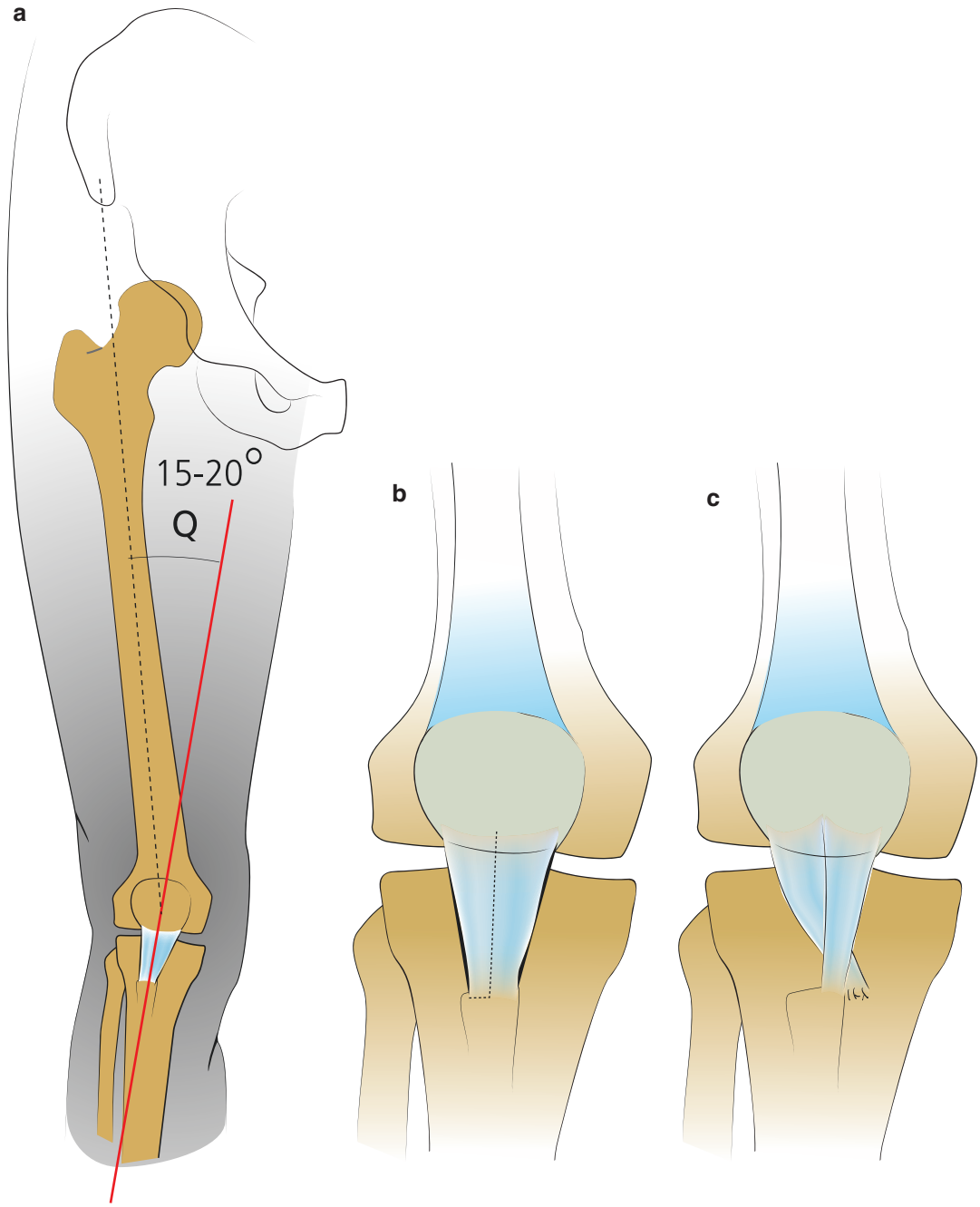



**Fig. 10.35** (**a**) Q-angle. This is the angle between the upper leg and the patellar ligament. The patellar ligament normally deviates 15–20° laterally with respect to the quadriceps. (**b**, **c**) Roux-Goldthwait procedure. The lateral half of the patellar ligament is transposed medially

Differential diagnosis:

**recurrent patellar subluxation/dislocation**
voluntary patellar subluxation/dislocation
habitual patellar subluxation/dislocation
recurrent posttraumatic patellar subluxation/dislocation

Explanatory note: **recurrent patellar subluxation/dislocation.** There are two conditions in which non painful repeated dislocation of the patellar may occur: voluntary patellar subluxation/dislocation and habitual patellar subluxation/dislocation. Recurrent posttraumatic patella subluxation/dislocation occurs after an initial significant trauma and is painful.

*Voluntary patellar subluxation/dislocation* This type is due to hypermobility disorders such as in Down[7] and Ehlers-Danlos syndromes[7].

*Habitual patellar subluxation/dislocation* This a subluxation/dislocation in normal children with no associated ligamentous laxity. In general there are knee anomalies, especially a misalignment of the extensor apparatus. The first dislocation in a habitual patellar subluxation or dislocation is generally not caused by a trauma. The patella dislocates upon knee flexion. The dislocation is a consequence of a developmental disorder with the following possible manifestations: atrophy of the vastus medialis of the quadriceps femoris muscle; a more proximal insertion of the vastus medialis: external torsion, combined with a valgus position of the leg, causing a larger Q-angle (Fig. 10.35); a shallow femoral groove; a proximally localized patella; or combinations of the above listed developmental disorders. In a proximally localized patella (patella alta) the distance between the apex of the patella and the attachment to the tibial tuberosity is greater than the length of the patella. Normally, the distance between the apex of the patella and the tibial tuberosity is as great as the length of the patella. Variation of more than 20 % indicates an abnormal position.

*Recurrent posttraumatic patellar subluxation/dislocation* A traumatic patellar dislocation affects 3–4 in 10,000 children between 10 and 17 years of age. In the first instance there has been an extensive valgus/flexion trauma, causing a laxity in the medial retinaculum that can lead to recurrent dislocations. Recurrences are seen in 30–50 % of cases after an initial traumatic patellar subluxation/dislocation,.

Supplementary assessment: anteroposterior and lateral X-rays of the knee and an axial X-ray of the patellofemoral joint. In a recurrent posttraumatic subluxation and dislocation there are often calcifications at the level of the medial retinaculum. Such calcifications are caused by a traumatic tear in the medial retinaculum. Sometimes there is an avulsion fracture on the medial part of the patella. There is an osteochondral fracture of the patellofemoral joint in 40 % of cases.

Primary care treatment: in the case of patellar subluxation/dislocation you should encourage the patient to extend the knee. The patella will tend to reduce spontaneously. If this is not the case, analgesics and muscle relaxants can be given. After which the patient can again be encouraged to extend the knee while the examiner pushes the patella from lateral to medial. A repetetive subluxation is best treated conservatively. Children with no surgical treatment after repetetive subluxations had less pain and osteoarthritis and fewer repetetive subluxations than did those who had proximal or distal realignment procedures. In patients treated conservatively the number of subluxations decreased dramatically as they approached 30 years of age.

When to refer: as a rule, after at least three subluxations/dislocations. A natural healing tendency is minimal after that. Dislocations of the patella will continue if no treatment is carried out.

Secondary care treatment: **voluntary patellar subluxation/dislocation.** In the first instance one should try quadriceps exercises and a patellar anti-dislocation orthosis. An operation should be avoided as much as possible. In problem cases in children a

[7] See Appendix.

Galeazzi procedure can be performed. After the child stops growing heighten the lateral part the patellofemoral groove by means of an osteotomy.

**Habitual patellar subluxation/dislocation** The growth plates will be damaged in a child if one carries out an operation on bone. For this reason, soft-tissue surgery should be chosen with a Galeazzi or modified Galeazzi operation for a proximal realignment (Fig. 10.34) possibly combined with a Roux-Goldthwait procedure for distal realignment (Fig. 10.35). However, despite these interventions patellar subluxations/dislocations may keep occurring. In that case, a bony operation can be considered after the child stops growing.

The tuberosity is displaced medially in the case of a Q-angle larger than 20°. Combining a medial with distal transposition (Hauser procedure) may only be carried out if there is a proximally localized position of the patella (patella alta) (Fig. 10.36). If this is not the case, distal transposition of the tibial tuberosity will cause premature degenerative changes in the patellofemoral joint. However, even when the tibial tuberosity is only displaced medially, it is in fact also displaced posteriorly, which can cause patellofemoral problems. An alternative is the Elmslie-Trillat procedure, in which a segment of the cortex of the tibial tuberosity is rotated medially. Early degenerative patellofemoral joints changes developed in those with distal realignment. Those with proximal realignment had fewer patellofemoral changes, but the recurrence was 25 %. A correction osteotomy proximal to the tibial tuberosity may be carried out if the excessive Q-angle is caused by a valgus position of the knee or by internal torsion of the lower leg. In the case of a shallow femoral groove, the lateral part of the groove can be raised anteriorly by means of an osteotomy.

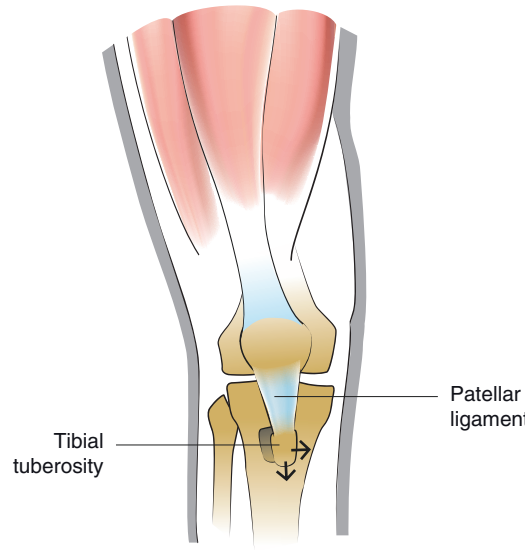



**Fig. 10.36** Hauser procedure: medial and distal displacement of the tibial tuberosity. A combination of medial and distal displacement may only take place if the patella has a proximal position (patella alta). Only a medial displacement of the tibial tuberosity is performed if there is no elevated patella position (Redrawn from: Hauser EDW. Total tendon transplant for slipping patella. A new operation for recurrent dislocation of the patella. Surg Gynecol Obstet. 1938;199–214)

**Recurrent posttraumatic patellar subluxation/dislocation** An arthroscopy after the first dislocation should first be considered if osteochondral fractures are suspected. Small fragments can be removed, and large fragments can possibly be reduced and fixed. The knee is immobilized for 5–10 days in a plaster of Paris cast or a patellar anti-dislocation orthosis if there is no osteochondral fracture. This is followed by prescribing quadriceps and hamstring exercises. A release of the lateral retinaculum can be carried out and the medial capsule can be reefed and sutured if recurrent subluxations/dislocations occur after this. This treatment is based on the situation after at least three dislocations.

**Differential Diagnosis Knee Misalignment**

| | |
|---|---|
| **Bowleg** | |
| Bilateral | |
| Up to the age of 2 years. | **Physiological genua vara** |
| After the age of 2 years without skeletal abnormalities (with a tibiofemoral varus >5°). | **Idiopathic excessive genua vara** |
| Cranial thinning, broad wrists. | **Rickets (infantile osteomalacia)** |
| Short body length. | **Skeletal dysplasia** |
| Bilateral or unilateral genu varum | **Blount disease (tibia vara, osteochondrosis deformans tibiae)** |
| After the age of 2 years with skeletal abnormalities. | Infantile type |
| Metadiaphyseal angle >16°. | |
| After the age of 8 years with skeletal abnormalities. | Adolescent type |
| Unilateral | |
| Bone defect on the medial side in the proximal part of the tibia. | **Focal fibrocartilaginous dysplasia** |
| Swelling on the lateral side of the knee. | **Dysplasia epiphysealis hemimelica (Trevor disease)** |
| Caused by infection or fracture. | **Infected or fractured growth plate** |
| **Knock knee** | |
| Bilateral | |
| Between ages 2 and 7. | **Physiological genua valga** |
| After the age of 7 years without skeletal abnormalities with a tibiofemoral valgus angle >10°. | **Idiopathic excessive genua valga** |
| Cranial thinning, broad wrists. | **Rickets (infantile osteomalacia)** |
| Short body length. | **Skeletal dysplasia** |
| Swellings around the knee. | **Multiple osteochondromata (exostoses)** |
| Unilateral | |
| Swelling on the medial side of the knee. | **Dysplasia epiphysealis hemimelica (Trevor disease)** |
| Caused by infection or fracture. | **Infected or fractured growth plate** |
| After a fracture in the proximal part of the tibia just below the growth plate. | **Fractured proximal tibial metaphysis** |
| **Overstretched knee deformity** | |
| Newborn after a breech birth full flexion is possible. | **Postural knee hyperextension** |
| Congenital ligamental laxity. | **Hypermobility** |
| Hyperextension 10°, passive flexion up to 45–90°. | **Congenital knee hyperextension** |
| Hyperextension 25–45°, passive flexion up to a neutral (0°) position. | **Congenital knee subluxation** |
| Hyperextension 25–45°, passive flexion is impossible. | **Congenital knee dislocation** |
| After fracture or infection of the tibial tuberosity or in Osgood-Schlatter disease. | **Acquired recurvatum deformity of the proximal part of the tibia** |
| **Bent knee deformity** | |
| Flexed, valgus position and external rotation of the knee. | **Congenital patellar dislocation** |
| Flexion contracture due to reduced quadriceps strength. | **Congenital absent patella** |
| **Repeated kneecap subluxation/dislocation** | |
| No preceding an extensive trauma. | **Habitual patellar subluxation/dislocation** |
| First dislocation due to an extensive trauma. | **Recurrent posttraumatic patellar subluxation/dislocation** |

## Knee Swelling

### Swelling Just Above and/or Below the Knee

Complaint: a swelling just above and/or below the knee.

Assessment: hard bony swelling.

Differential diagnosis:

**benign tumor**
**malignant tumor**

Explanatory note: **benign tumor.** The most common bone tumor at the level of the knee joint, which is accompanied by swelling, is an osteochondroma, This is also known as exostosis/osteocartilaginous exostosis (Fig. 10.37). This is a developmental disorder in the periphery of the growth plates in which a bony outgrowth can develop that usually has a cartilaginous cap. This irregularity can be observed as a hard, painless swelling, with symptoms arising due to friction caused by the displacement of muscles and tendons over the swelling. The anomaly increases in size until the child stops growing. An osteochondroma at the level of the head of the fibula can cause knock knees (Figs. 10.20 and 10.21). An osteochondroma located in the distal part of the lower leg between the tibia and the fibula can cause a valgus misalignment of the ankle.

The anomaly can occur in a solitary form (solitary osteochondroma) or in many areas, identified as multiple osteochondromata or exostoses and abbreviated as MO or ME. Multiple osteochondromata are hereditary in more than 60 % of cases. These are referred to as hereditary multiple osteochondromata or exostoses and are abbreviated as HMO or HME.

The familial form affects about 10–25 in 1,000,000 individuals. There is a 50 % chance that one of the children will get it if one of the patient's parents has the disease. Girls and boys are at equal risk. The number of osteochondromata varies per individual, but tends to hover between 10 and 20. The anomalies can develop in any bony part, but usually appear in the distal part of the femur, proximal and distal part of tibia and fibula. Localizations in the ribs, pelvis, the shoulder blade, proximal part of the humerus, forearm (usually the wrist), proximal part of the femur are also possible. The most frequent localization is around the knee.

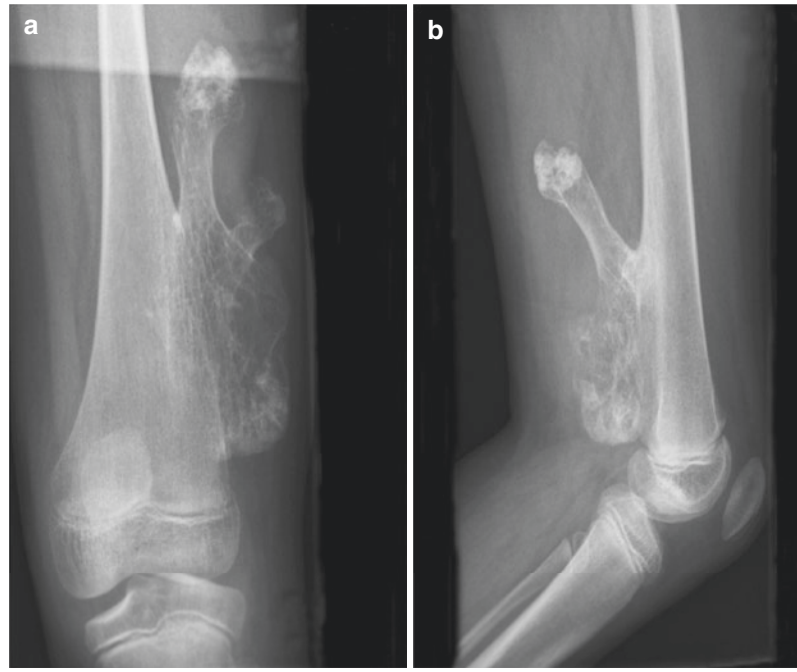



**Fig. 10.37** Osteochondroma (exostosis) of the femur

Osteochondromata can be detected at any age, but are usually discovered in infants and toddlers. In 50 % of children osteochondromata have been discovered by the age of 3½ years, and in nearly all children by the age of 12. It is very unlikely that osteochondromata will develop in a 12-year-old child who has no osteochondromata but does have a family history with osteochondromata.

Malignant transformation is almost nonexistent in youths, but they do happen in adulthood. The chances of this is less than 1 % for a solitary osteochondroma and 1–2 % in multiple osteochondromata. Malignant transformation is mostly associated with osteochondromata localized in the pelvis and in the scapulas. Malignancy should be suspected if an osteochondroma increases in size during adulthood.

**Malignant bone tumor** There are few symptoms in the beginning. Vague, intermittent pain, especially after sports activities may be the first symptom. Gradually the pain is present all day, and also at night. In general, only at a later stage is there a visible swelling that increases in size.

For explanatory note, supplementary assessment, primary care treatment, when to refer and secondary care treatment, see for an osteochondroma pp. 182–185 and for a malignant bone tumor pp. 223–227.

## Swelling at the Back of the Knee

Complaint: swelling at the back of the knee.

Assessment: swelling at the back of the knee (Fig. 10.38).

Differential diagnosis:

**popliteal cyst (Baker cyst)**
**soft tissue tumor**

Explanatory note: a nonpainful swollen bursa of the semi-membranous and/or medial gastrocnemius muscle can develop in the popliteal space between the third and the eighth years of life. It can be more easily felt with an extended knee with the patient lying prone than with a flexed knee. A cyst in children tends not to communicate with the joint, and in contrast to adults it is not related to any intra-articular irregularities. These bursae tend to disappear spontaneously after a few years.

Supplementary assessment: if there is doubt regarding the diagnosis, a lamp can be held against the swelling. We are dealing with a popliteal cyst if the swelling lets light through (transillumination). There may be a soft tissue tumor in the popliteal space if this is not the case. Softtissue tumors in the popliteal space are rare in children.

Primary care treatment: a wait-and-see policy can generally be adopted. Reassure the parents that we are dealing with a benign anomaly. If you cannot convince them, you can confirm the diagnosis by aspirating the cyst. You should however tell the parents that this procedure is intended to confirm the diagnosis after which the cyst will return.

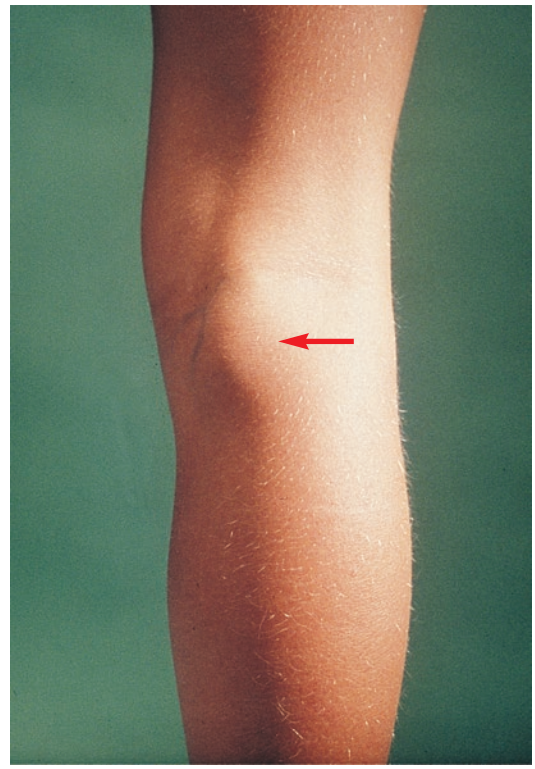


**Fig. 10.38** Popliteal cyst (*arrow*)

When to refer: only when you have doubts about the diagnosis.

Secondary care treatment: **popliteal cyst.** In case of doubt about the diagnosis, an ultrasound and if necessary a MRI should be carried out. Operative removal of a popliteal cyst is rarely indicated. Only in exceptional circumstances should one consider operative removal in the case of a large painful cyst. Risk of recurrence is very high.

*Treatment in a specialized center* **soft tissue tumor.** These tumors must be treated in a specialized center.

## Knee Movement Limitations

### Knee Stretch Limitation

Complaint: pain laterally or medially. The knee can no longer be fully stretched.

Assessment: the knee cannot be fully extended, actively or passively (lock phenomenon). Forced passive extension causes pain around the medial or lateral joint gaps. Sometimes there is hydrops. In a maximally flexed knee there is pressure pain on the anterior side of the medial or lateral femoral condyles (König pressure point). Meniscal tear tests are also performed (see pp. 203–204).

Differential diagnosis:

**osteochondritis dissecans of the femoral condyle**
**discoid meniscus**
**meniscal tear**

Explanatory note: **osteochondritis dissecans of the femoral condyle.** Radiological studies in symptomless children younger than 10 years of age frequently show an irregular ossification of the lateral femoral condyle. There is never a loose fragment. This radiological variant disappears spontaneously before the 10th year of life and should not be confused with a subchondral bony necrotic lesion, osteochondritis dissecans. The cause of osteochondritis dissecans is unknown; an overload-caused stress fracture is suspected, which leads to the development of subchondral bony necrosis due to diminished vascularisation. Osteochondritis dissecans occurs mainly in children who participate in intensive sports. Subchondral bony necrotic lesions can occur in multiple joints (elbow, hip, knee, ankle and the first metatarsophalangeal joint).

The knee is most frequently affected. In 85 % of cases the anomaly is localized at the level of the medial femoral condyle, in about 13 % of cases at the level of the lateral femoral condyle, and in a few percent around the patellofemoral joint (Fig. 10.39). Osteochondritis dissecans occurs between the ages of 5 and 15, peaking between ages 11 and 14. In 5 % of cases the anomaly is bilateral. Osteochondritis dissecans affects boys twice as often as girls, occurring particulary between ages of 10 and 20. After a trauma the entire subchondral bony fragment can become loose and becomes a loose body (corpus liberum) (Fig. 10.40).

The most important symptom is pain. At first, symptoms mostly resemble patellofemoral pain syndrome (see pp. 228–229). The pain shifts in stage II to the medial or lateral side of the knee depending on the localization. In stages II and III there may be a permanent or recurrent hydrops. In stages III and IV there may be a limitation in extension caused by the loose fragment between the femoral condyle and the tibial plateau. The limitation in extension sometimes develops also in stage II if the lesion protrudes to some degree but is still lying in the original location. There is in a maximally flexed knee pressure pain on the medial side of the femoral condyle. This is called the König pressure point. If this involves the lateral femoral condyle there is pressure pain on the lateral femoral condyle with natural maximum knee flexion. Osteochondritis dissecans of the lateral femoral condyle is accompanied by a discoid meniscus in 15 % of cases. Stages I and II are the most common. However, with small lesions spontaneous healing occurs between 12 and 18 months in 90 % of cases.

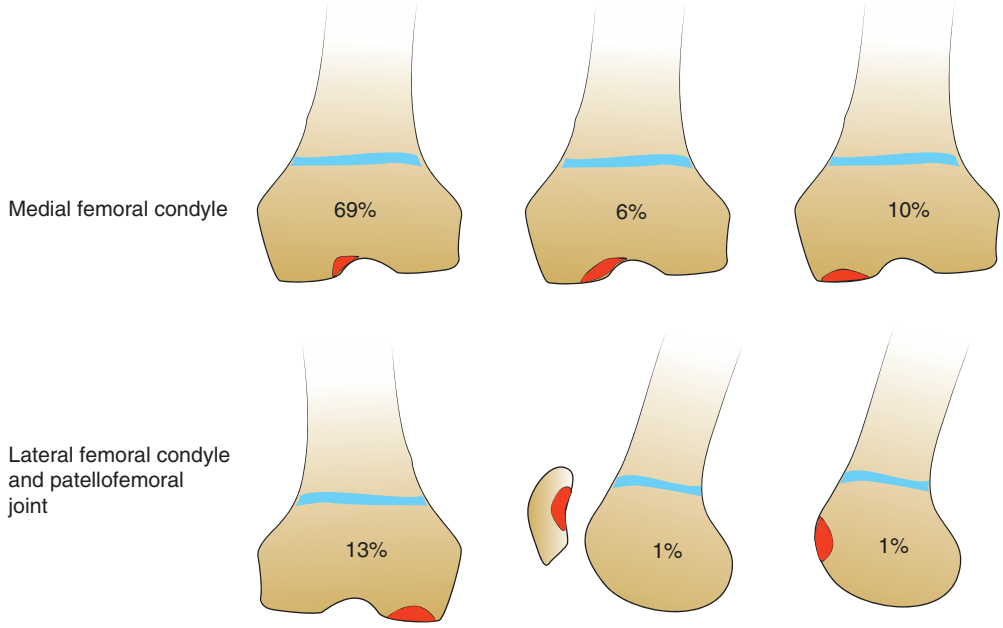



**Fig. 10.39** Localization of an osteochondritis dissecans lesion

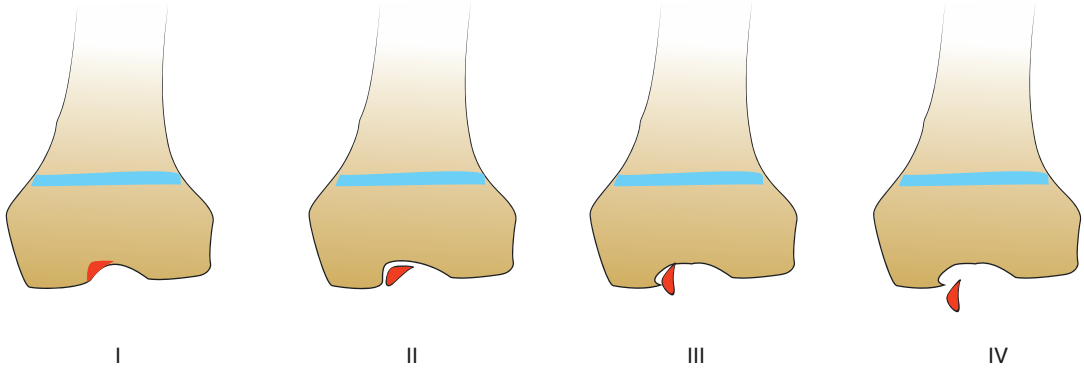



**Fig. 10.40** Classification of osteochondritis dissecans of the left knee. Grade *I*: non-displaced osteochondral fragment. Grade *II*: non-displaced loose osteochondral fragment. Grade *III*: partial displacement of the loose osteochondral fragment. Grade *IV*: fully displaced loose osteochondral fragment (corpus liberum)

**Discoid meniscus** In about 0.5 % of cases a lateral meniscus has a congenitally abnormal shape. Instead of a half moon shaped meniscus, it is shaped as a flat disk (Fig. 10.41). Such an abnormal shape is almost never seen in a medial meniscus. A discoid meniscus rarely leads to symptoms. Complaints can be temporary and usually disappear on their own. There is often a discoid meniscus in the other knee.

There are three types of discoid menisci: complete, incomplete, and a Wrisberg ligament type. A discoid meniscus is thicker than a normal meniscus and encompasses the entire or a great part of the articular surface of the tibia. The complete and incomplete types are normally anchored. The Wrisberg ligament type is anchored only on the posterior side with the Wrisberg meniscofemoral ligament. This makes the meniscus very mobile. It can cause audible snapping when the knee is extended, especially in young children. Due to its increased mobility the meniscus can double up and this can cause locking symptoms. There may be pressure pain around the lateral joint gap.

**Meniscal tear** Meniscal lesions are very rare in children. When they do occur, it is usually at the end of the growth period (older than 12). Medial meniscal tears are three times more common than lateral tears. In children this chiefly involves a vertical lengthwise tear in the posterior part of the medial meniscus which gradually expands ventrally. The torn, centrally located part can become sandwiched between the femoral condyle and the tibial plateau, causing a limitation in extension. The knee is locked. The torn meniscus part can be compared with the handle of a bucket (Fig. 10.42), and that is why it's called a "bucket handle tear".

Tests administered to check for a meniscal tear are the joint line tenderness test (Fig. 10.43), Steinman test (Fig. 10.44), McMurray test (Fig. 10.45), Appley test (Fig. 10.46), and Thessaly test (Fig. 10.47). No single meniscus test is accurate enough to prove a meniscal tear, but there are indications that combining tests leads to higher diagnostic accuracy.

Supplementary assessment: anteroposterior and lateral X-rays as well as a flexed "tunnel" view of both knees in case of limited knee extension. It is often easier to see an osteochondritis dissecans lesion on a "tunnel" X-ray than on an anteroposterior or lateral X-ray (Fig. 10.48). An MRI should be requested if the X-rays are not abnormal and a meniscal tear or a discoid meniscus is suspected.

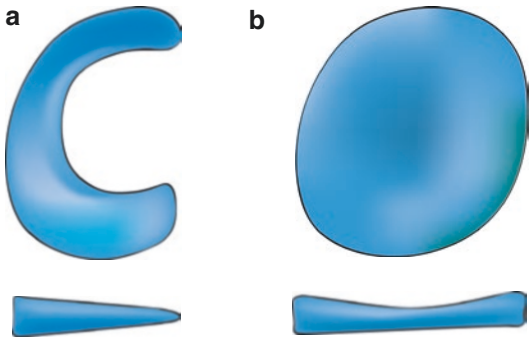



**Fig. 10.41** In about 0.5 % of cases the lateral meniscus has a congenitally abnormal shape. Instead of a half moon shaped piece of cartilage (**a**) we find a disc-shaped meniscus. This is called a discoid meniscus (**b**)

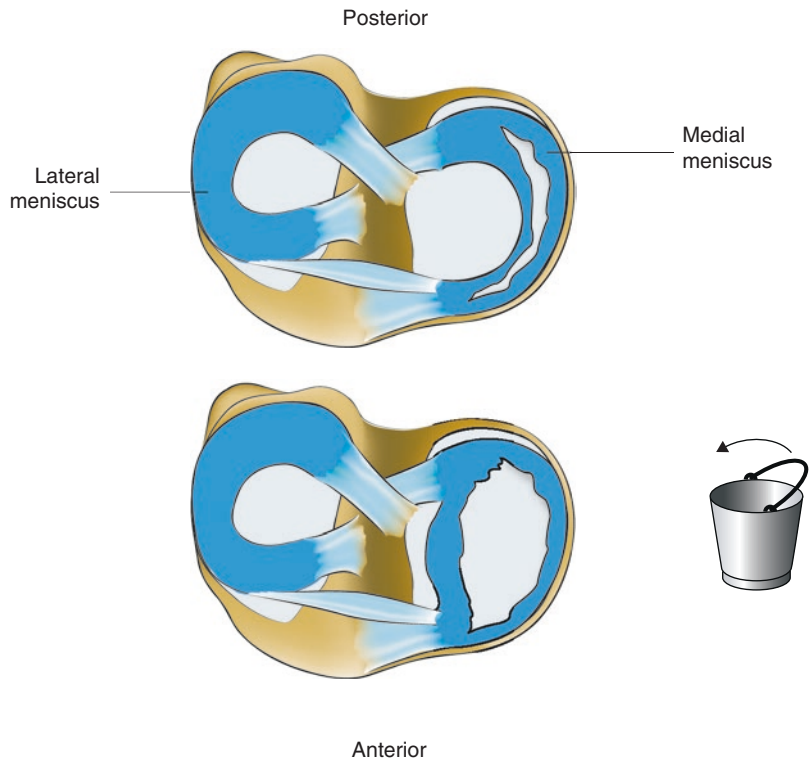



**Fig. 10.42** (**a**) Vertical tear in the medial meniscus of the right knee. (**b**) The torn, centrally located part can dislocate laterally and get sandwiched in between the femoral condyle and the tibial plateau, causing limitation in extension. The torn central part is known as a "bucket handle"

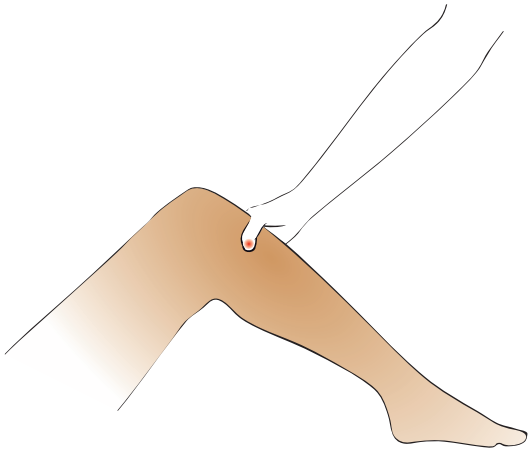


**Fig. 10.43** Joint line tenderness test. The medial and lateral joint lines can be palpated separately with the knee in 90° flexion. The test is positive when the patient cannot tolerate pressure pain at the medial joint line in a medial meniscal tear and at the lateral joint line in a lateral meniscal tear

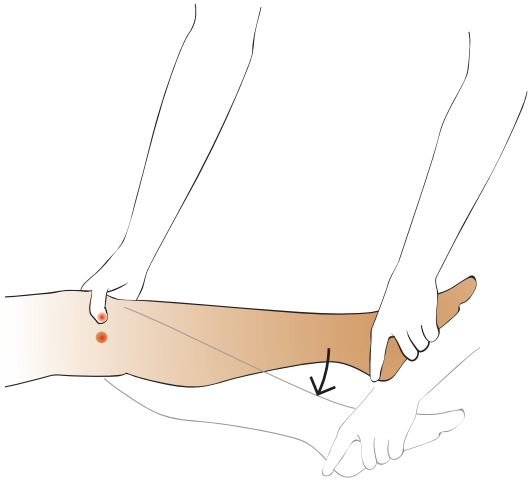


**Fig. 10.44** Steinmann test II. The joint line is palpated. A positive test is indicated if the tenderness moves posteriorly with increasing flexion or anteriorly when the knee is extended

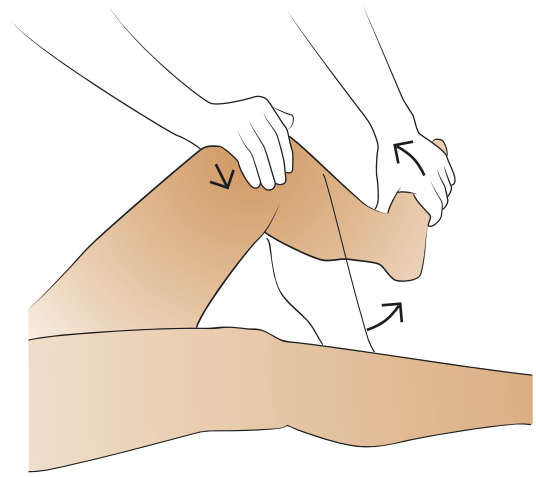


**Fig. 10.45** Mc Murray test. To examine a tear of the posterior part of the medial meniscus the examiner brings with one hand the knee in maximum flexion and external rotation of the lower leg with simultaneously palpating the medial jointline with the other hand. Next the knee is extended. During extension a torn and sandwiched posterior part of the medial meniscus may shoot back into place. At that moment a click may be heard and/or felt. To examine the posterior part of the lateral meniscus the procedure is performed with the lower leg in internal rotation and with lateral joint line palpation

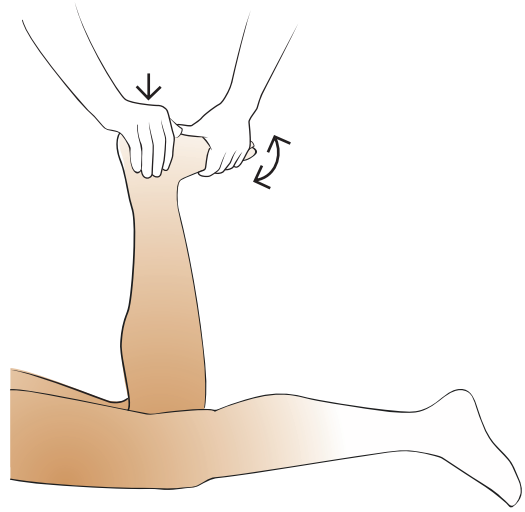


**Fig. 10.46** Appley test. The patient lies prone with the knee bent 90°. The examiner rotates the leg externally and internally several times under simultaneous vertical pressure. A painful snap can point to a meniscal injury

Primary care treatment: the initial advice is to avoid knee-loading sports (such as American football, rugby, soccer, volleyball, basketball, and tennis) in children between the ages of 10 and 15 in the case of an osteochondritis dissecans in which the osteochondral fragment is still fixed (stages I or II). If there are many complaints kneeloading can be relieved by walking with crutches. There are no objections to cycling or swimming. X-rays of the knee should be repeated after 6 months. If the images show improvement, a

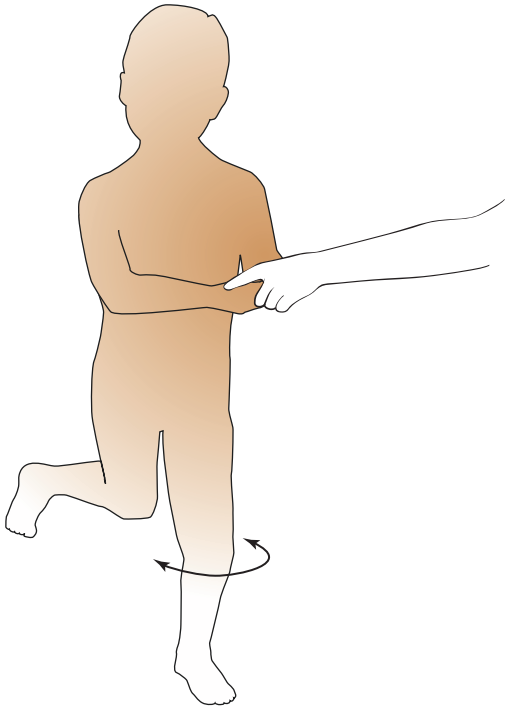


**Fig. 10.47** Thessaly test: the patient stands on the leg to be examined with the knee bent 20° and the foot flat on the ground. The thigh is internally and externally rotated three times by turning the body with respect to the lower leg. The test is considered to be positive if the patient has pain around the medial or lateral joint area. There may also be locking symptoms

wait-and-see policy can be adopted with follow up every 6 months until full healing is achieved (within 18 months).

One should bear in mind the possibility of a meniscal tear or a discoid lateral meniscus if the X-rays show no indication for osteochondritis dissecans.

When to refer: if extension limitation does not disappear spontaneously, if the X-rays show loosening of the osteochondral fragment, or if in children between 10 and 15 years of age the X-rays show no improvement after avoiding knee-loading sports. Patients should be referred who also have persistent complaints from a discoid meniscus or a proven meniscal tear.

Secondary care treatment: **osteochondritis dissecans of the femoral condyle.** The lesion should be fixed with one or several compression screws in a stage II osteochondritis dissecans that shows no improvement or a stage III that shows partial loosening of the fragment (Fig. 10.48). The screws are removed after consolidation. If the corpus liberum hasn't stabilized in stage IV fixation can also be considered, with or without a bone transplant. A corpus liberum will be removed operatively if it is rounded off and cannot be repositioned. The defect in the femoral condyle is curetted and bone marrow stimulated by micro fracture technique. Arthrosis is seen within 20 years in more than 75 % of cases when the fragment cannot be repositioned. One may consider removing autologous bone with cartilage from the non weight bearing part of the knee and placing this in the defect. Recent studies have focused on what is known as biological repair of the cartilage, using cultivated or the body's own chondrocytes. The effects of this possibility are still unclear.

**Discoid meniscus** If this still keeps giving problems, it can be reshaped operatively to create a more or less half moon shaped meniscus. However, this type of procedure can never give a normal shaped meniscus especially because the inner border will always be thicker than in a normal meniscus. Sometimes it is necessary to remove the entire meniscus. The results of a total meniscectomy in children are poor due to premature wear and tear.

**Meniscal tear** An arthroscopy is performed if a meniscal tear is strongly suspected. An arthroscopy can remove the torn portion, leaving the intact part alone. A meniscal tear can be sutured under certain conditions (Table 10.2). Good revascularization around the tear is the most important precondition for the healing of a sutured meniscal tear. A meniscal tear must lie in what is known as the red-red zone, which has good vascularization on both sides of the tear, or in any event in a red-white zone that has a good blood supply to the capsular side of the tear. A tear that is localized less than 2 mm from the joint

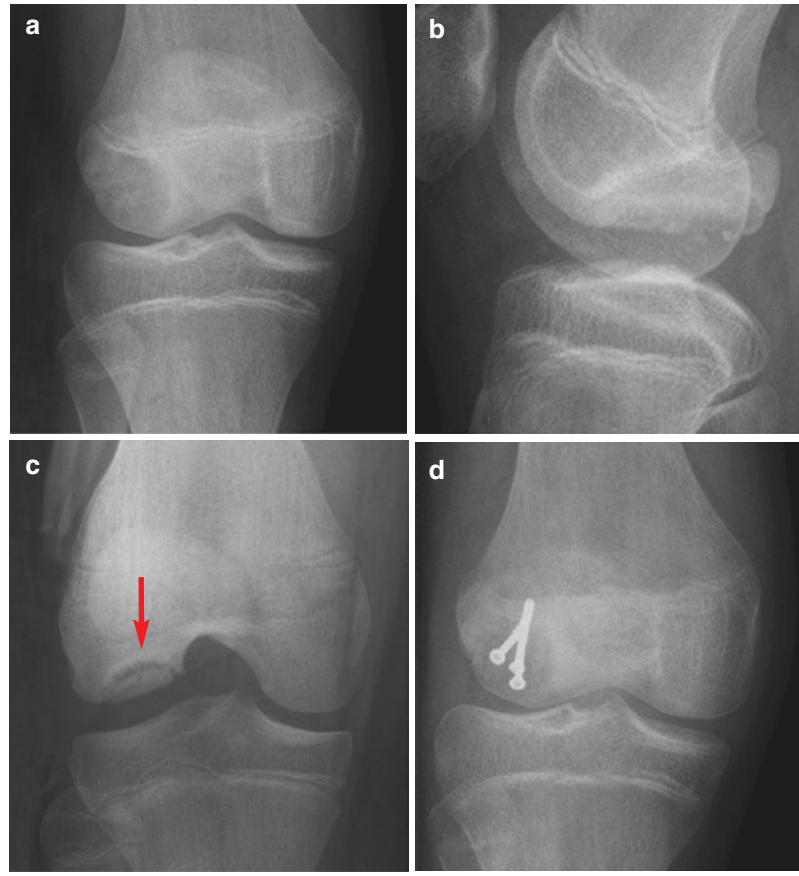



**Fig. 10.48** (**a**) Anteroposterior, (**b**) lateral, and (**c**) tunnel view images of the knee. In contrast to the anteroposterior and lateral images the osteochondritis dissecans lesion can be clearly seen on a tunnel view X-ray (*arrow*). (**d**) Fixing the osteochondral lesion with three screws

**Table 10.2** Preconditions for suturing a meniscal tear

| |
|---|
| Meniscal tear in red-red zone |
| Vertical meniscal tear |
| Meniscal tear less than 4 mm distance from the joint capsule |
| Meniscal tear longer than 1 cm and shorter than 4 cm |
| Patient younger than age 40 |
| Neutral knee-axis |
| Meniscal tear is present for less than 6 weeks |

From Laible C, Stein DA, Kiridly DN. Meniscal repair. J Am Acad Orthop Surg. 2013;21:204–13

capsule has the greatest chance of healing. The chances of healing are minimal if a tear is located more than 4 mm from the joint capsule. Meniscal tears of less than 1 cm are considered stable and will generally heal spontaneously. Tears longer than 4 cm are unstable and heal poorly after suturing. Vertical tears, which tend to affect young patients, heal the best. Radial tears are located mostly in the avascular zone and are not suitable for suturing. Horizontal tears are generally not sutured either because they are difficult to suture and are mostly of a degenerative nature. There must be a neutral axial position in the knee. A distraction will occur at the level of the sutured tear, and the chances of healing are minimal if a medial meniscal tear in a varus knee is sutured. The suturing of meniscal tears within 6 weeks after they appear has a better prognosis than tears that have been present for longer than that. It is assumed that younger patients have greater chances of healing than in older individuals. A sutured meniscal tear has greater chances of successful healing if an anterior cruciate ligament reconstruction is carried out simultaneously. This is probably due to an increased vascularization resulting from the anterior cruciate ligament procedure.

## Knee Bending Limitation

Complaint: the knee cannot be fully flexed.

Assessment: the knee cannot be fully flexed, actively or passively. There are no other locomotor irregularities. The knee can be normally extended.

Diagnosis: **quadriceps fibrosis.**

Explanatory note: in this anomaly there is a progressive extension contracture resulting from a progressive fibrosis of one or several components of the quadriceps femoris muscle. The fibrosis is in the distal part of the quadriceps femoris muscle. The vastus intermedius of the quadriceps femoris muscle is most frequently involved. The fibrosis makes the muscle less elastic and knee flexion becomes difficult. A shortening of the quadriceps can also occur, pulling the patella superiorally (patella alta). The anomaly affects young children, especially girls. The cause is unknown. A congenital solitary muscle abnormality is suspected. In some cases the administration of antibiotics into the quadriceps femoris muscle at a young age may help.

Supplementary assessment: X-rays of the knee can show a patella alta.

Primary care treatment: there is no point in going to a physiotherapist. The contracture of the quadriceps femoris muscle does not react to active or passive exercises.

When to refer: if knee flexion is less than 90°.

Secondary care treatment: **quadriceps fibrosis.** Operative lengthening of the quadriceps tendon if knee flexion is less than 90°.

**Differential Diagnosis: Knee Swelling**

| **Swelling just above and/or below the knee** | |
|---|---|
| Hard bony painless swelling. | **Benign bone tumor** |
| Hard bony painful swelling. | **Malignant bone tumor** |
| **Swelling at the back of the knee** | |
| Fluctuating and translucent swelling. | **Popliteal cyst (Baker cyst)** |
| Non-fluctuating, non-translucent swelling. | **Soft tissue tumor** |

**Differential Diagnosis: Knee Movement Limitations**

| **Knee stretch limitation** | |
|---|---|
| Pressure pain on the distal side of the medial femoral condyle in a maximally flexed knee. | **Osteochondritis dissecans of the medial femoral condyle** |
| Pressure pain on the distal side of the lateral femoral condyle in a maximally flexed knee. | **Osteochondritis dissecans of the lateral femoral condyle** |
| Positive meniscal tests. | **Medial meniscal tear** |
| Pressure pain at the level of the lateral joint line and/or | **Discoid lateral meniscus** |
| Positive meniscal tests. | **Lateral meniscal tear** |
| **Knee bending limitation** | |
| The knee cannot be fully flexed, actively or passively. | **Quadriceps fibrosis** |

## Knee Effusion

### Knee Effusion Within a Few Hours After Trauma

Complaint: the knee swells up within a few hours after an obvious trauma.

Assessment: a large swelling can be seen on inspection. The swelling is usually located above and on both sides of the knee, and is horseshoe-shaped (Fig. 10.49). There tends to be a limitation of movement, particularly flexion and to a lesser degree extension. The knee is held in a slightly flexed position (Bonnet position, Fig. 10.50) if the swelling

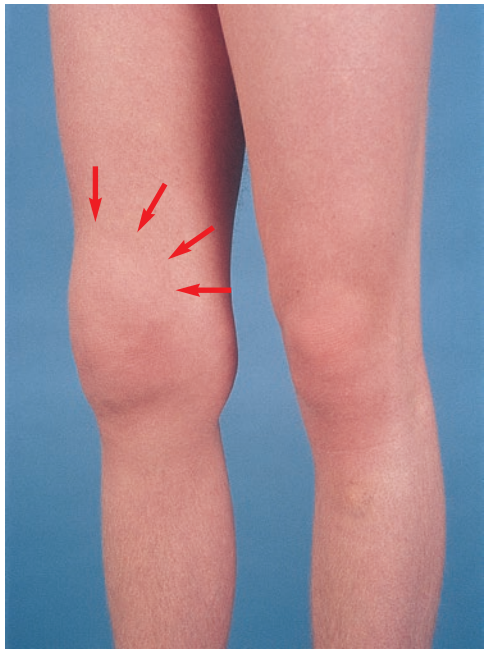


**Fig. 10.49** Hemarthrosis (or hydrops) of the right knee, manifested by a horseshoe-shaped swelling (*arrow*) above and on both sides of the patella

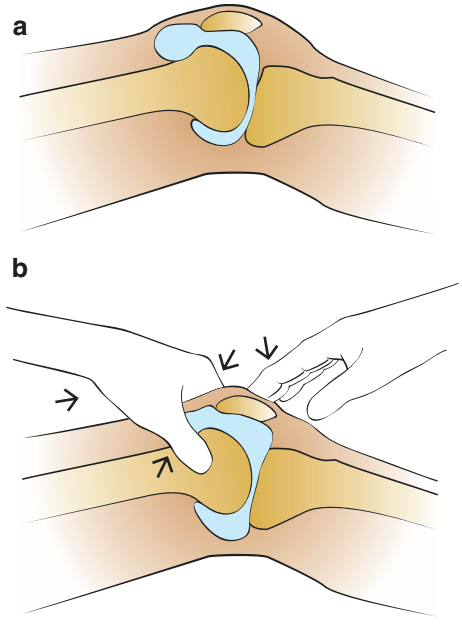



**Fig. 10.50** (**a**) Bonnet position (slight flexion). (**b**) Patellar tap test. In this test the suprapatellar recess is pressed empty distally with one hand, and with the fingers of the other hand the patella is pressed posteriorly. Normally speaking, the patella cannot be pressed posteriorly because it lies on the femoral condyles. The patella is elevated if there is a lot of blood or fluid in the knee, which allows some posterior movement

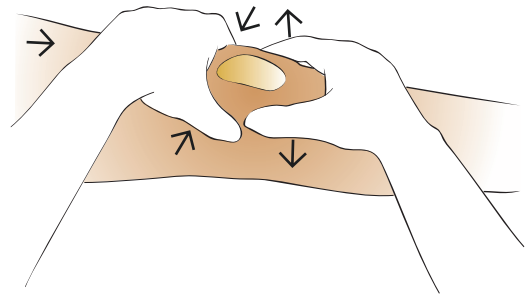


**Fig. 10.51** Cross fluctuation test. Place the thumb and index finger of the right hand on both sides of the patella. If you empty the suprapatellar recess with the other hand, the thumb and finger of the right hand will become separated from each other after the displacement of the fluid

is extreme. In this position the contents in the knee joint are the greatest and tension in the joint is at its lowest. The effusion in the knee is caused by hemorrhage or an abnormal accumulation of synovial fluid. The patellar tap test (Fig. 10.50), the cross fluctuation test (Fig. 10.51) and the dimple or bulge test (Fig. 10.52) are applicable when assessing hemarthrosis or hydrops.

A swollen knee is very painful. The patient is grateful if the pressure is reduced and the pain is less after a joint puncture with aspiration (Fig. 10.53); this allows better assessment. When assessing knee stability, examination of the medial and lateral ligaments is particularly important (Fig. 10.54). The severity of a collateral ligament injury can be classified into three degrees (Table 10.3). The tests for anterior, posterior and rotatory instability are not very reliable at an early stage, due to the swelling and because the knee is painful. Such tests may have to be done again after a week, when the knee is less painful (Figs. 10.54, 10.55, 10.56, 10.57, 10.58, 10.59, and 10.60 and Tables 10.4 and 10.5). Assessment on pressure pain points is also unreliable because the knee is sensitive everywhere immediately after the trauma,

Differential diagnosis:

**capsular tear**
**intra-articular fracture**
**ligamental rupture**

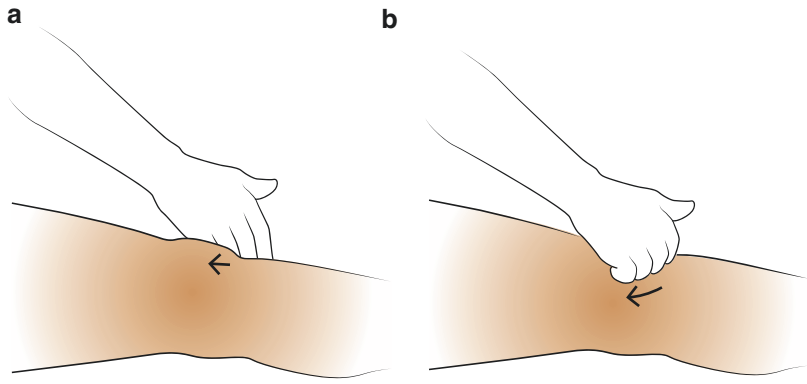



**Fig. 10.52** Dimple or bulge test. (**a**) Thoroughly stroke the medial dimple empty in a proximal direction with the back of the hand. (**b**) Next, you do the same on the lateral side, and the medial dimple will fill up again. This test is only practical in an asthenic patient with high-viscosity fluid in the knee

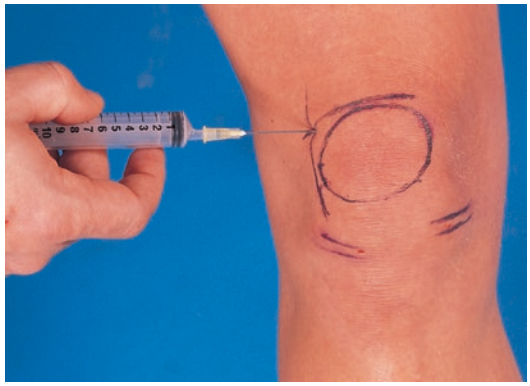


**Fig. 10.53** Aspiration of the right knee. It is best to aspirate the knee at the intersection of the parallel lines drawn on the proximal edge and lateral side of the patella if there is a hemarthrosis or hydrops

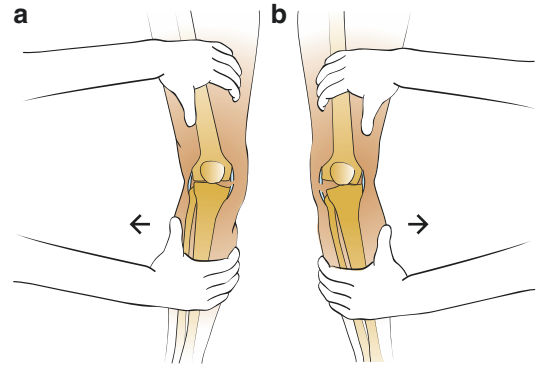



**Fig. 10.54** (**a**) Abduction (valgus) and adduction (varus) stress test for medial and lateral collateral ligaments ruptures. Test for medial ligamental instability: the knee is brought into 30° of flexion. The medial ligament can be tested by doing a valgus stress test. In a grade I anomaly, the pain is only indicated after a trauma around the medial ligament. In cases of grades II and III medial ligament instability you feel the distance increasing between the medial tibial and the medial femoral condyles. The medial tibial condyle rebounds into its previous position once you remove the valgus stress. (**b**) Test for lateral ligamental instability: the knee is brought into 30° of flexion. The lateral ligament can be tested by exerting varus stress. In a grade I anomaly, pain is indicated only after a trauma around the lateral ligament. In cases of grades II and III lateral ligament instability you feel the distance increasing between the lateral tibial and lateral femoral condyles. The lateral tibial condyle rebounds into its previous position once you remove the varus stress. The medial and lateral ligaments are not tested in an extended position. An extended knee will not diverge the tibial and femoral condyles even in cases of a complete rupture of the medial and lateral ligaments if the posterior cruciate ligament is intact

Explanatory note: the knee swells up within a few hours after trauma, this is usually caused by a hemarthrosis. There is usually an abnormal amount of joint fluid in the knee (hydrops) if the swelling occurs in the course of 24 h. A hemarthrosis resulting from a trauma is caused by a capsular tear, an intra-articular fracture or a ligamental rupture.

**Capsular tear** One can only speak of a capsular tear if other injuries have been ruled out.

**Intra-articular fracture** Intra-articular fractures in children can be growth-plate fractures, avulsion fractures of the anterior and posterior cruciate ligaments, or osteochondral fractures. Osteochondral fractures are caused by rotation trauma to the knee. A piece of cartilage with underlying bone can break off from the joint surface by shearing forces under load. Such injuries tend to affect adolescents.

**Table 10.3** The medial and lateral ligamental instability of the knee is expressed in three grades

| Grade of instability (valgus) stress | Abduction (varus) or adduction |
|---|---|
| Grade I | There is only pain at the medial or lateral ligament. |
| Grade II | The tibial and femoral condyles deviate, yet this is felt to be limited. There is an end point. |
| Grade III | The tibial and femoral condyles deviate, it feels as if nothing can stop the displacement. |

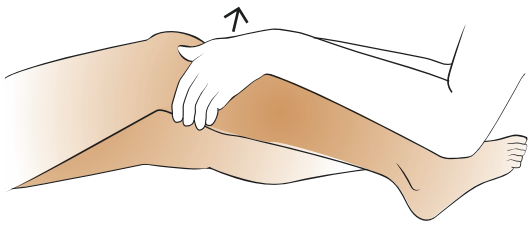


**Fig. 10.55** Lachman test for an anterior cruciate ligament rupture. The knee is flexed 20°. The tibial tuberosities are at an equal height. The examiner places both hands around the proximal part of the tibia with the thumbs parallel to the patellar tendon, and exerts a forward force on the lower leg. No displacement will be felt if the anterior cruciate ligament is intact. There is anterior cruciate ligament instability if anterior displacement is greater than 3 mm. The Lachman test is positive in more than 90 % of cases in an anterior cruciate ligament rupture in a non painful knee. The test is much more reliable than the anterior drawer test with the knee in 90° of flexion

**Ligamental rupture** Ruptured ligaments are much less common in children than in adults thanks to ligamental elasticity. A severe rotational trauma of the knee in children with open growth plates will be more likely to cause an avulsion fracture or a fracture around the growth plate rather than ligamental ruptures. Ligamental ruptures are usually found in children older than 12 years of age. ! Hemarthrosis can cause adhesions which reduce the mobility of the knee. The hemarthrosis will cause a painful synovitis that may last up to 6 weeks even if there are no adhesions. In 75 % of cases this involves an anterior cruciate ligament rupture; 70 % of such patients hear or feel a loud snap during the trauma. This can also occur with an osteochondral fracture. An anterior cruciate ligament rupture is accompanied by a medial ligament rupture or a meniscal tear in 25 % of cases. There is usually a combination of injuries.

*Posteromedial rotatory drawer test* The same manoeuver will be performed. In this manoeuver the medial tibia plateau will, beside shifting posteriorly, rotate posteriorly if there is a rupture of the medial ligament, oblique popliteal ligament and the posteromedial capsule. The internal rotation in the knee is increased. The test is considered positive if the shift exceeds 3 mm.

*Posterolateral rotatory drawer test* In this manoeuver the lateral tibia plateau will, beside shifting posteriorly, rotate posteriorly if there is a rupture of the arcuatum complex. The external rotation in the knee is increased. The test is considered positive if the shift exceeds 3 mm.

Supplementary assessment: anteroposterior and lateral X-rays of the knee to identify or rule out fractures.

Primary care treatment: it can be assumed that there is a capsular tear if physical assessment gives no indication of a ligamental rupture and in the case where radiological assessment shows no fractures. A pressure bandage is sufficient in that case, if necessary after knee aspiration. A large hemarthrosis is very painful. The aspiration is intended to alleviate pain.

No treatment is necessary for a grade I and II medial or grade I lateral ligament injury. The patient can be mobilized using elbow crutches, with the amount of weight bearing depending on the symptoms. The symptoms usually disappear within 6 weeks.

When to refer: the patient should be referred (Table 10.6) in a grade III medial and grade II and III lateral instability. It is also advisable to refer patients with an anterior or posterior cruciate ligament injury who still have a sprained feeling after exercise therapy.

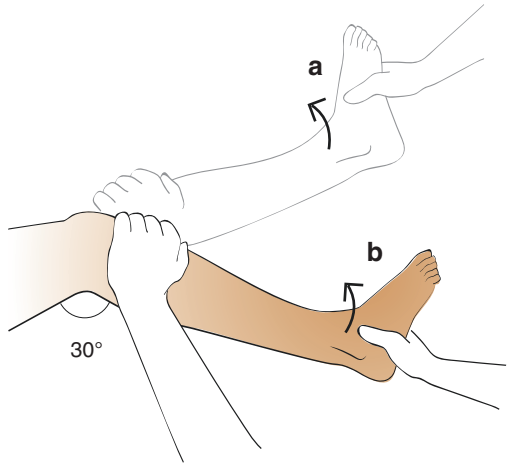



**Fig. 10.56** Lateral pivot shift test. This test also serves to determine anterolateral rotatory instability. The extended leg is lifted by the examiner and rotated internally at the hip and the knee (**a**). In the case of an anterior cruciate ligament rupture an enhanced internal rotation of the tibia will be present and the lateral tibial plateau has a more anterior position. Next, the knee is flexed slowly. In 30° of flexion the lateral tibial plateau is displaced abruptly in a posterior direction, causing the powerful iliotibial muscle to act as a flexor (**b**). After an acute injury the test is very difficult or impossible to perform without anesthesia, so the reliability in acute lesions is low. In chronic lesions the reliability is considerably greater, as long as the patient can relax. Stability assessment under anesthesia gives the best results. The pivot shift test has a sensitivity of 82 % and a high specificity of 98 %

Secondary care treatment: **intra-articular fracture.** One should aim for anatomical reduction in the case of an intra-articular fracture. Arthroscopic or operative reduction is carried out with fracture fixation by means of an osteosynthesis if a closed (noninvasive) reduction is not possible.

**Acute medial and lateral collateral ligament rupture** An acute medial ligament rupture grade III is treated conservatively with cast immobilization from the ankle to halfway up the thigh, with the knee in 10° of flexion for 6 weeks. Medial meniscal tears, which are present in 20 % of grade III medial ligament ruptures, tend to be located close to the capsule and in 90 % of cases recover during the 6-week period of cast immobilization. A lateral ligament rupture or a rupture of the arcuate complex grade II and III degrees need to be restored operatively.

**Chronic medial ligamental instability** There can still be instability in the acute period after a medial ligamental rupture, giving the child a sprained feeling around the knee. Chronic medial ligamental instability usually causes no symptoms. In exceptional cases the medial ligaments can be tightened by detaching them at the level of the femoral condyle with a thin piece of bone, with proximal displacement and fixation.

**Chronic lateral ligamental instability** Chronic lateral ligamental instability does tend to cause complaints. Isolated lateral ligamental instability is rare. Reconstruction by using part of the biceps tendon can be performed in the case of an isolated lesion.

**Acute anterior and posterior cruciate ligament rupture** Operative reduction and stabilization is done only for dislocated avulsion fractures of the cruciate ligaments. A wait-and-see approach can be adopted if the anterior or posterior cruciate ligament is ruptured in the ligamental part. Acute operative reattachment of the cruciate ligament is pointless because after a trauma the ends become necrotic and retract. Suturing such a rupture does not produce a functional anterior cruciate ligament for this reason. Physiotherapy can be prescribed after the acute period in the case of a sprained feeling in the knee. Hamstring exercises are prescribed for an anterior cruciate ligament rupture, and quadriceps exercises for the posterior cruciate ligament. Most patients with a posterior cruciate ligament rupture have few symptoms after the acute phase without treatment.

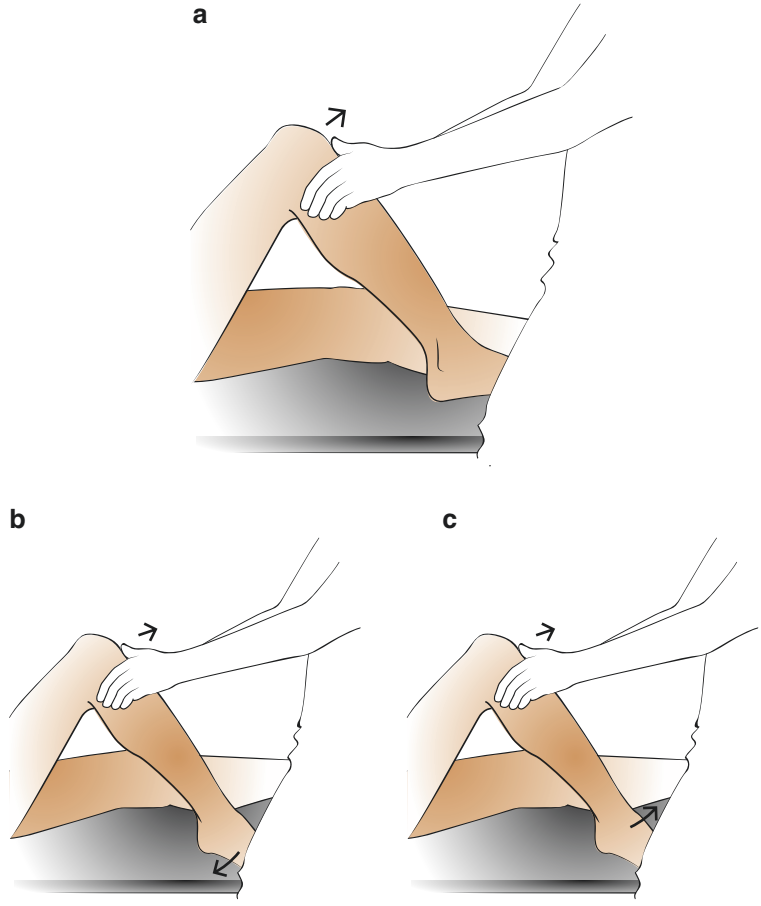



**Fig. 10.57** (**a**) Anterior drawer test for an anterior cruciate ligament rupture. The patient lies supine with the hip flexed at 45° and the knee at 90°. The foot points directly forward and is fixed by the examiner by sitting on the forefoot. The tibial tuberosities are at an equal height. The examiner places both hands around the proximal part of the tibia with the thumbs parallel to the patellar tendon, and exerts an anterior force on the lower leg. The patient is not allowed to tighten his hamstrings. No movement will be felt if the anterior cruciate ligament is intact. There is anterior cruciate ligament instability if the forward displacement is more than 3 mm. Sensitivity of the anterior drawer test for the anterior cruciate ligament is low. In acute cases it is 22 %, and in chronic cases 50 % with a specificity of 95 %. The anterior drawer test is generally only positive if there is also an accompanying medial or lateral ligamental injury. The anterior drawer test tends to be negative in isolated anterior cruciate ligament ruptures. (**b**) Anteromedial rotatory drawer test. The patient lies supine with the hip flexed 45° and the knee 90°, the foot is fixed in external rotation on the examination table. Next, the anterior drawer test is performed. In this manoeuver the medial tibial plateau will, besides shifting anteriorly, rotate anteriorly, if there is a rupture of the medial ligament, including the oblique popliteal ligament. This may be enhanced by the presence of an anterior cruciate ligament rupture. The test is considered positive if the anteromedial shift exceeds 3 mm. (**c**) Anterolateral rotatory drawer test. The patient lies supine with the hip at 45° and the knee bent 90°, while you fix the foot in internal rotation. In this test the lateral tibial plateau will, beside shifting anteriorly, rotate anteriorly if there is a rupture of the lateral collateral ligament. This is possibly enhanced by a pre existing anterior cruciate ligament rupture. The test is positive if the anterolateral shift exceeds 3 mm

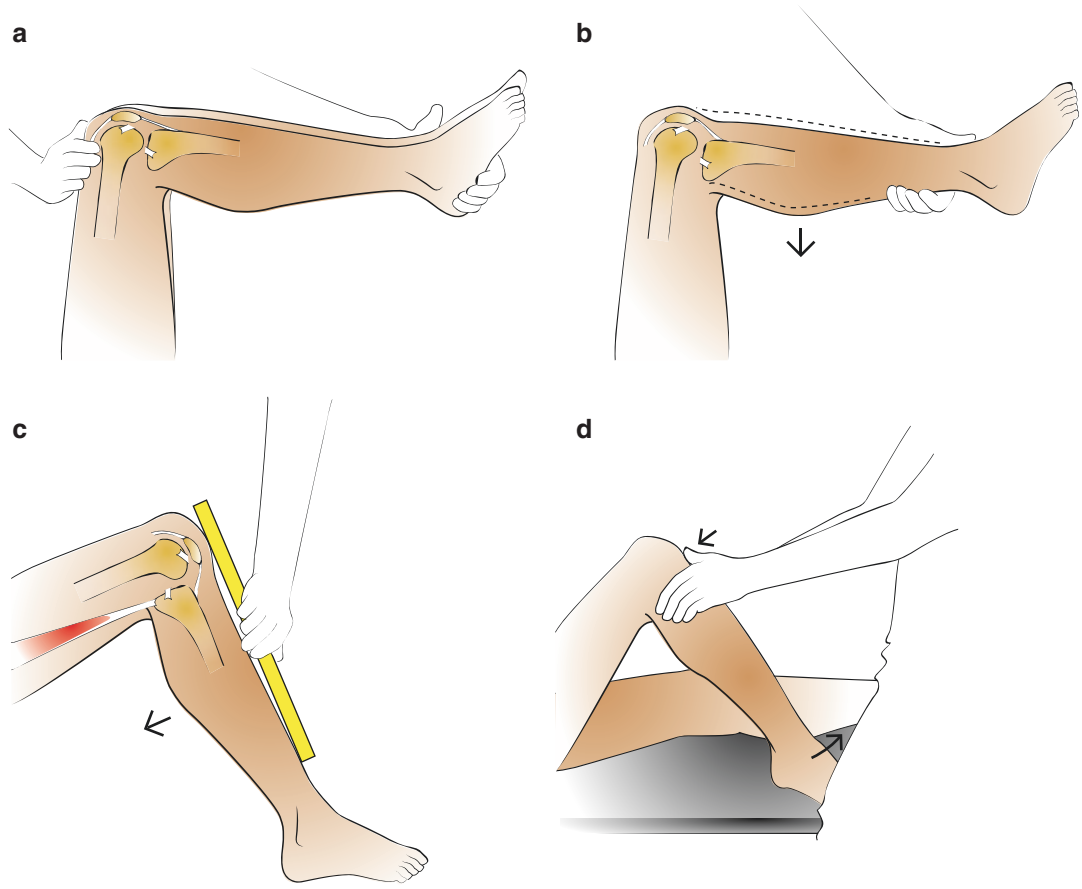



**Fig. 10.58** Gravity test for a posterior cruciate ligament rupture. The knee is flexed 90°: (**a**) with an intact posterior cruciate ligament the tibial tuberosity is at an equal height in both knees. (**b**) If there is posterior cruciate ligament instability the tibial tuberosity of the affected side will be posteriorly positioned when compared with the contralateral side with an intact posterior cruciate ligament. (**c**) An alternative way to do the gravity test is with the hips at 45° and the knees flexed 90°. The lower legs are in a neutral rotational position with the feet on the examination table. Both knees are compared laterally with each other. If the tibial tuberosity of the injured knee has collapsed posteriorly we are dealing with a posterior cruciate ligament rupture. In a knee that is bent 90°, if the posterior cruciate ligament is intact the tibial tuberosity will be positioned 1 cm anterior to the medial femoral condyle. If there is a posteriorly oriented shift of more than 1 cm, the tuberosity will lie behind the distal boundary of the medial femoral condyle. (**d**) Posterior drawer test for a posterior cruciate rupture. The patient lies supine with the hip 45° flexed an the knee 90°. The foot points straight forward and is fixed by the examiner sitting on the foot. The tibial tuberosity of the affected side will be posteriorly positioned compared to the contralateral side. The examiner places both hands around the proximal part of the tibia with the thumb parallel to the patellar tendon, and exerts a forward force on the lower leg until the tibial tuberosities are at the same level. Next the examiner exerts a posterior force on the lower leg. The patient is not allowed to tighten the hamstrings. There is an posterior cruciate rupture if there is a posterior shift more than 3 mm

**Chronic anterior cruciate ligament, anteromedial rotatory and anterolateral rotatory instability** In general, an isolated ruptured anterior cruciate ligament produces no instability. There is also medial or lateral ligament instability in the case of instability symptoms. A orthosis can be prescribed (Fig. 10.61) if the patient only has complaints when carrying out certain sport activities. Reconstruction may be considered if symptoms persist. An anterior cruciate ligament reconstruction is performed for anterior cruciate ligament instability in the case with anteromedial and/or anterolateral instability. This procedure involves drilling through the growth plate in the distal part

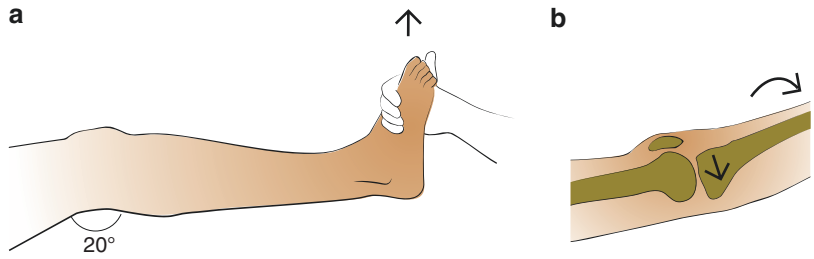



**Fig. 10.59** External rotation recurvatum test. This test serves to determine a posterolateral rotatory instability. (**a**) The examiner lifts the entire leg with the knee 20° flexed. Next, the knee is slowly extended. (**b**) There is external rotation of the lower leg and a recurvation of the knee in the case of a posterolateral rotatory instability. This instability is caused by a rupture of the arcuate complex (lateral ligament, popliteal tendon, arcuate popliteal ligament). Usually there is also a posterior cruciate ligament rupture

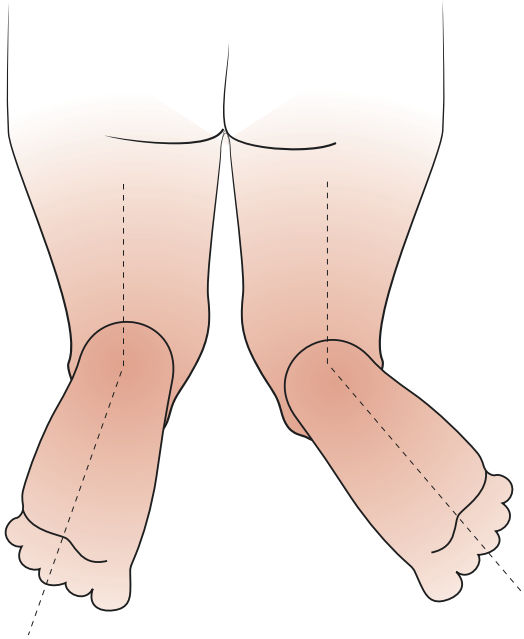


**Fig. 10.60** Dial test. This test is also known as a posterior late-ral external rotation test, also serves to determine posterolateral instability. The patient lies prone with the knees touching. The examiner encompasses the feet and holds them in dorsal extension, then both lower legs are turned in external rotation and one assesses the range of rotation. The test is performed twice, once with the knees in 30° of flexion and once with the knees in 90° of flexion. An increased external rotation of more than 10° in the injured knee compared with the uninjured knee in 30° but not in 90° of flexion indicates a possible isolated injury of the arcuate complex. An increased external rotation of more than 10° in 90° but not in 30° of flexion points to a lesion of the posterior cruciate ligament. In a combined injury of the arcuate complex and the posterior cruciate ligament the external rotation in the injured knee compared to the uninjured knee is increased by more than 10° in 90° and 30° of flexion. The sensitivity and specificity of this test are not known. The figure shows a positive dial test with the knees in 90° of flexion

**Table 10.4** The severity of the anterior or posterior cruciate instability is expressed in three grades. In an anterior cruciate ligament rupture the drawer test produces an anterior shift of the tibia with respect to the distal part of the femur, and in a posterior cruciate ligament rupture a posterior shift. In a knee flexed to 90° the tibial tuberosity will lie 1 cm in front of the medial femoral condyle if the ligamental apparatus of the knee is intact

| Degree of instability | Anterior or posterior shift with respect to the healthy side |
|---|---|
| Grade I | 3–5 mm |
| Grade II | 5–10 mm |
| Grade III | More than 10 mm |

of the femur and the proximal part of the tibia. The operation is preferably postponed until the child is fully grown in order to prevent a growth disorder as much as possible. The anterior cruciate ligament is reconstructed using tendons from the gracilis and semitendinosus muscles (Fig. 10.61). The reconstructed anterior cruciate ligament is positioned in the same area as the original anterior cruciate ligament. The success percentage of this operation is about 80 %. The patient is not allowed to do sporting activities for 6 months after such an operation.

**Chronic posterior cruciate ligament, posterolateral rotatory instability and posteromedial instability** Symptoms chiefly occur if there is posterolateral rotatory instability. A orthosis can be prescribed in the first instance. The results of a posterior cruciate ligament reconstruction vary. Reconstruction is only carried out in extreme cases. Most patients do well without operative treatment.

**Table 10.5** Schematic overview of knee instability, corresponding ligamental lesion(s) and diagnostic manoeuvers. Posteromedial (rotatory) instability is rarely seen in practice

| Instability | Ligamental lesion(s) | Diagnostic manoevers |
|---|---|---|
| Medial | Medial ligament | Abduction (valgus) stress in 30° flexion |
| Lateral | Lateral ligament | Adduction (varus) stress in 30° flexion |
| Anterior cruciate ligament | Anterior cruciate ligament | Lachman test |
| | | Anterior drawer test |
| Anteromedial rotatory | Medial ligament, including the posterior oblique ligament | Anterior drawer test in external rotation |
| | Mostly the anterior cruciate ligament | |
| Anterolateral rotatory | Lateral ligament | Anterior drawer test in internal rotation |
| | Nearly always the anterior cruciate ligament | Lateral pivot shift test |
| Posterior cruciate ligament | Posterior cruciate ligament | Gravity test |
| | | Posterior drawer test |
| Posteromedial rotatory | Medial ligament, oblique popliteal ligament, posteromedial capsule | Posteromedial rotatory drawer test |
| | Often posterior cruciate ligament | |
| Posterolateral rotatory | Arcuate complex (lateral ligament, popliteal tendon and arcuate popliteal ligament) | Posterolateral rotatory drawer test |
| | | External rotation recurvation test |
| | Mostly the posterior cruciate ligament | Dial test |

**Table 10.6** Decision scheme for a knee effusion in children after a trauma

| |
|---|
| **Knee effusion within a few hours after a trauma (hemarthrosis):** |
| DD: capsular tear |
| Intra-articular fracture |
| Ligamental rupture |
| Primary care treatment: possible knee aspiration, ligamental assessment, X-rays of the knee. |
| When to refer to an orthopedic surgeon: |
| Fractures |
| Grade III medial and grades II and III lateral ligamental ruptures |
| For the rest, wait. A hemarthrosis, just like grade I and II medial and grade I lateral ligament instability, causes pain during 6 weeks. |
| **Knee effusion within 24 h after a trauma (hydrops):** |
| DD: distortion |
| Meniscal tear |
| Primary care treatment: wait, more than 90 % of these knee problems disappear spontaneously within three months. |
| When to refer to an orthopedic surgeon: |
| Extension limitation |
| Persistent hydrops |
| If symptoms persist for more than 3 months |

A very common reconstructive technique is detachment of the arcuate complex at the level of the femur and transposing this proximally and anteriorly (‘advancement’ technique of the arcuate complex). This may also be combined with an intra-articular reconstruction of the posterior cruciate ligament using the tendons of the gracilis and semitendinosus muscles.


**Fig. 10.61** (**a**) Orthosis for knee instability. (**b**) Anterior cruciate ligament reconstruction using tendons from the gracilis and semitendinosus muscles

If necessasary reconstruction of a chronic posteromedial rotatory instability is frequently performed using the semitendinosis tendon.

## Knee Effusion Within 24 h After Trauma

Complaint: painful and swollen knee that has developed within 24 h after a trauma.

Assessment: the knee is usually not very swollen; the cross fluctuation test or the dimple test are usually indicated for confirming the presence of fluid in the joint. There is pressure pain around the joint gaps, and the meniscal tear tests can be positive (Figs. 10.42, 10.43, 10.44, 10.45, 10.46, and 10.47).

Differential diagnosis**:**

**distortion**
**meniscal tear**

Explanatory note: **distortion.** There is usually a distortion if the swelling does not develop immediately after a trauma but in the course of the first 24 h.

**Meniscal tear** A medial or lateral meniscal tear is rare in children under the age of 12 (Fig. 10.62). A meniscal tear should be suspected in older children if the symptoms have not disappeared after 3 months (for more on meniscal tears, see pp. 202–205).

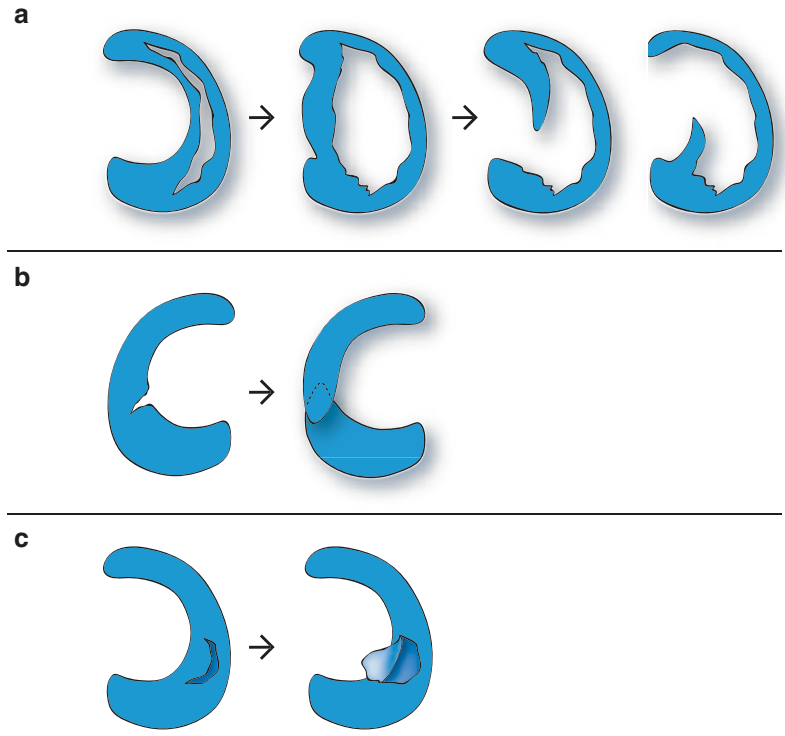



**Fig. 10.62** Different types of meniscal tears. (**a**) A vertical lengthwise tear can end up as a 'bucket handle' tear, which mostly starts in the posterior end of the meniscus and gradually expands in an anterior direction. A vertical lengthwise tear can also tear out centrally, causing a posterior or anterior frayed tear. (**b**) In a radial tear there is a vertical tear perpendicular to the concave central side of the meniscus. Around the popliteal tendon, such a radial tear in the lateral meniscus can run all the way into the joint capsule, possibly causing the anterior and posterior sides to slide over each other. Because of its shape, it is also known as a "parrot beak" tear. In children we generally see a vertical lengthwise tear. (**c**) A horizontal (cleavage) tear can end in a flap tear. Horizontal tears tend to be degenerative and occur mainly after age 40. They can end up taking on the shape of a complex tear made up of horizontal as well as vertical components

Supplementary assessment: is not indicated during the first 3 months.

Primary care treatment: a wait-and-see approach can be adopted for 3 months. The symptoms disappear spontaneously in more than 90 % of cases. A torn meniscus should be suspected if the symptoms haven't disappeared after 3 months.

When to refer: if the symptoms haven't disappeared after 3 months (Table 10.5).

Secondary care treatment: **meniscal tear** (for secondary treatment, see pp. 204, 205).

## Knee Effusion Without a Preceding Trauma

Complaint: the child is not ill and has no fever. There is however a painful swollen knee without a preceding trauma.

Assessment: the cross fluctuation test or dimple test (Figs. 10.50 and 10.51) reveals that there is fluid in the knee.

Differential diagnosis:

**osteochondritis dissecans of the femoral condyle**

**juvenile idiopathic arthritis**
systemic type (Still disease)
pauciarticular type (oligoarticular type)
polyarticular type
**hemophilia bleeding disorder**
**pigmented villonodular synovitis**
focal type (latent nodular type)
diffuse type
**reactive arthritis**
**Lyme disease**

Explanatory note: **osteochondritis dissecans of the femoral condyle.** There are no or few symptoms in an early stage of an osteochondritis dissecans. Sometimes there is a hydrops or an extension limitation. (see pp. 200, 201 and 204)

**Juvenile idiopathic arthritis** This is a condition that starts before the sixteenth year of life. This is an arthritis of one or several joints that persists for longer than 6 weeks and for which other causes have been ruled out. It may begin with a swollen and warm knee. Pain isn't always in the foreground, and a swollen knee is often the first symptom. We distinguish three types depending on the trajectory in the first 6 months: the systemic type, also known as Still disease; the pauciarticular type in which one to four joints are affected; and the polyarticular type, which affects five or more joints (Table 10.7).

Juvenile idiopathic arthritis should always be considered if several joints are involved in the process. The diagnosis becomes more difficult if only one joint, for example the knee, is involved in the pauciarticular type. The incidence of juvenile idiopathic arthritis is 2 in 10,000 children per year.

*Systemic juvenile idiopathic arthritis* This type was described in 1897 by the English pediatrician Still. It is a combination of chronic arthritis with at least one of the following anomalies: fleeting small-spot exanthema without fixed localization, generalized lymph gland enlargement, hepatosplenomegaly or serositis (pericarditis, pleuritis or peritonitis). This type involves 20 % of children with juvenile idiopathic arthritis. The anomaly is distributed equally in boys and girls, and occurs at any age. The situation is dominated by general malaise, and is characterized by fever peaks up to 40 °C that occur late in the afternoon and last several hours, accompanied by fleeting small-spot exanthema without fixed localization. Infectious parameters are elevated and there is a microcytic anemia. Rheumatic factors and antinuclear antibodies are usually not demonstrable. Polyarthritis may develop sometimes immediately at the start or several weeks later. The course of the disease varies considerably. There are moderate to severe articular abnormalities in 20 % of children. Mortality rate is less than 0.5 %.

**Table 10.7** Classification of juvenile idiopathic arthritis

| |
|---|
| **Systemic type (still disease)** |
| Polyarthritis with or preceded by daily intermittent fever for at least 2 weeks combined with at least one of the following four characteristics: |
| Fleeting exanthema |
| Generalized lymph gland enlargement |
| Hepatomegaly or spenomegaly |
| Serositis (pericarditis, arthritis or peritonitis) |
| **Pauciarticular type (oligoarticular type)** |
| Arthritis in one to four joints during the first 6 months. |
| **Polyarticular type** |
| Arthritis in five or more joints during the first 6 months. |

*Pauciarticular juvenile idiopathic arthritis* This type is seen in 50 % of all cases. One to four joints are affected during the first 6 months. The number of affected joints does not exceed four in 80 % of cases after 6 months. The number is five or more after 6 months in 20 % of cases. This type is 4–5 times more common in girls than in boys. The arthritis begins at a young age, before age 8, with a peak incidence between the ages of 1 and 3. The polyarticular type will occasionally be seen in teenage girls. It tends to be insidious and doesn't usually cause many symptoms. The disease often begins in the knee. Tendovaginitis of the flexor tendon of the middle or ring finger is common, as a first sporadic sign.

Chronic uveitis affects 20 % of patients. This produces no symptoms or external manifestations, and can only be determined with a slit lamp. The antinuclear antibody test (ANA) is often positive, but rheumatoid factors are negative.

Infectious parameters are normal. Although a specific HLA can be found, it has no significance up to the present time.

*Polyarticular juvenile idiopathic arthritis* In this type, five or more joints are affected during the first 6 months; 30 % of all children with juvenile idiopathic arthritis fall into this category. The first peak incidence is found between the ages of 2 and 5, affecting boys as well as girls. The second peak affects primarily girls aged 12–16.

This type tends to be insidious, affecting initially one or two joints, and gradually affecting more. This is often preceded by a period of subfebrile temperature, excessive transpiration, weight loss and joint stiffness. The arthritis is predominantly distributed symmetrically and is localized in large joints such as knees, wrists, elbows and ankles. The cervical spine and the mandibular joints are often also affected. The small joints of the hand and foot can become inflamed at an earlier or later stage. Chronic uveitis affects 10 % of patients. The inflammatory parameters are elevated. There is usually hypocytic or normocytic anemia. In 10 % of patients the rheumatoid factor is positive, and in 25 % the ANA test is positive; 25 % of patients have HLA-B27. The disease has an aggressive course in 80–90 % of patients, nearly all girls, and develops similar to patients with the adult type of rheumatoid arthritis.

**Hemophilia bleeding disorder** Intra-articular hemorrages can occur as a result of clotting disorders and the knee is frequently involved.

**Pigmented villonodular synovitis** This is an inflammatory tumor growth in the synovial joint capsule (Fig. 10.63). There are two types, focal and diffuse. The incidence of the focal type is two cases and in the diffuse type nine cases per million population.

*Focal type* The focal type or latent nodular type is the least aggressive and typically occurs in the smaller joints of the hands and feet. This type is predominantly seen in females. Although rare the focal type may develop in large joints.

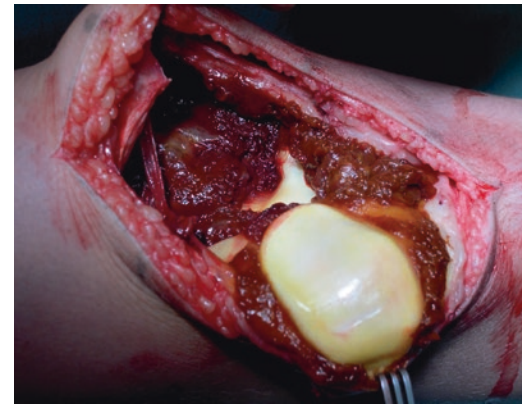


**Fig. 10.63** Pigmented villonodular synovitis of the knee (diffuse form) with characteristic *brown-colored* proliferation of the joint capsule

*Diffuse type* The preferred localization of this tumor is the knee (80 %) and to a lesser extent in the hip, ankle and shoulder. Radiological assessment may reveal small bony defects around the attachment of the joint capsule.

**Reactive arthritis** An infection elsewhere in the body can give rise to a type of arthritis known as reactive arthritis. It often occurs as a result of a viral infection elsewhere in the body. Different viruses, such as hepatitis-B, rubella, human parvovirus B19, herpes simplex, varicella, cytomegalia and Epstein-Barr, can cause arthritis directly or as a result of an immunological reaction. The course of such an arthritis tends to be less severe than that of a septic arthritis. The examiner usually gets clues from other manifestations of specific viral infections. Most viral arthritides are self-limiting and of a short duration, usually less than 6 weeks.

A reactive arthritis can also develop 2–8 weeks after a bacterial infection elsewhere in the body. It usually involves a urogenital tract infection or an intestinal infection. It usually begins between 20 and 40 years of age, but reactive arthritis can also occur at the end of puberty. Between 70 and 80 % of these patients are carri-

ers of the HLA-B27 antigen. In the initial phase these arthritides are difficult to distinguish from a septic arthritis, but their course is much less severe. Most patients are symptom-free within 1 year. Normally reactive arthritis is considered to be a self-limiting condition, but in 30–50 % of patients multiple episodes occur and in 20 % of cases the disease becomes chronic, which can lead to joint destruction.

**Lyme disease** A special form of reactive arthritis is Lyme disease. This disease is provoked by Borrelia burgdorferi, which is transmitted after a tick bite in less than 1 % of cases. A clear sign of infection is a red ring on the skin around the bite, known as erythema migrans. This red ring, with an often-paler center, appears 2–30 days after the bite. Within several months 60 % of cases show a reactive arthritis if the disease remains untreated and usually involves the knee (90 %) or other joints such as elbow, ankle, hip and wrist. This arthritis is often difficult to distinguish from a septic arthritis, especially as there is fever in 20–50 % of cases. Several joints (One to four) may also be involved. The oligoarticular type is difficult to distinguish from the oligoarticular type in juvenile idiopathic arthritis. There is a recurrence after several months up to a year after the disease appears in half of the cases, even after treatment with antibiotics. Neurological conditions such as facial nerve paresis or paralysis or meningitis appear several days or even months later in 15–20 % of cases of untreated Lyme disease,

Supplementary assessment: an anteroposterior, a lateral and a tunnel view X-ray of the knee should be requested if osteochondritis dissecans is suspected. The diagnosis of hemophilia is usually already known. Bleeding into the joint is very probable if a swollen knee appears in a patient who is known to have hemophilia. It is not a good idea to do a knee aspiration without current information about the patient's clotting factors. The knee can be aspirated in the other cases. A clear joint aspirate is seen in the case of an osteochondritis dissecans. The joint fluid will be cloudy in the event of juvenile idiopathic arthritis and reactive arthritis. A brown aspirate is seen in pigmented villonodular synovitis. Blood tests can be done for rheumatic factors, antinuclear antibodies (ANA), HLA-B27, BSE, CRP, leukocyte numbers, Hb and Ht if juvenile idiopathic arthritis is suspected. An MRI should be requested if pigmented villonodular synovitis is suspected. Serological testing can be done if reactive arthritis is a possibility.

Primary care treatment: children with swollen knees after a preceding trauma will not generally be treated by the general practitioner, they must be referred for further assessment.

When to refer: an osteochondritis dissecans with a persistent hydrops must be referred to an orthopedic surgeon. Children with an unexplained swollen knee or those with hemophilia will generally be referred to a pediatrician for treatment. The pediatrician will usually refer the child to a pediatric rheumatologist, or to an ophthalmologist in case of uveitis in cases of juvenile idiopathic arthritis.

Secondary care treatment: **osteochondritis dissecans of the femoral condyle** (for secondary treatment, see pp. 204, 205).

**Juvenile idiopathic arthritis** Juvenile idiopathic arthritis is initially treated with NSAID's. An intra-articular injection with corticosteroids can be given in isolated cases. In addition, children should receive intensive physiotherapy to prevent contractures.

An orthopedic surgeon should be involved in 10 % of children with juvenile idiopathic arthritis in which severe joint abnormalities develop. Serial plaster casts will be used to correct a 10–20° flexion contracture. Sometimes lengthening the tendons of semimembranosus, gracilis, biceps and semitendinosus muscles are indicated if there is a flexion contracture of 20° or more. The indication for synovectomy is very limited.

This may be considered for children with an oligoarticular arthritis. The frequently seen valgus deformity of the knee can be corrected by performing a temporary hemi-epiphysiodesis on the medial side of the distal part of the femur using clamps, an eight-plate or screws, between the ages of 11 and 12. The materials can be removed as soon as the knee is straight. If there is still a valgus position a supracondylar corrective osteotomy can be performed once the child has finished growing. A total knee replacement arthroplasty is reserved for severe destruction of the knee joint, in which the child has pain at rest and when walking is severely limited. Such arthroplasties can be carried out from the approximate age of 12 years. Although the short-term results of a joint replacement arthroplasty are excellent, many of the inserted prostheses will loosen up after 10 years. Both patient and practitioner have little choice: be wheelchair-bound with a lot of pain, or walk for 10 years without pain.

**Hemophilia bleeding disorder** Sufficient hemostasis should be insured to stop hemorrhage and prevent recurrent hemarthrosis.

**Pigmented villonodular synovitis** The treatment of choice is an arthroscopic partial synovectomy for the focal type and an open procedure for the diffuse type (a total knee). A total synovectomy is technically impossible in a knee. This is why the diffuse variant has a major risk of recurrence after a synovectomy.

**Reactive arthritis** A reactive arthritis resulting from a viral infection will be treated expectantly. The bacterial infection should be treated first in the case of reactive arthritis after a bacterial infection elsewhere in the body. Treatment for the reactive arthritis itself is not necessary. A 30-day antibiotic cure is prescribed in cases of arthritis resulting from Lyme disease. One or two antibiotic treatments result in recovery in 80 % of cases. An intra-articular injection with corticosteroids can be considered if the arthritis persists after the second treatment with antibiotics.

## Knee Effusion Skipping to Other Joints

Complaint: These are usually sexually active teenagers. The joint symptoms are sometimes preceded by a short period with fever and a fleeting skin rash, especially around the ankles and lower legs. The pain and swelling are mostly localized on the dorsal side of the hands, wrists, ankles and feet, but can also appear in the knee.

Assessment: the joints in the upper extremities, especially those of the fingers and the wrist, are more frequently involved than the lower extremities joints.

Diagnosis: **gonococcal arthritis**

Explanatory note: **gonococcal arthritis.** One should consider gonorrhea in sexually active teenagers who have skipping joint complaints (polyarthritis migrans). A polyarthritis begins usually 2–4 weeks but sometimes after years after the infection and usually begins slowly, but there can also be a acute type. In an acute type more joints can sometimes be involved, however the sternoclavicular joint is amongst others one of the most commonly involved joint.

The patients complain in 80 % of cases about skipping joint pain and swelling. The joints of the upper extremities, particularly the fingers and wrists are more involved than the joints of the lower extremities.

Supplementary assessment: the temperature is often subfebrile. Laboratory assessment: the WBC count, CRP and ESR is less elevated than in a septic arthritis. A gonococcus (Neisseria gonorrhoeae) can be demonstrated in aspirated joint fluid only in 50 % of cases.

Primary care treatment: none.

When to refer: if gonococcal arthritis is suspected.

Secondary care treatment: gonococcal arthritis is penicillin-sensitive. Generally there is no need for joint aspiration or rinsing. The

patient is commonly given 3 days of penicillin intravenously and 4 days orally.

## Knee Effusion Accompanied by Nocturnal Sweating

Complaint: there is a painful and swollen knee joint. The patient is ill, has subfebrile temperature, suffers from night sweating and is losing weight (emaciation). There is pulmonary, intestinal or renal tuberculosis in the medical history.

Assessment: there is considerable joint capsule thickening and a pronounced atrophy of the quadriceps muscle. The joint capsule thickening is easiest to palpate around the synovial fold of the suprapatellar recess above the knee. Mobility of the joint is limited in all directions. Considerable muscle atrophy is noticeable.

Diagnosis: **tuberculous arthritis**

Explanatory note: **tuberculous arthritis.** More than two million people die of tuberculosis worldwide every year. Tuberculosis, caused by Mycobacterium tuberculosis, is rare in more developed countries. Usually there is a hematogenic dissemination from a pulmonary, renal or intestinal tuberculosis in this type of osteomyelitis or infected arthritis. 5% of cases end up with bone or joint infections following pulmonary tuberculosis,. The spinal column is the most commonly affected, followed in decreasing frequency in the hip, knee, ankle, sacroiliac joint, shoulder or wrist. Bone and joint tuberculosis affects children as well as adults. Many years ago it were mainly young children who had bone and joint tuberculosis. The peak has shifted to adults between the ages of 40 and 60 during the last 40 years.

There are two types of tuberculosis, a primary osseous type and a primary synovial type. The primary osseous type is the most common. The bone lesion tends to appear in the metaphysis of a long bone, mostly in the hip and the knee in children and can penetrate into the joint and opposing side of the joint.

Joint tuberculosis can also start with a synovitis (primary synovial type), but usually occurs because a lesion breaks through into the joint from the metaphysis (secondary synovial type). The condition can be broadly classified into three stages. The boundaries of the stages are not too well defined, and the various types often overlap.

**First stage: hydrops stage** In this stage there are para-articular bony lesions with symptomatic (reactive) fluid accumulation in the joint, or one is dealing with a condition in which the synovial capsule has tuberculous inflammatory lesions (secondary or possibly primary).

**Second stage: fungal** The capsule swells substantially due to tuberculous granulations. The joint becomes spongy, like a mushroom. The surrounding musculature becomes strongly atrophic. As a result of this the spindle-shaped swollen knee becomes even more noticeable. Mobility is extremely limited and the patient complains about joint pain. The skin around the spindle-shaped swollen knee is pulled tight and is white. This appearance of the knee led to the nomenclature tumor albus. Contractures can be caused by distruction of the joint cartilage or the joint ligaments. A flexion contracture develops in the knee. A subluxation occurs in the knee joint in which the tibia is displaced posteriorly if the joint ligaments and part of the capsule are destroyed.

**Third stage: purulent caseous** Necrosis occurs in the granulation tissue, transforming this into a cheeselike mass (caseous is Latin for cheesy). In this stage pus formation and fistulae occur. An open fistula is in this case usually located posteriorly.

The final result of untreated joint tuberculosis is a permanent loss of mobility (fibrous ankylosis) as well as the development of contractures. This means a flexion/abduction/external rotation contracture in the hip and flexion contracture in the knee, in the ankle and foot a pes equinus valgus position, in the elbow a contracture in 90° of flexion, and in the wrist palmar flexion contrac-

ture. There is often a special type in the shoulder, known as caries sicca (caries is Latin for rotting, sicca is Latin for dry).

Little granulation tissue is formed here, causing a destruction of the joints (primarily of the humeral head) without the formation of abscesses or fistulas.

The diagnosis is difficult to establish at the beginning because of the slow progression. Substantial quadriceps atrophy is very noticeable in knee tuberculosis. The Mantoux test is positive and the ESR and CRP may be elevated.

Supplementary assessment: X-rays and a MRI of the knee, ESR, CRP and Mantoux reaction.

Primary care treatment: tuberculosis should be considered particularly if there is substantial quadriceps atrophy.

When to refer: as quickly as possible.

Secondary care treatment: **tuberculous arthritis.** A joint capsule or bone biopsy are generally carried out to confirm the diagnosis. It can take a few weeks to get the results. However, treatment can already be initiated before the results of the biopsies are known if tuberculosis is strongly suspected. The treatment consists of a combination of tuberculostatic drugs. These are usually administered for 9 months. In more than 50 % of cases the joint will recover fully if the therapy is started early. A synovectomy can be considered for severe synovitis. An arthrodesis or a joint-replacement arthroplasty can be considered if the joint surface is fully destroyed.

N.B.: ! There is multi-resistant tuberculosis in Eastern Europe and Africa.

**Differential Diagnosis: Knee Effusion**

| | |
|---|---|
| **Knee effusion within a few hours after trauma** | |
| Hemarthrosis. | **Capsular tear** |
| Unstable knee. | **Intra-articular fracture** |
| | **Ligamental rupture** |
| **Knee effusion within 24 h after trauma** | |
| Hydrops. | **Distortion** |
| Positive meniscal tear tests. | **Meniscal tear** |
| **Knee effusion without a preceding trauma** | |
| Clear knee aspirate. | **Osteochondrosis dissecans of the femoral condyle** |
| Cloudy knee aspirate (usually several joints are swollen). | **Juvenile idiopathic arthritis (see Table 10.7)** |
| Hemophilia patient. | **Hemophilia bleeding disorder** |
| Brown knee aspirate. | **Pigmented villonodular synovitis** |
| | Focal type |
| | Diffuse type |
| Cloudy knee aspirate with infection elsewhere in the body. | **Reactive arthritis** |
| **Knee effusion which skips to other joints** | |
| Sexually active teenagers. | **Gonococcal arthritis** |
| **Knee effusion accompanied by nocturnal sweating** | |
| Considerable quadriceps atrophy. | |
| Subfebrile temperature. | **Tuberculous arthritis** |

# Knee Pain

## Vague Pain Just Above or Below the Knee

Complaint: the child complains about pain just above or just under the knee. The nature of the pain keeps changing and occurs mainly after doing sports. There may be night pain. The child is not ill and has no fever.

Assessment: no abnormalities are found upon initial assessment of the knee. Sometimes there is slight pressure pain just above or just under the knee.

Differential diagnosis:

**stress fracture**
**subacute osteomyelitis (Brodie abscess)**
**malignant bone tumor**
osteosarcoma
Ewing sarcoma

Explanatory note: **stress fracture.** A stress fracture occurs in persons with normal bone tissue, without a clear preceding trauma. In children a stress fracture occurs usually in the proximal part of the tibia or the distal part of the fibula, but can occur in any bone except the ribs, shoulder blades and pelvis. Complaints tend to start gradually. The child complains of pain in the lower legs on the medial side just below the knee. The pain increases if the leg is bearing weight, and at rest it decreases but does not disappear completely. There is no night pain. Physical assessment reveals a painful spot on pressure on the medial side of the lower leg under the knee, which feels warm, and sometimes there may be slight swelling.

**Subacute osteomyelitis** A subacute osteomyelitis is a cavity filled with pus. Just as in acute osteomyelitis and septic arthritis it is more common in the lower than in the upper extremities and particularly in the proximal and distal part of the tibia and distal part of the femur but also in the talus, calcaneus and in the vertebral column. The abscess cavity generally lies in the metaphysis, but can also lie in the epiphysis or the diaphysis in less fulminant cases of acute hematogenic osteomyelitis. In addition, the abscess can be located in either the metaphysis or the epiphysis, and a part of the epiphyseal plate will have disappeared (Fig. 10.64). The abscess can be centrally or eccentrically located and may cause sclerosis or erosion. There may be new subperiosteal bone formation, resulting in a layered structure if the abscess is located eccentrically in the diaphysis. This layered structure (which on X-rays resembles onion layers) can be confused with a Ewing sarcoma. The most common causes of subacute hematogenic osteomyelitis are Staphylococcus aureus and Staphylococcus epidermidis. The child usually has had 3–4 weeks of pain before he

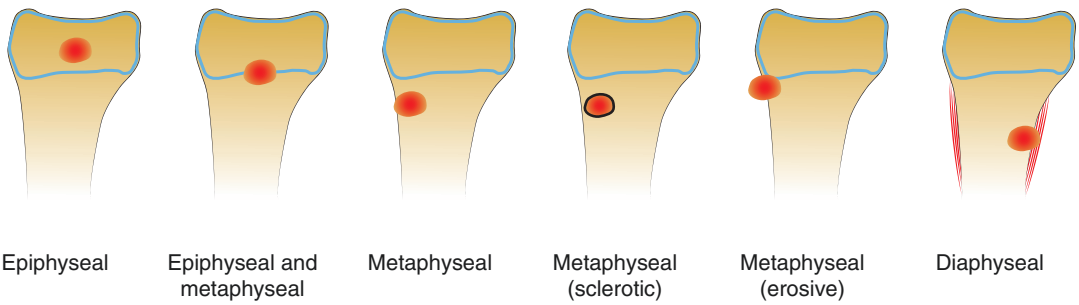



**Fig. 10.64** Types of subacute osteomyelitis (Brodie abscess). The abscess cavity normally lies in the metaphysis, but can also be found in the diaphysis, in the epiphysis, and in both the metaphysis and the epiphysis. It can be located centrally or eccentrically, with or without a sclerotic edge. If the abscess is positioned eccentrically in the diaphysis new bone formation can occur subperiostally, creating a layered structure (Redrawn from: Staheli RT. Fundamentals of pediatric orthopaedics. 4th ed. Philadelphia/Baltimore/New York/London/Buenos Aires/Hongkong/Sydney/Tokyo: Lippincot Williams & Wilkins; 2008. p. 107)

or she is seen by a the doctor. The pain at the location of the abscess tends to intensify gradually, but sometimes diminishes gradually on a periodical basis. The pain may also increase at night. On examination we see a patient who has neither fever nor general disease symptoms. Sometimes a slightly warm swelling can be felt under the skin at the location of the abscess. The function of the adjoining joints is normal. White cell count is usually normal, and the ESR and CRP are elevated in half of all cases. Cultures from the abscess cavity are positive in 60 % of cases.

**Malignant bone tumor** Primary malignant bone tumors are very rare (about 8 in 1 million inhabitants per year). Primary malignant bone tumors often develop around puberty and adolescence. The most frequent location for these tumors is the metaphyseal part of the knee joint. The diagnosis tends to be established relatively late. Sometimes the patient waits several months before consulting the general practitioner. There are few symptoms in the initial stage. The first symptoms are varying pain, especially after sports activities. The pain becomes gradually present day and night. The occurrence of night pain is certainly a reason for further assessment.

One should suspect a primary malignant tumor if there is persistent, atypical pain around the knee. Early swelling usually only occurs if the tumor is located near the surface, for example in an osteochondroma, a periosteal or a paraosseal osteosarcoma, or if the bone is being expanded by a tumor, as is the case in an aneurysmal bone cyst. The occurrence of pathological fractures is the last sign in malignant tumors, and it is often the first sign in benign bone tumors such as unilocular bone cysts. The most common malignant tumors in children are osteosarcoma and Ewing sarcoma (Table 10.8).

**Table 10.8** Tumors around the knee, lower leg and foot. The tumors identified by a § are rare. Giant cell tumors can be benign as well as malignant

| Location | Benign bone tumors | Malignant bone tumors |
|---|---|---|
| Femur | Fibroma | Non-ossifying |
| (distal) | Osteochondroma (exostosis) | Osteosarcoma |
| Epiphysis | Chondroblastoma | |
| | Endochondroma | |
| | Eosinophilic granuloma | |
| | Fibrous dysplasia | |
| | Osteoid osteoma | |
| Tibia | Non-ossifying fibroma | Osteosarcoma |
| (proximal) | Osteochondroma (exostosis) | |
| Tibial shaft | Eosinophilic granuloma | Adamantinoma§ |
| | Fibrous dysplasia | Ewing sarcoma |
| | Neurofibromatosis (Von Recklinghausen disease) | Osteosarcoma |
| | Osteoid osteoma | |
| Foot | Aneurysmal bone cyst | Angiosarcoma§ |
| | Chondroblastoma§ | Ewing sarcoma |
| | Chondromyxoid fibroma§ | Osteosarcoma§ |
| | Endochondroma | |
| | Osteoblastoma | |
| | Osteoid osteoma | |
| | Unilocular bone cyst | |
| | (juvenile/solitary bone cyst) | |

From Adler CP, Kozlowski K. Primary bone tumors and tumorous conditions in children. Heidelberg: Springer Verlag; 1993

*Osteosarcoma* This tumor can occur at all ages. The most common age is between 10 and 30 years, with 25 % of patients being younger than 16. Pain in a malignant bone tumor generally proceeds very slowly but is progressive, and in 80 % of cases is related to activity. Night pain, which affects 20 % of cases, should be an alarm signal and further assessment is required. A palpable swelling occurs in about 40 % of cases, mostly at a later stage. An osteosarcoma can occur in any skeletal part. In 50–60 % of cases the tumor is localized around the knee. Flat bones are involved less often than long tubular bones, but osteosarcoma of the pelvis is not uncommon. In long tubular bones the localization is metaphyseal or metadiaphyseal. The growth plate is involved in 75 % of metaphyseal localizations. The serum alkaline phosphatase is elevated in more than half of the cases.

*Ewing sarcoma* This is a very malignant round cell tumor that often metastasizes. The tumor occurs most commonly between ages 5 and 14, and 95 % of patients are younger than 25.

The most common symptom is pain, in 65 % of cases during activities. Night pain affects 20 % of patients. A considerable soft-tissue swelling that often begins quickly with localized pressure pain and an increase in skin temperature is found in about 35 % of cases. The tumor is localized in the pelvis or in long tubular bones in 75 % of cases. The anomaly is usually located in the long tubular bones in young children under 10 years of age. The most common localizations in older children are the pelvis, shoulder girdle and axial skeleton. The tumor is almost always located in the diaphysis in tubular bones. In about one-third of cases the adjoining metaphysis is also affected.

Supplementary assessment: anteroposterior and lateral X-rays of the knee, CRP, ESR and alkaline phosphatase. A stress fracture does not reveal irregularities radiologically when symptoms begin. A callus is vaguely visible two weeks after symptoms begin (Fig. 10.65). After 3–4 weeks a fracture line is present that can generally be shown only on a CT or MRI. After 8 weeks the fracture has healed.

A subacute osteomyelitis is generally visible as a well-defined cavity (Fig. 10.66). New bone formation is seen periosteally, creating a layered structure if it lies eccentrically in the diaphysis. This layered structure, which on X-rays resembles onion layers, can be confused with a Ewing sarcoma. In subacute hematogenic osteomyelitis the WBC count is usually normal, and the CRP and ESR are elevated in half of the cases. Blood cultures are rarely positive. A technetium scan shows increased activity at the location of the abscess, mostly at an early stage. Characteristics of a malignant tumor on an X-ray can be: an osteolysis, destruction of the cortex, a lamellar subperiosteal reaction (onion peel), a Codman triangle, spiculae, growth outside the periostium and the appearance of a moth-eaten bone (Fig. 10.67). As a rule, it isn't possible to distinguish a malignant from a benign bone tumor with certainty on X-ray. Only in osteochondromata and nonossifying fibromas is radiological diagnosis sufficient.

In a Ewing sarcoma, the ESR is usually elevated. Alkaline phosphatase can be elevated in osteosarcomas and in Ewing sarcomas. This is of limited significance because during growth, especially in adolescence, alkaline phosphatase is always elevated.

Primary care treatment: the possibility of a stress fracture, acute hematogenic osteomyelitis or a primary malignant bone tumor should always be considered if there is vague pain.

When to refer: if a bone lesion is seen on the X-rays.

Secondary care treatment: **stress fracture.** A stress fracture in the proximal part of the tibia is treated with a plaster cast immobilization for 6 weeks. Normal weight bearing is possible.

**Subacute hematogenic osteomyelitis** A subacute osteomyelitis is treated with antibiotics until the CRP and the ESR levels are normal (usually 4–6 weeks). This is sufficient in 90 % of

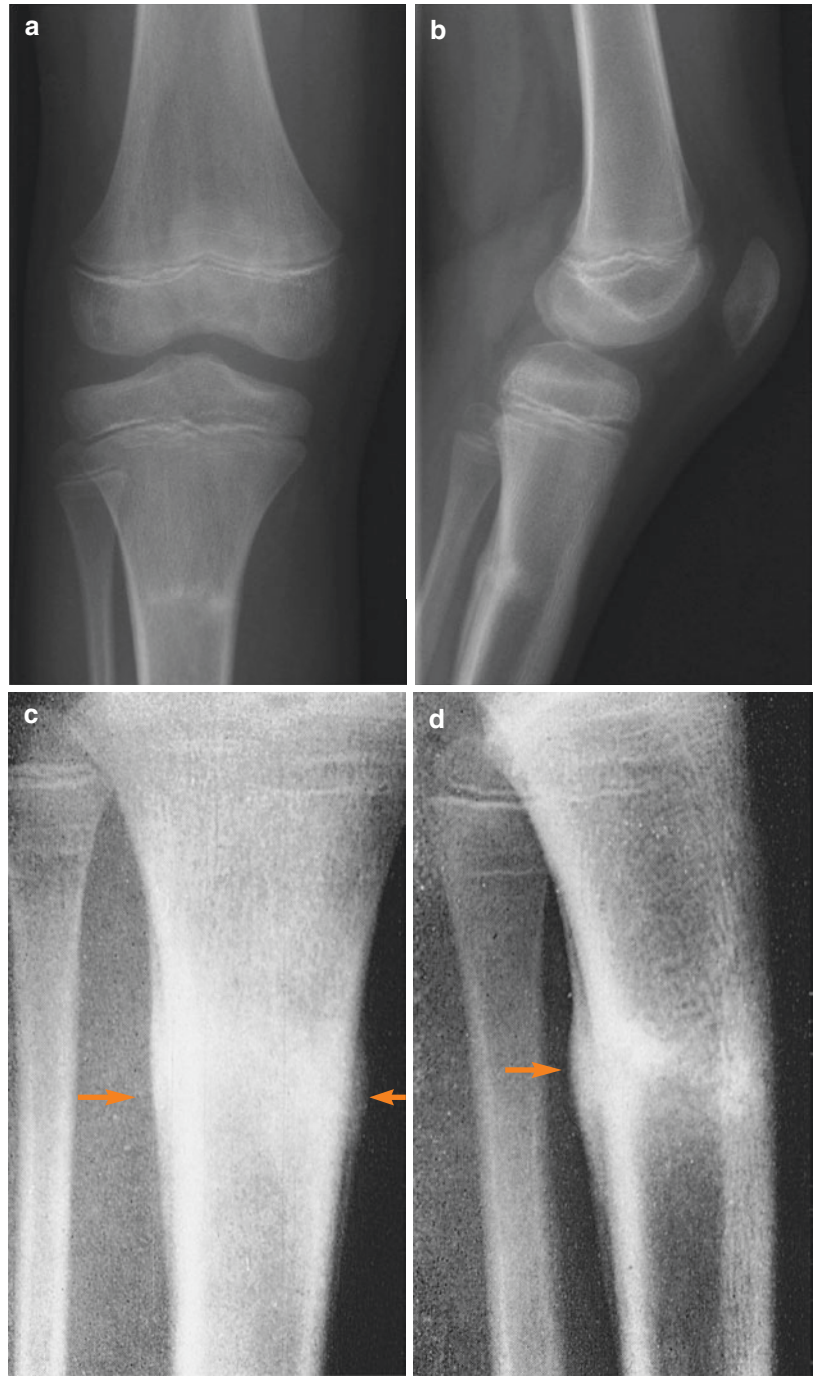



**Fig. 10.65** (**a**) Anteroposterior and (**b**) lateral X-ray of the proximal part of the right lower leg. A tibial stress fracture can hardly be recognized. (**c**) Anteroposterior and (**d**) lateral X-ray of the proximal part of the right lower leg. The tibial stress fracture healed after 8 weeks. Callus formation is visible (*arrows*)

cases. The cavity is curetted if the abscess does not react to antibiotics. In the case of epiphyseal involvement growth will be undisturbed if the abscess has affected less than one-third of the growth plate. A CT-scan or an MRI should be requested in case of doubt, or if there isn't a malignant bone tumor after all.

**Malignant bone tumor** Treatment of primary malignant tumors takes place in a center

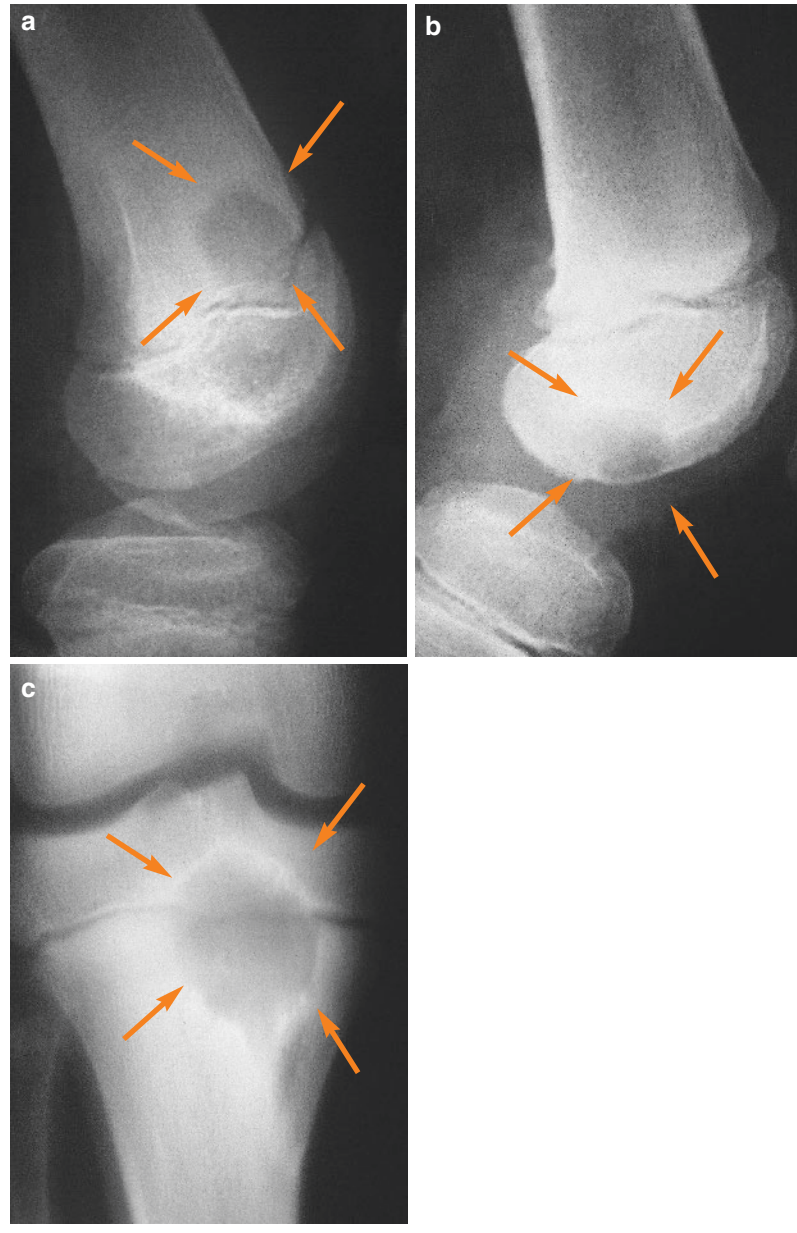



**Fig. 10.66** (**a**, **b**) Lateral X-rays, and (**c**) antero-posterior tomogram of a knee with subacute osteomyelitis. (**a**) Positioned in the distal femoral metaphysis. (**b**) Located in the distal femoral epiphysis. (**c**) Positioned both in the metaphysis and the epiphysis in the proximal part of the tibia

specialized in bone tumors. A bone biopsy has to be performed to establish the diagnosis. The biopsy, which may be carried out using CT technology, should be done by an orthopedic surgeon specialized in bone tumors. An open biopsy is usually recommended in order to obtain sufficient material.

*Osteosarcoma* The treatment consists of a combination of chemotherapy and surgery. An amputation or an exarticulation takes place or the tumor is radically resected; this is followed by a Van Nes rotation plasty or reconstruction of the bone defect by means of an endoprosthesis or a fibular transplant (Fig. 10.67). Prognosis: the chances of metastases are considerable. The process is hematogenic or lymphogenic, spreading to the lungs or other parts of the skeleton. Intramedullary metastases can also appear, known as ‘skip lesions’. The 5-year survival rate

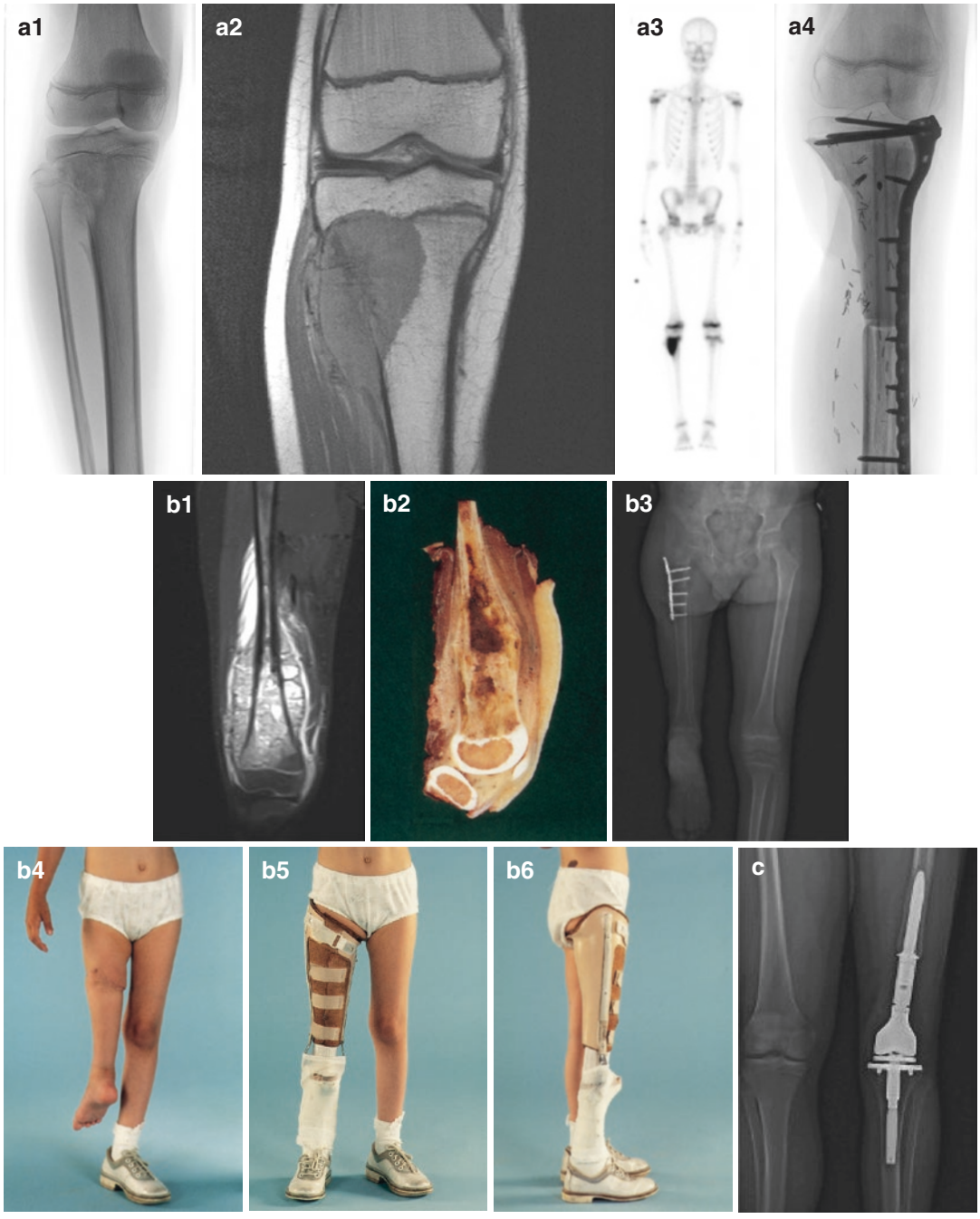



**Fig. 10.67** (**a1**) Osteosarcoma on the lateral side in the proximal part of the tibia. Characteristic is the osteolysis with destruction of the cortex, subperiostal reaction (onion peel), Codman triangle and a motheaten-away part. (**a2**) MRI image. (**a3**) Bone-scan. (**a4**) Resection of the tumor, vascularized fibula allograft to bridge the defect supported with a plate osteosynthesis. (**b1**) MRI with osteosarcoma in the distal part of the right femur. (**b2**) Sagittal cross-cut of the pathological anatomical preparation. (**b3**–**b5**) After resection of the tumor a Van Nes rotation plasty is carried out. In this procedure the leg is rotated through180° and fixed after resection of the tumor. The foot is now pointing backwards. Next, a prosthesis can be given to make the ankle joint function as a knee joint. (**c**) Implant after removal of a tumor in the distal part of the femur

for osteosarcoma after treatment lies between 70 and 80 %. Sometimes lung metastases can still be removed. The 5-year survival chances are 10–20 % after removal of a lung metastasis. The prognosis in the case of bone metastases is very poor.

*Ewing sarcoma* Chemotherapy followed by radical resection and, if possible, followed by radiotherapy. The 5-year survival chances lie around 50 %. There are metastases at the moment of diagnosis in 15–30 % of cases. Metastases spread equally to the lungs and to other bones.

## Pain in the Inner or Outer Side of the Knee

Complaint: pain in the medial or lateral side of the knee.

Assessment: there is pressure pain on the anterior side of the medial or lateral femoral condyle in a maximally flexed knee, or the meniscal tear tests are positive (Figs. 10.42, 10.43, 10.44, 10.45, 10.46, and 10.47).

Differential diagnosis:

**osteochondritis dissecans of the femoral condyle**
**discoid meniscus**
**meniscal tear**

Explanatory note: these three diagnoses are usually not accompanied by an extension limitation or a hydrops in the knee. Most of the time there are only pain periods on the inner or outer side of the knee.

For explanatory note, supplementary assessment, primary care treatment, when to refer and secondary care treatment (see pp. 200–205).

## Pain at the Front of the Knee

Complaint: pain symptoms at the front of the knee occur mostly when standing up after sitting for a long time with flexed knees, cycling into the wind, walking, and sporting activities involving a lot of jumping, such as in volleyball and basketball.

Assessment: pressure pain around the upper-outer side of the patella. Pressure pain around the medial patellar facet. Pressure pain around the lateral patellar facet. Pressure pain around the lower pole of the patella. Swelling and pressure pain around the tibial tuberosity (Fig. 10.68).

Differential diagnosis:

**bipartite and multipartite patella**
**patellofemoral pain syndrome**
**osteochondritis dissecans of the patellofemoral joint**
**lateral patellofemoral compression syndrome**
**Sinding-Larsen-Johansson disease**
**Osgood-Schlatter disease**

Explanatory note: **bipartite and multipartite patella.** There is generally a rounded bony fragment on the upperouter side of the patella in bipartite patella (Figs. 10.69 and 10.70).

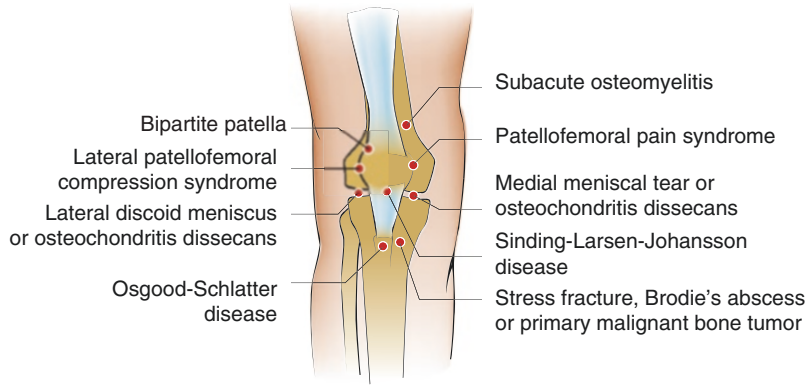



**Fig. 10.68** Pressure-pain points on the anterior side of the right knee in children

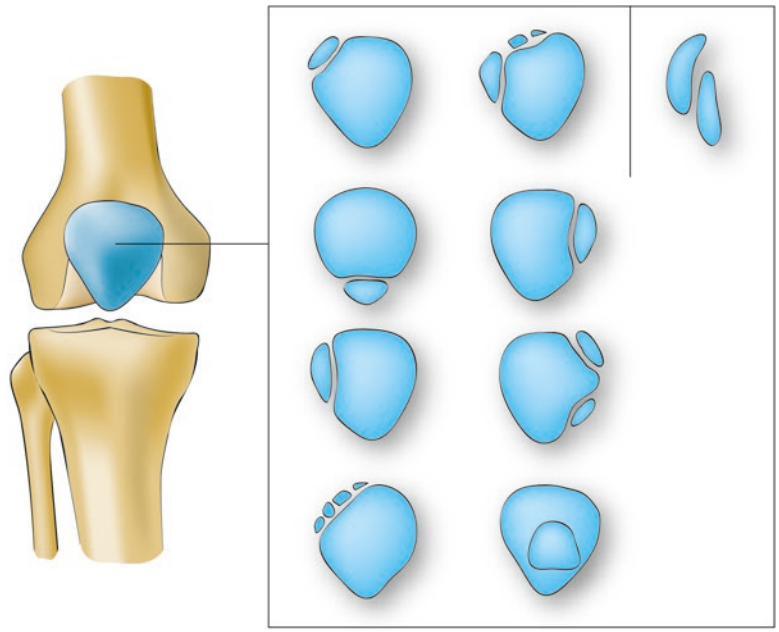


**Fig. 10.69** In 1–3 % of cases the patella is not made up of one single bone but of two or more parts and is known as bipartite or multipartite patella. The most common form is a bipartite patella, which has a rounded bone fragment on the upper-outer side of the patella (*left-upper figure in the frame*). The uppermost figure on the right in the frame is a lateral representation, the others are anteroposterior representations of the patella

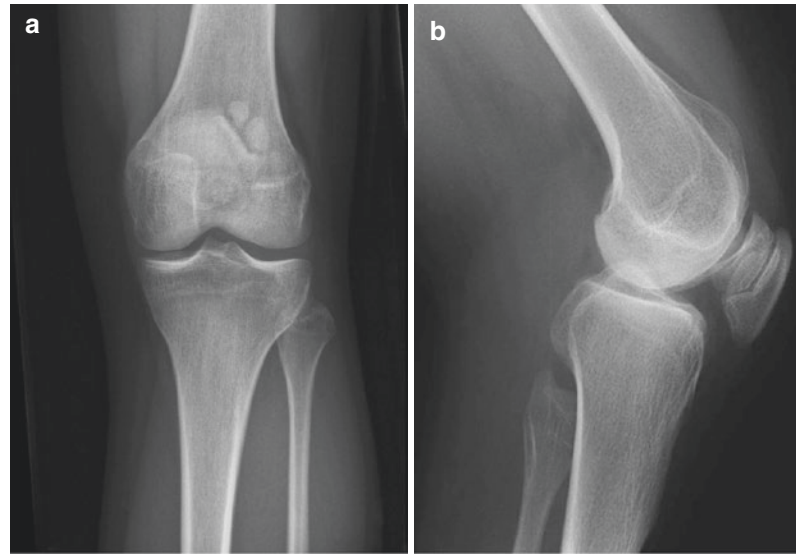



**Fig. 10.70** (**a**) Anteroposterior X-ray of the left knee with a tripartite patella. (**b**) Lateral X-ray of the knee with the rarest form of bipartite patella, which shows fragmentation in the sagittal plane

This type tends to be bilateral. The fragment is fibrous or cartilaginous and connected with the rest of the patella. Bipartite patella is present in 1–3 % of children. Incidence between girls and boys is 9:1. Complaints caused by this are rare in children. Only 2 % is symptomatic. Sometimes a bipartite patella is incorrectly identified as a fracture.

**Patellofemoral pain syndrome** The terms chondropathy and chondromalacia of the patella are often used for symptoms in a patellofemoral pain syndrome. These terms suggest cartilage disease of the patella. This cannot always be confirmed in children in most cases and it is better to avoid these terms. The cause of the symptoms is not clear. Symptoms mostly affect children

between the age of 13 and 15. The symptoms are sometimes so severe that sporting acitvities become impossible and the child can only walk with crutches. There may also be a lock phenomenon. The patellofemoral pain syndrome causes pressure pain around the medial patellar facet (Fig. 10.71), and the patellar grind test or Clarke sign is positive. In the patellar grind test the examiner places the first web space of his hand just superior to the patella while applying pressure on the patella with the other hand. The patient is instructed to gently and gradually contract the quadriceps muscle. A positive sign on this test is pain in the patellofemoral joint.

Pain is usually initiated while ascending or descending stairs or slopes, squatting, kneeling, cycling, running or prolonged sitting with bent knees. The latter feature is sometimes termed the "movie sign" or" theatre sign" because one might experience pain while sitting to watch a film or play. The symptoms can persist for several years but in most cases disappear spontaneously. In isolated cases the symptoms can be caused by an osteochondritis dissecans of the patella. Sometimes patellofemoral symptoms are the initial signs of an osteochondritisdissecans of the medial or lateral femoral condyle (see pp. 201, 204 and 205).

**Osteochondritis dissecans of the patellofemoral joint** The knee is mostly affected. In 85 % of cases the anomaly is localized at the level of the medial femoral condyl, in about 13 % of the cases at the level of the lateral femoral condyl, and in a few percent around the patellofemoral joint (see pp. 201).

The symptoms are the same as in a patellofemoral pain syndrome.

**Lateral patellofemoral compression syndrome** The pain in this case is lateral and not medial as in the patellofemoral pain syndrome. There is pressure pain around the lateral patellar facet and the patella does not fit neatly into the femoral groove, but supports the lateral patellar facet rather unevenly and forcefully on the lateral femoral condyle.

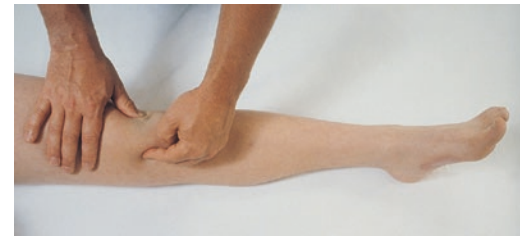


**Fig. 10.71** In patellofemoral pain syndrome there is pressure pain around the medial patellar facet. The examiner presses the patella in medial direction with one hand and palpates the posterior edge of the patella on the medial side with the index finger of the other hand

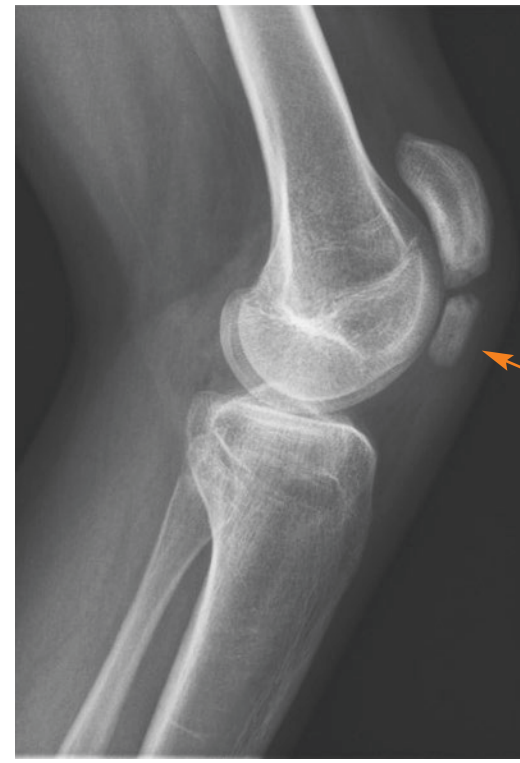


**Fig. 10.72** Sinding-Larsen-Johansson disease. On the X-ray we see calcifications at the apex of the patella (*arrow*)

**Sinding-Larsen-Johansson disease** This is an overloading syndrome around the attachment of the patellar ligament at the patella apex. On X-rays we see a bone fragment or fragments around the apex of the patella (Fig. 10.72). Symptoms usually appear between the ages of 10 and 15.

**Osgood-Schlatter disease** In all cases there is a painful swelling around the tibial tuberosity

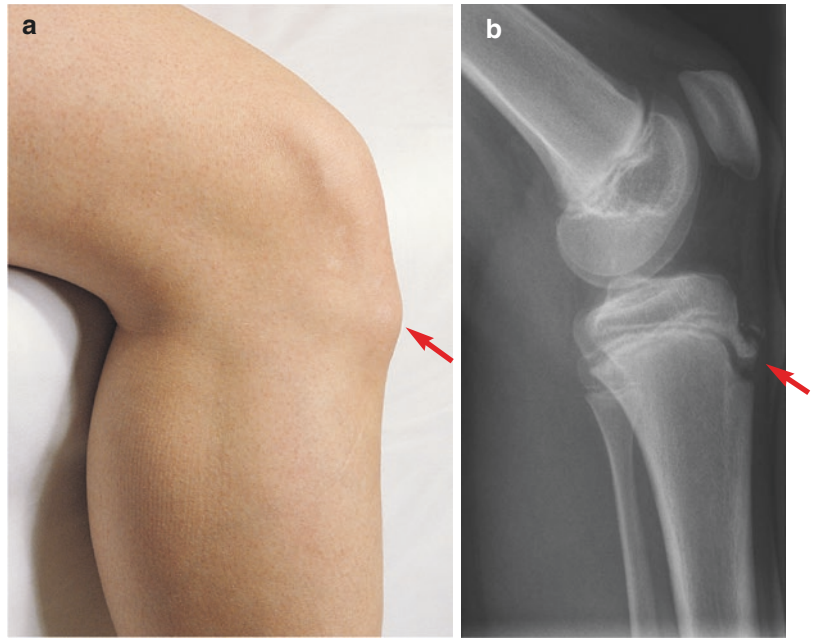



**Fig. 10.73** Osgood-Schlatter disease. (**a**) Swelling around the tibial tuberosity (*arrow*). (**b**) Ossification disorder of the apophysis of the tibial tuberosity (*arrow*)

(Fig. 10.73). The pain is at its most severe when the patient kneels. The pain is a result of overloading at the location of the attachment of the patellar ligament to the tibial tuberosity, which causes an ossification disorder of the apophysis. Symptoms begin between the ages of 10 and 15, usually in boys. The symptoms are bilateral in 30 % of cases. The pain tends to last for 12–18 months and then disappears spontaneously. The swelling always remains, even after the pain has disappeared. It can persist in about 5 % of cases as a result of a loose bone fragment at the level of the tibial tuberosity. A premature closure of the anterior part of the growth plate in the proximal part of the tibia is rare but does cause a recurvatum deformity.

Supplementary assessment: radiological assessment is only indicated for persistent pain on the anterior side of the knee. An X-ray of the patellofemoral joint may be requested in addition to anteroposterior and lateral X-rays of the knee joint. No radiologically anomalies are seen in the patellofemoral pain syndrome.

In a bipartite patella there is usually a rounded fragment on the upper-outer side of the knee. Sometimes there are several fragments or fragmentation distally all on the medial side. There may even be fragmentation in the sagittal plane. In this case this type of bipartite patella can be best seen on lateral X-rays of the knee.

An axial X-ray of the patellofemoral joint can help detect osteochondritis dissecans lesions of the patella: an axial X-ray of the patella shows a joint gap of the patellofemoral joint that is narrower on the lateral side than the medial side in the case of the lateral patellofemoral compression syndrome.

We see a bone fragment or fragments around the apex of the patella in the Sinding-Larsen-Johansson disease.

X-rays sometimes show, in addition to a soft tissue swelling, a loose bone fragment around the apophysis of the tibial tuberosity in Osgood-Schlatter disease.

Primary care treatment: the symptoms of the previous mentioned conditions can be treated by the general practioner. First of all lifestyle rules are prescribed, such as not sitting with flexed knees for a prolonged period of time, avoiding cycling into the wind as much as possible, and climbing stairs. Sporting activities that incite pain

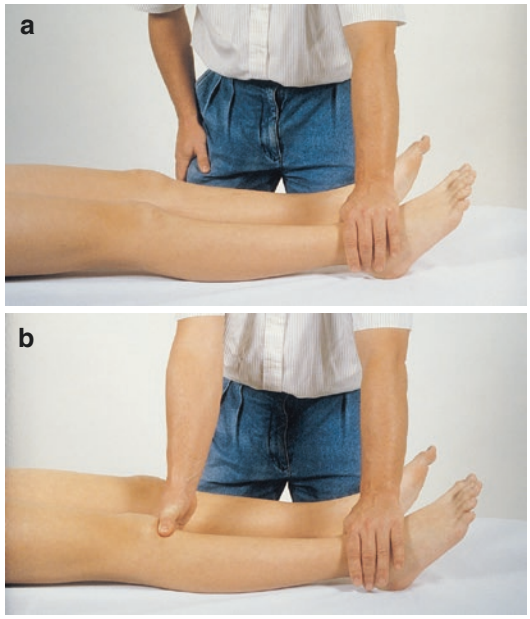



**Fig. 10.74** (**a**) Patellar tendon test. The child lies supine. In the first part of the patellar tendon test the patient must lift the extended leg while the examiner applies counterpressure on the lower leg. A child with patellar pain syndrome will generally identify pain on the anterior side of the knee. (**b**) In the second part of the patellar tendon test the examiner also presses on the patellar tendon just below the patella with the thumb of the other hand. A good indication for a patellar tendon bandage or orthesis is when the child has little or no sign of pain

should be less intense or discontinued. It goes without saying that these rules do not have to be followed strictly. The child cannot "mess up" the knee, there will be no damage to the knee, but there will be pain. The symptoms almost always disappear spontaneously in the long run. The knee can be taped or a patellar tendon bandage may be prescribed if it isn't possible to keep the complaints within bounds by following the lifestyle rules, the patellar tendon test is the best method to assess the situation (Fig. 10.74). The child lies on the back. Counterpressure on the lower leg is applied while the child lifts the extended leg. A child with patellofemoral pain syndrome and lateral patellofemoral compression syndrome will tend to have pain on the anterior side of the knee, a child with Sinding-Larsen-Johansson disease will have pain around the lower pole, and a child with Osgood-Schlatter disease will have pain around the tibial tuberosity. Then the test is repeated, pressing the thumb of the other hand on the patellar ligament between the apex of the patella and tibial tuberosity. There is a good indication for a tape or patellar tendon bandage to reduce symptoms if the child has no pain or less pain (Fig. 10.75). An alternative for a patellar tendon bandage is a Hamburger patellar orthesis (Fig. 10.75) or a Q-brace (Fig. 10.76). A decrease in the symptoms using tape or ortheses is achieved in 50–60 % of cases.

When to refer: if the symptoms on the anterior side have been present for more than a year, despite lifestyle rules and any treatment using a patellar tendon bandage or a patellar orthesis.

Secondary care treatment: **Bipartite patella.** Small fragments are removed and in large fragments a tension band wire is applied. The success rate of these operations is unknown.

**Patellofemoral pain syndrome** There is no adequate operative treatment available for this syndrome. There is no indication for arthroscopy. A loose bone fragment can be operatively removed if there is an osteochondritis dissecans of the patellofemoral joint.

**Osteochondritis dissecans of the patellofemoral joint** Operation is indicated for persistent pain, intra-articular loose bodies and subchondral sclerosis. Excision of the fragment and curettage of the crater, with or without drilling, is recommended.

**Lateral patellofemoral compression syndrome** If there is a stubborn lateral patellofemoral compression syndrome, one may consider cleaving the lateral retinaculum lengthwise ("lateral release").

**Sinding-Larsen-Johansson disease** One may consider operatively removing the bone fragments around the apex of the knee in cases of persistent Sinding-Larsen-Johansson disease.

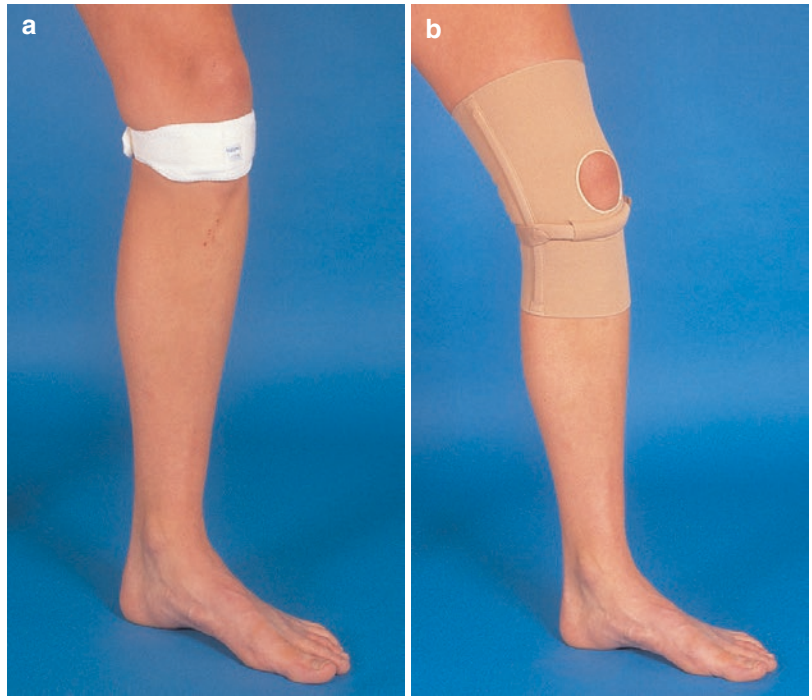



**Fig. 10.75** (**a**) A patellar tendon bandage is worn just under the patella. (**b**) Hamburger patellar orthesis. This orthesis has the same function as a patellar tendon bandage but there is a reduced chance of venous engorgement. The patella is left free and reinforcement is provided between the kneecap and the tibial tuberosity

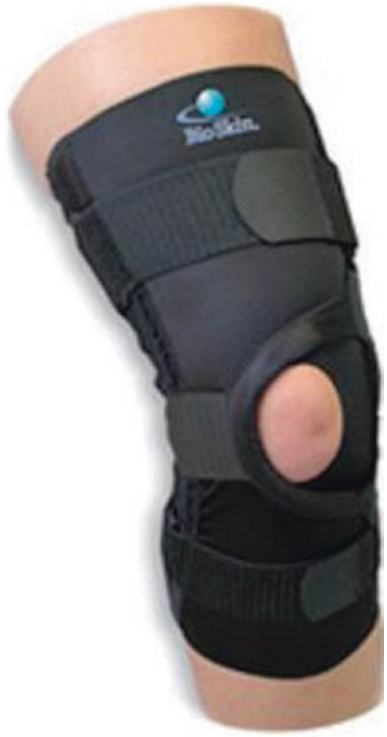


**Fig. 10.76** Q-brace. Patella-orthesis with displaceable support

**Osgood-Schlatter disease** In adult cases that present with a rounded bone fragment around the attachment of the patellar tendon with the tibial tuberosity accompanied by complaints, especially when kneeling and crawling one can consider removing the loose bone fragment operatively.

## Thigh and Knee Pain

Complaint: pain around the upper leg and/or the knee.

Assessment: the child has a hip-unburding gait pattern (see addendum). The child lies prone with a knee flexed to 90° and there is a painful and/or limited internal rotation of the hip.

Differential diagnosis:

**coxitis fugax**
**Legg-Calvé-Perthes disease (Perthes disease, coxa plana, Waldenström disease)**
**chronic epiphysiolysis capitis femoris**

Explanatory note: hip anomalies, especially in young children, can cause pain in the upper leg or even in the knee. The problem is not an anomaly in the knee itself but an anomaly in the hip in 5 % of knee complaints in children. The first movement that will become limited in the hip joint is internal rotation. The anomaly is very probably located in the hip joint and not in the knee joint if there is limited and/or painful hip joint internal rotation.

For explanatory note, supplementary assessment, primary care treatment, when to refer and secondary care treatment, see Chap. 9.

## Extremely Painful Immobile Knee

Complaint: the parents have noticed that their child younger than 18 months no longer moves the leg in question. The child is not ill and has no fever. In older children: the child is ill and has a fever; refuses to use the affected leg and will certainly not stand or walk on it. There is pain around the knee, or just proximal or distal to the joint.

Assessment: a newborn obviously cannot indicate where the pain is localized; but one does notice that the involved extremity is not being moved. The knee tends to be held in light flexion (Bonnet position). There is already redness in an early stage. On examination the knee or the area proximal or distal to the knee will be warmer than the surroundings. There is also severe local pressure pain. An older child will also be febrile.

Differential diagnosis:

**acute osteomyelitis**
**septic arthritis**

For explanatory note, supplementary assessment, primary care treatment, when to refer and secondary care treatment, see Chap. 18.

### Differential Diagnosis: Knee Pain

| | |
|---|---|
| **Vague pain just above or below the knee** | |
| Pressure pain on the medial side under the knee. | **Stress fracture** |
| Pressure pain just above or below the knee. | **Subacute osteomyelitis (Brodie abscess)** |
| | **Malignant bone tumor** |
| | Osteosarcoma |
| | Ewing sarcoma |
| **Pain in the inner or outer side of the knee** | |
| Pressure pain on the anterior side of the medial femoral condyle in a maximally bent knee. | **Osteochondritis dissecans of the medial femoral condyle** |
| Pressure pain on the anterior side of the lateral femoral condyle in a maximally bent knee. | **Osteochondritis dissecans of the lateral femoral condyle** |
| Pressure pain at the lateral joint line. | **Discoid lateral meniscus** |
| Positive medial meniscus tear tests. | **Medial meniscal tear** |
| Positive lateral meniscus tear tests. | **Lateral meniscal tear** |
| **Pain at the front of the knee** | |
| Pressure pain on the upper-outer side of the patella. | **Bipartite and multipartite patella** |
| Pressure pain on the medial patellar facet. | **Patellofemoral pain syndrome** |
| Lock phenomenon. | **Osteochondritis dissecans of the patellofemoral joint** |
| Pressure pain on the lateral patellar facet. | **Lateral patellofemoral compression syndrome** |
| Pressure pain at the lower pole of the patella. | **Sinding-Larsen-Johansson disease** |
| Pressure pain at the tibial tuberosity. | **Osgood-Schlatter disease** |
| **Thigh and knee pain** | |
| Internal rotation limitation of the hip for several days in a child between ages 2 and 12. | **Coxitis fugax** |
| Internal rotation limitation of the hip for a prolonged period of time in a child between ages 2 and 12. | **Legg-Calvé-Perthes disease (Perthes disease, coxa plana, Waldenström disease)** |
| Internal rotation limitation of the hip for a prolonged period of time in a child between ages 10 and 16. | **Chronic slipped capital femoral epiphysis** |
| **Extremely painful immobile knee** | |
| The knee is severely painful and is no longer mobile. | **Acute osteomyelitis** |
| | **Septic arthritis** |

# Lower Leg

# 11

## Lower Leg Misalignment

### Forwards and Outwards Lower Leg Bowing

Complaint: the parents come with their newborn child because the lower leg is bowed anteriorly and laterally.

Assessment: there is an anterolateral lower leg bowing (Fig. 11.1).

Differential diagnosis:

**congenital anterolateral tibial and possibly fibular bowing**
**camp(t)omelia dysplasia (Cumming dysplasia syndrome)**
**congenital tibial pseudarthrosis**
type I
type II
type III
type IV
type V
**fibrous dysplasia of the tibia**

Explanatory note: **congenital anterolateral tibial and possibly fibular bowing.** This is a rare deformity in which the tibia and possibly the fibula are bowed anterolaterally. On X-ray's the cortex of the central part of the tibia is thickened particularly on the medial side. Therefore the bone marrow cavity is narrowed (Fig. 11.2). The deformity improves with time but never completely.

**Camp(t)omelic dysplasia**[1] The deformity is often described without the "t" (campomelic dysplasia). Both femurs and tibias with fibulas are bowed anterolaterally (Fig. 11.3). There are tibial cutaneous dimples and an equinous or clubfoot. Furthermore the patient is conspicuous because of a big head, low implanted ears, micrognathia and a small and flat face.

**Congenital tibial pseudarthrosis** There is an anterolateral tibial bowing at birth. Only in a small minority of cases there is already a pseudarthrosis at birth. However in the vast majority this occurs later on. The incidence is 1 in 200,000 births. Bilateral cases are even more exceptional. The cause is unknown. In three quarters of the cases we are dealing with a neurofibromatosis (Von Recklinghausen disease[1]). The periosteum is thickened and covered with a thick layer of soft tissue at the level of the pseudarthrosis. Five types can be classified (Fig. 11.4):

Type I. Simple congenital tibial pseudarthrosis: the lower leg is straight and at best slightly short. There is no narrowing in the diameter of the tibula and fibula on X-ray. After a minimal trauma a midshaft stress fracture of the tibia occurs which finally ends up in a pseudarthrosis (Fig. 11.5). This generally occurs after 5 years of age. The pseudarthrosis is often

[1] See Appendix.


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_11

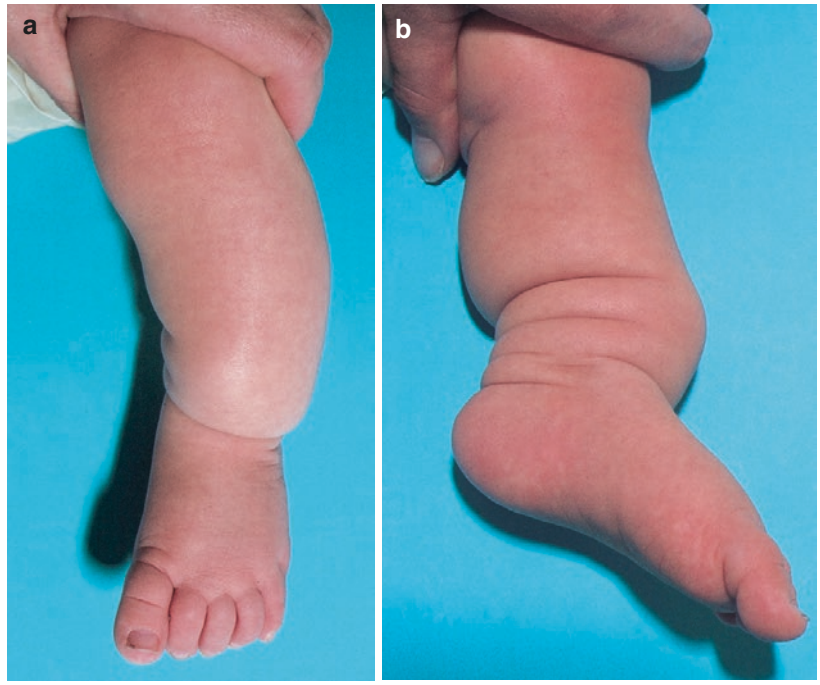



**Fig. 11.1** Congenital anterolateral bowing of the left lower leg. (**a**) Left lower leg from in front and (**b**) viewed from the side

called the late type. A neurofibromatosis (Von Recklinghausen disease[2]) is not present.

Type II. Cystic congenital tibial pseudarthrosis: in this type there is also no narrowing in the diameter of the tibia and fibula. Neurofibromatosis is not present. In the area of transition between the central and distal third of the tibia cystic abnormalities are present in the tibia and also occasionally in the fibula (Fig. 11.6). This appearance looks like a fibrous dysplasia. The lower leg is not bowed at birth. Angulation occurs during the first months after birth.

Type III. Sclerotic congenital tibial pseudarthrosis: the pseudarthrosis occurs in a sclerotic segment. The bone marrow cavity is completely or partially obliterated. A stress fracture occurs in the cortex at the level of a sclerotic fragment which partially extends into the sclerotic bone (Fig. 11.7). There may be a neurofibromatosis in this type.

Type IV. Dysplastic congenital tibial pseudarthrosis without fibular pseudarthrosis: in this case there is an intensification of the sclerosis with complete obliteration of the bone marrow cavity. The hourglass shaped tibia is bowed anteriorly and laterally (Fig. 11.8). The pseudarthrosis can be present at birth, but generally occurs after the child starts to walk. In this type a neurofibromatosis is almost always present.

Type V. Dysplastic congenital tibial and fibular pseudarthrosis. This is considered to be the most difficult type to treat. There is almost always a neurofibromatosis in these cases.

**Fibrous dysplasia of the tibia** In fibrous dysplasia one or more fibrous tissue foci are found. There is a monostotic and a polyostotic type. The most common locations are in the ribs, tibia, femur and mandible. The abnormality is usually discoverd just before puberty (Fig. 11.9).

Supplementary assessment: anteroposterior and lateral X-rays of both legs.

Primary care treatment: none.

When to refer: if there is an anterolateral bowing of the leg or if there is a stress fracture present in a straight leg.

Secondary care treatment: **congenital anterolateral tibial and possibly fibular bowing.** A wait and see policy will be adopted in the case

[2]See Appendix.

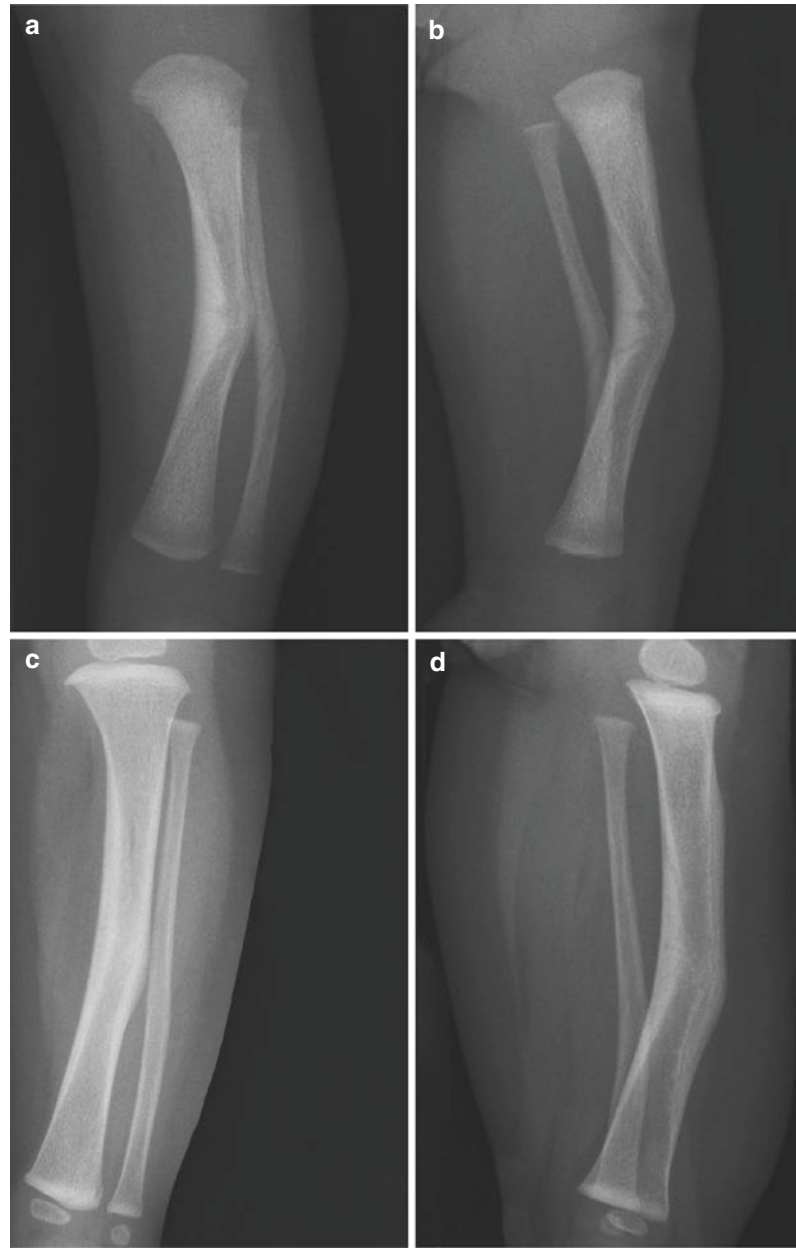



**Fig. 11.2** (**a**) Anteroposterior and (**b**) lateral X-ray of a congenital anterolateral tibial bowing in a 4 weeks old child. On the anteroposterior X-ray the cortex in the central area of the tibia is thickened particularly on the medial side and the bone marrow cavity is narrowed. (**c**, **d**) Situation at 2½ years of age. A spontaneous improvement has occurred

of a congenital anterolateral bowing. If the remaining deformity is more than 10° a correction osteotomy may be considered after the age of 4.

**Camp(t)omelic dysplasia**[3] The prognosis is often poor. They often die shortly after birth as a result of aspiration pneumonias. A correction of the bowed upper and lower leg and the clubfeet may be considered if the child survives.

**Congenital tibial pseudarthrosis** An ankle-foot orthosis (AFO) may be prescribed during the prepseudarthrosis period if there is no evidence of a fracture. In the case of a pseudarthrosis the bowing can be corrected in young children with fixation of the tibia with an intramedulary rod

[3]See Appendix.

(Fig. 11.10), and a spongy autogenous bone graft will be applied at the level of the pseudarthrosis. If consolidation does not occur a bone transport technique using Ilizarov fixation can be carried out after about 6 years of age. The pseudarthrosis will be widely resected. The tibia will be divided proximally and transported distally. The defect which occurs proximally will be filled up with a good quality autogenous bonegraft. During this procedure any axial deformity will also be corrected. The defect can also be corrected with a vascularised fibulatransplant from the normal side (Fig. 11.11). Success is achieved using these methods in about 90 % of cases. Type I, II and III have a better prognosis than type IV and V. Sometimes even after several procedures the pseudarthrosis cannot be corrected and finally even a lower leg amputation may be considered.

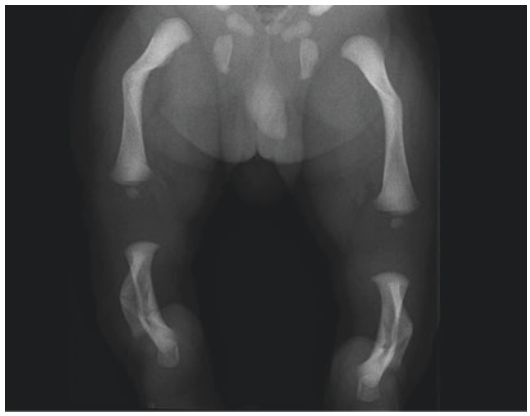


**Fig. 11.3** Camp(t)omeliac dysplasia. Anteroposterior X-ray of both legs. Anterolateral bowing of both femurs and lower legs with equinus feet in a newborn child

**Fibrous dysplasia of the tibia** Corrective osteotomy with curettage can be carried out filling the cavity up with a bone graft or calcium phosphate chips.

## Backwards and Inwards Lower Leg Bowing

Complaint: the lower leg is bowed posteriorly and medially. The foot is in hyperextension and sometimes to such an extent that the

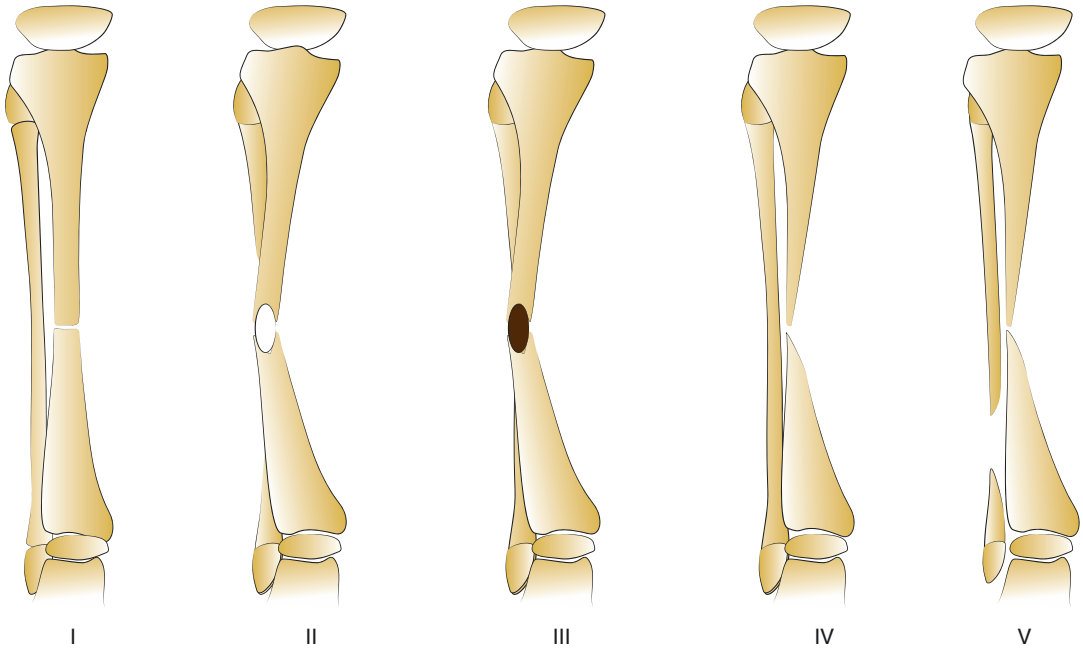



**Fig. 11.4** Classification congenital tibial pseudarthrosis. Type *I*: simple. Type *II*: cystic. Type *III*: sclerotic. Type *IV*: dysplastic. Type *V*: dysplastic with pseudarthrosis of the fibula (Redrawn from: Staheli RT. Fundamentals of pediatric orthopaedics. 4th ed. Philadelphia/Baltimore/New York/London/Buenos Aires/Hongkong/Sydney/Tokyo: Lippincot Williams & Wilkins; 2008)

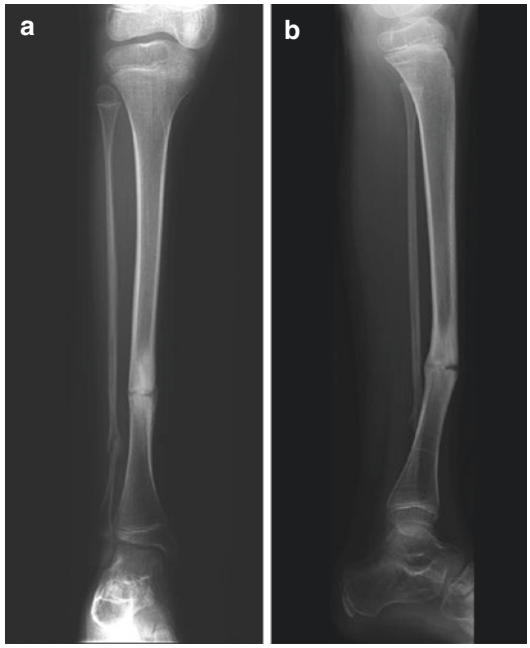



**Fig. 11.5** (**a**) Antroposterior and (**b**) Lateral X-ray of simple congenital tibial pseudarthrosis (type I) in a 10 year old boy

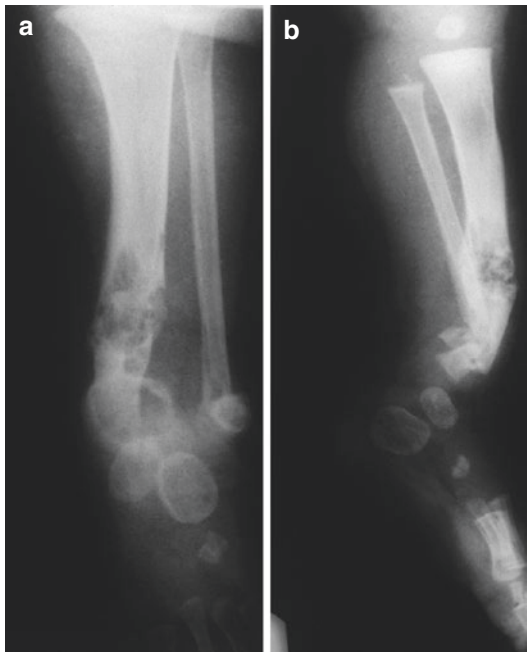



**Fig. 11.6** (**a**) Anteroposterior and (**b**) Lateral X-ray of a cystic congenital tibial pseudarthrosis (type II) in a 2 month old child

dorsum of the foot lies against the front side of the lower leg.

Assessment: the lower leg in the area between the central third and distal third is bowed posteriorly (in recurvation) and medially (valgus) (Fig. 11.12). The foot is in dorsiflexion. Plantar flexion of the foot is limited, but can usually be passively moved to a neutral position between dorsal extension and plantar flexion. It is usually a unilateral deformity and the involved leg is usually 1 cm less in circumference compared to the normal leg. At birth there is a leg length difference of at least 1 cm on the involved side.

Diagnosis: **congenital posteromedial tibial and fibular bowing.**

Explanatory note: **congenital posteromedial tibial and fibular bowing.** The cause is unknown. The degree of bowing, medially and posteriorly is finally usually the same and varies between 25° and 65°. The difference in leg length will be greater depending on the rate of growth. This leg length difference is caused by slow growth in the growth plate of the distal part of the tibia. On average there is a leg length difference of 4 cm at the end of the growth period (varying from 3 to 7 cm). During growth the bowing of the tibia and fibula disminishes and this occurs more quickly in the posterior component than in the medial component (Fig. 11.12). This spontaneous correction is greatest during the first 6 months after birth. At 2 years of age about half of the misalignment as corrected spontaneously. Correction is slower after the age of 3. Correction of the dorsiflexion deformity of the foot will be spontaneous during this period.

Supplementary assessment: anteroposterior and lateral X-rays of the lower leg.

Primary care treatment: none.

When to refer: as soon as possible after birth.

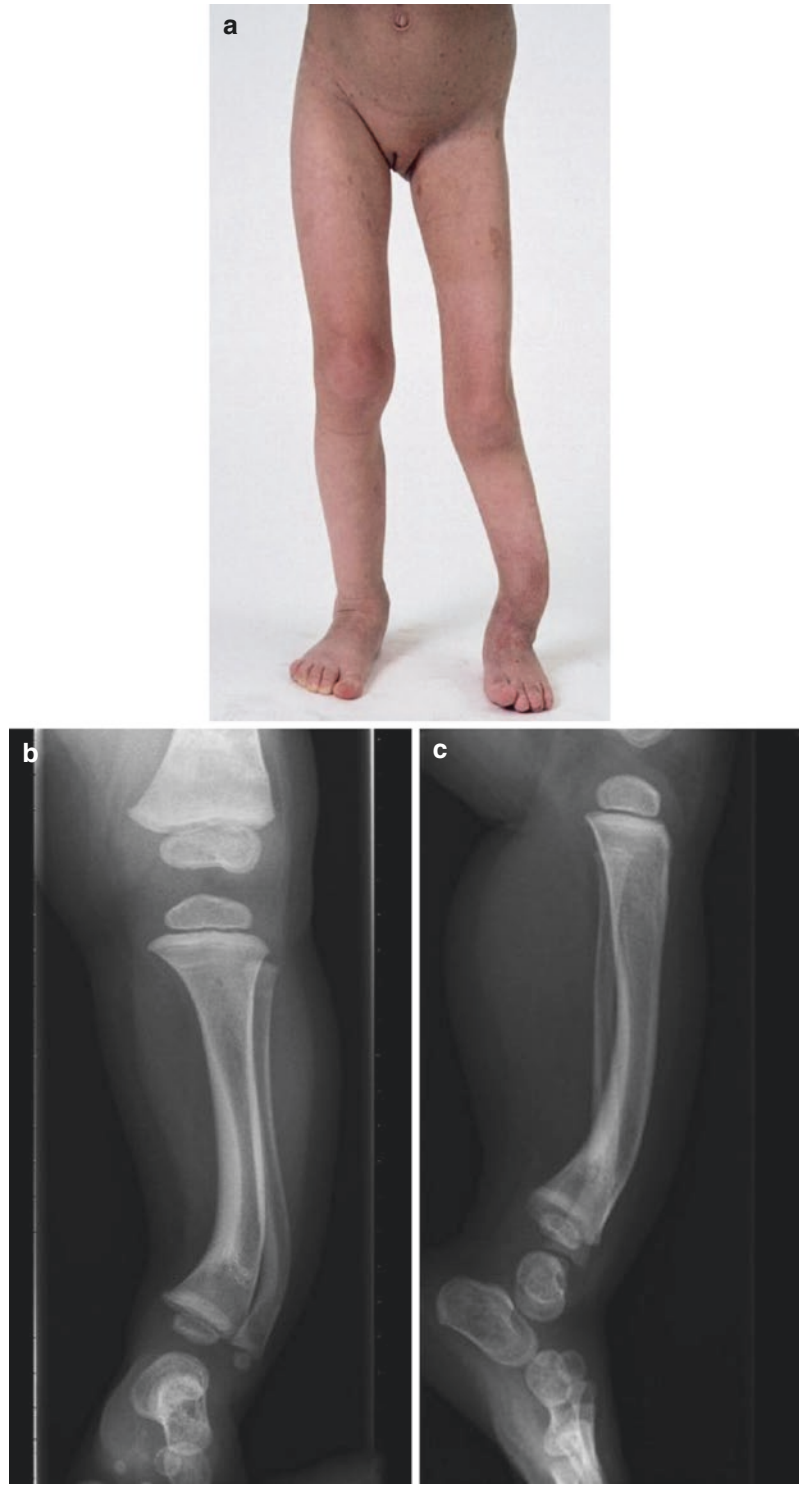



**Fig. 11.7** (**a**) Anterolateral bowing of the left lower leg in a sclerotic congenital tibial pseudarthrosis (type III). (**b**) Anteroposterior X-ray and (**c**) Lateral X-ray of the left lower leg. Sclerotic congenital tibial pseudarthrosis which finally will ends up in a pseudarthrosis

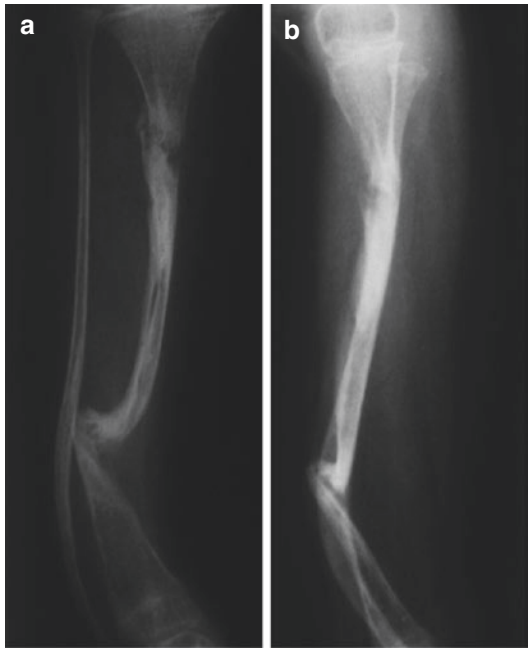



**Fig. 11.8** Dysplastic congenital tibial pseudarthrosis without fibular pseudarthrosis (type IV). (**a**) Anteroposterior and (**b**) Lateral X-ray of the right lower leg

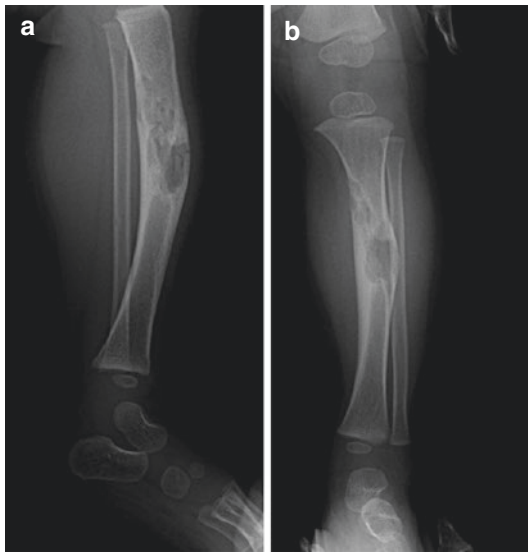



**Fig. 11.9** Fibrous dysplasia of the tibia (**a**) lateral and (**b**) anteroposterior X-ray of the left lower leg

Secondary care treatment: **congenital posteromedial tibial and fibular bowing.** Manual corrections will be carried out with plaster of Paris immobilization (normally 3–6 weeks) in the case of a serious foot dorsiflexion deformity followed by an orthosis in order to keep the foot in maximal plantar flexion and inversion. The brace will only be worn at night if the child is walking. In some cases in which a serious angulation is still present a corrective osteotomy may be considered after 4 years of age (Fig. 11.12). A difference in leg length of 2–4 or 5 cm can be corrected with an epiphysiodesis of the contralateral tibia (and if necessary the fibula). However, it should be pointed out that proximal tibial growth from 8 years of age until the completion of growth is on average 6 mm. In the case of a leg length difference of more than 4–5 cm a leg lengthening procedure may be considered.

## Lower Leg Pain

### Severe Lower Leg Pain

Complaint: severe lower leg pain.

Assessment: the lower leg is swollen and painful on pressure. The child cannot move or hardly move the foot.

Diagnosis: **acute compartment syndrome.**

Explanatory note: **acute compartment syndrome.** This occurs as a result of a sudden increase in pressure within one or more lower leg muscle compartments. The cause is a fracture or contusion or skintraction (Bryant) for treatment of a femural fracture in small children (Fig. 11.13). In Bryant traction children lie on their back with 90° of hip flexion and the legs undergo skintraction. The risks of an acute compartment syndrome are greater in children older than 2 years of age. They will develop a clubfoot deformity if the compartment syndrome is not promptlytreated.

Supplementary assessment: an acute compartment syndrome is diagnosed on physical examination.

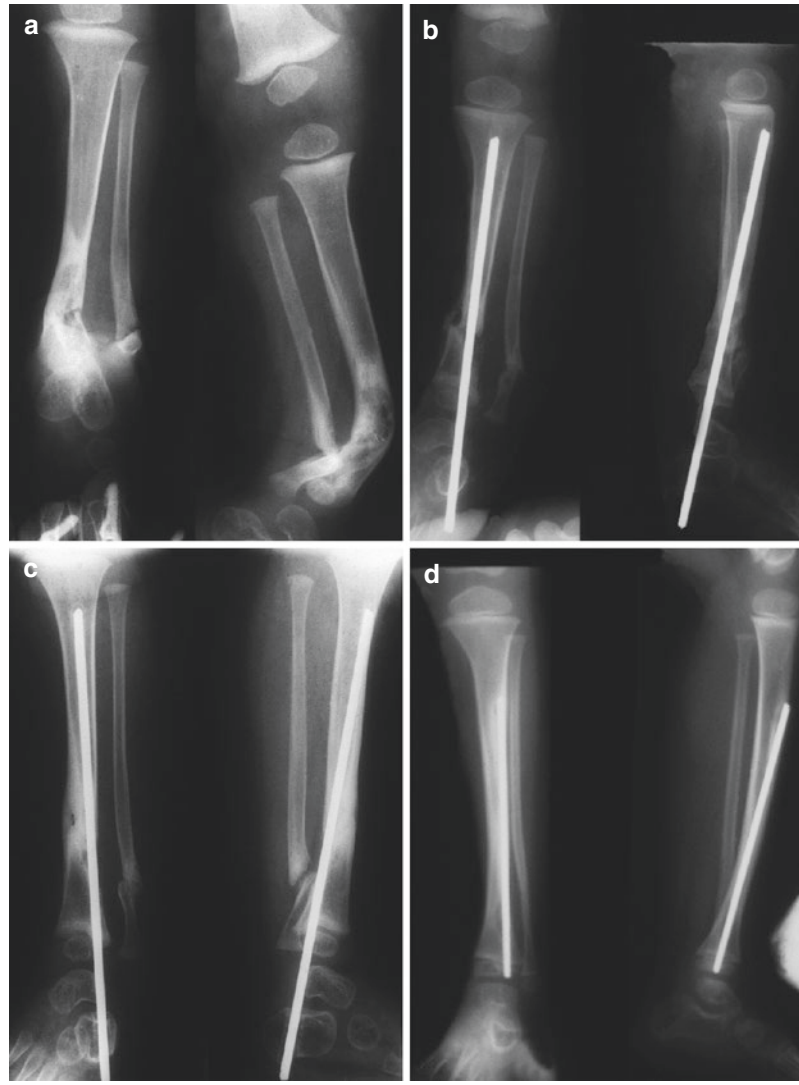



**Fig. 11.10** (**a**) Cystic congenital tibial pseudarthrosis at 10 months of age. (**b**) At 1 year (the same patient as in Fig. 11.11a). Correction and fixation using an intramedullary rod in the tibia inserted through the heel. With the passage of time the tibia grows in length and in this situation the distal end of the intramedullary rod finally ends up lying just above the ankle joint. (**c**) At 13 months of age. (**d**) At 3 years of age

Primary care treatment: none.

When to refer: an acute compartment syndrome should be referred as quickly as possible.

Secondary care treatment: **acute compartment syndrome.** The fascia of the involved compartments will be divided: anterior extensor muscle compartment, peroneal lateral compartment, superficial and deep posterior flexor muscle compartments (Fig. 11.14).

## Pain in the Outer Side of the Lower Leg

Complaint: there is pain on the outer side of the lower leg.

Assessment: the outer side of the lower leg is sensitive to pressure. An increase in pain occurs with repeated dorsiflexion movements against resistance.

Diagnosis: **chronic anterior tibial syndrome (chronic anterior compartment syndrome)**

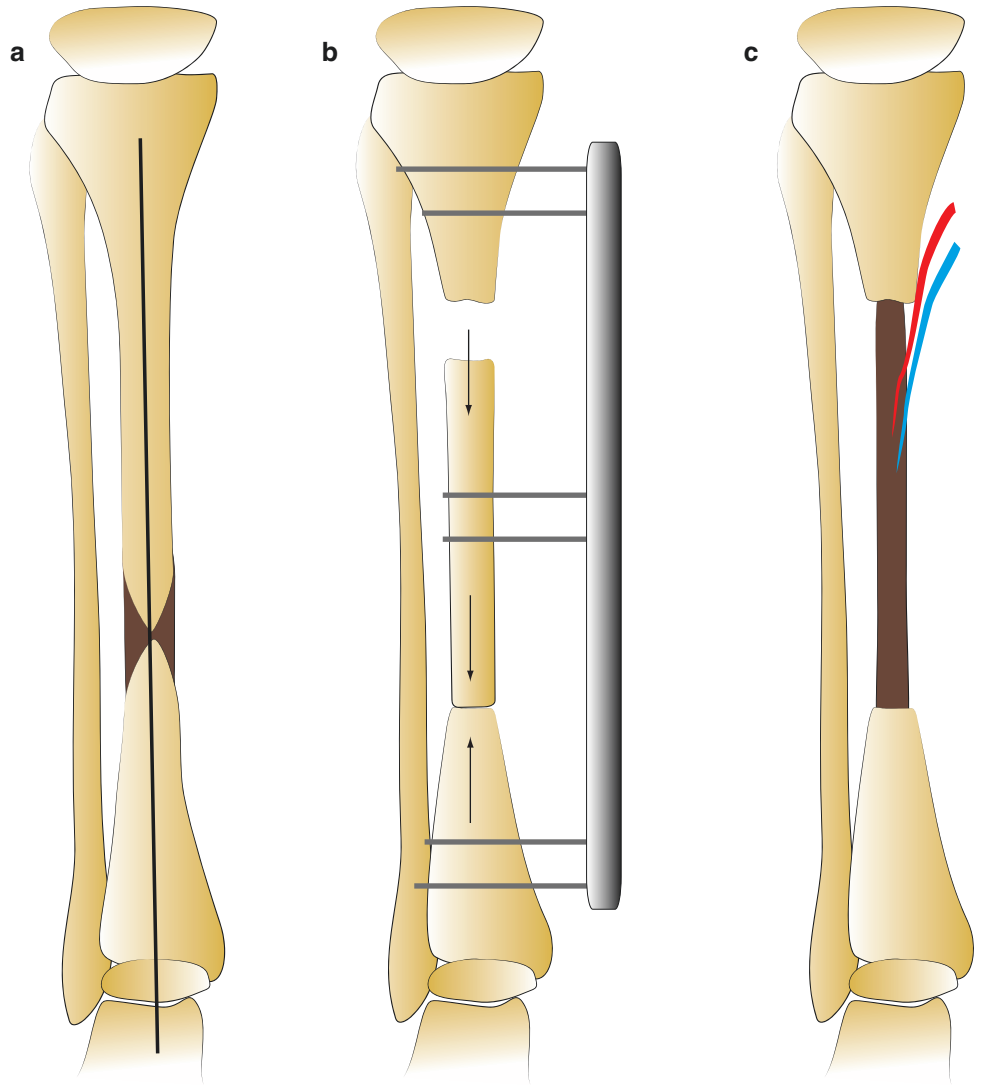



**Fig. 11.11** (**a**) Treatment of a congenital tibial pseudarthrosis using an intramedullary rod inserted through the heel with an autogenous bonegraft at the level of the curreted pseudarthrosis. (**b**) Bone transport technique with an Ilizarov frame. (**c**) Free vascularised fibula transplant in the affected leg taken from the normal side (Redrawn from: Staheli RT. *Fundamentals of pediatric orthopaedics*. 4th ed. Philadelphia/Baltimore/New York/London/Buenos Aires/Hongkong/Sydney/Tokyo: Lippincot Williams & Wilkins; 2008)

Explanatory note: **chronic anterior tibial syndrome.** A chronic compartment syndrome is almost always localized in the anterior compartment and is generally caused by excessive sport activities in which the pressure in the anterior compartment increases and causes pain (Fig. 11.14). Pain in this area can also be stimulated by repetitive dorsiflexion movements against foot resistance in which the examiner provides dorsal counter pressure.

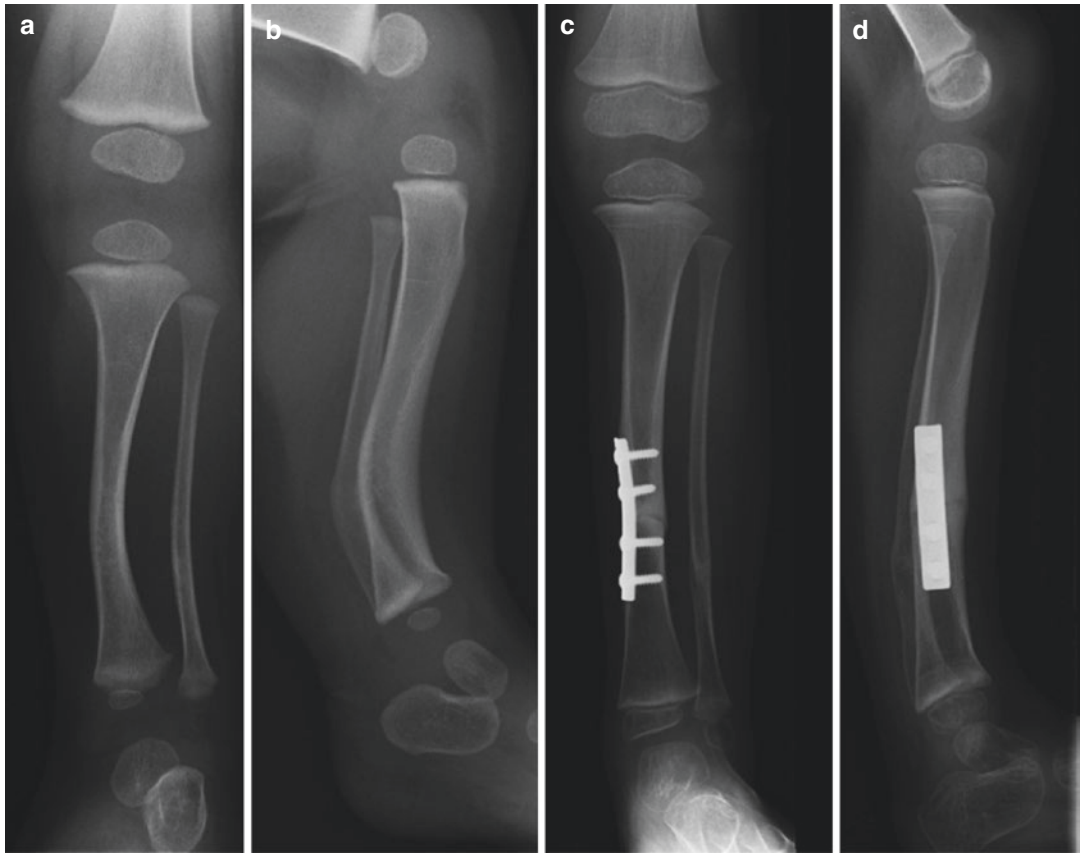



**Fig. 11.12** Congenital posteromedial bowing of the tibia and fibula of the left lower leg. (**a**) Anteroposterial X-ray. (**b**) Lateral X-ray. (**c**) Anteroposterior X-ray after corrective osteotomy. (**d**) Lateral X-ray after corrective osteotomy

Supplementary assessment: a chronic anterior tibial syndrome will usually be confirmed on physical examination. An intracompartmental anteriorcompartment pressure measurement may be considered if necessary. An intracompartmental pressure of 15 mmHg or more is suspicious for an anterior tibial syndrome.

Primary care treatment: reduce sport activities and provide a foot arch support.

When to refer: if reduction in sport activities and the arch support do not lead to reduction of the complaints.

Secondary care treatment: **anterior tibial syndrome.** Fasciotomy of the anterior compartment.

## Pain in the Inner Side of the Lower Leg

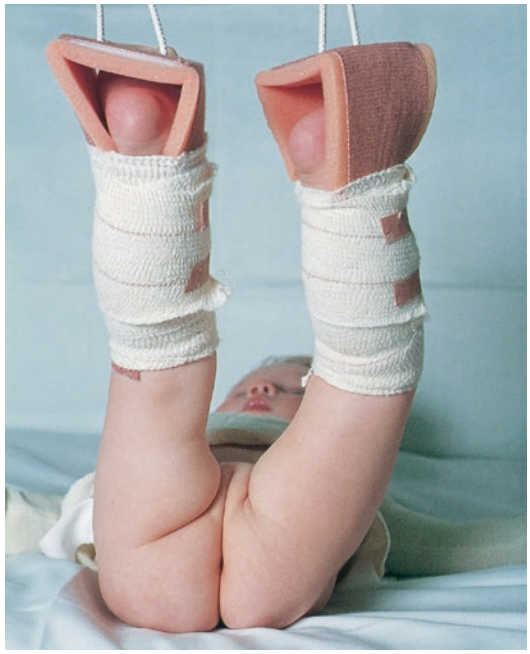
Complaint: pain on the inner side of the lower leg.

Assessment: the medial side of the tibia is sensitive to pressure.

Diagnosis: **tibial periostitis (shinsplints).**

Explanatory note: **tibial periostitis.** Periostitis is often called shinsplints. It is an overload syndrome that can occur in marathon runners but also in sportsmen who train on a hard surface with inadequate footwear. This problem seldom occurs under the age of 16.

Supplementary assessment: no abnormalities are seen on X-rays of the lower leg.



However, on a bone scan a diffuse increased activity will be seen on the medial side of the tibia in the case of periostitis.

Primary care treatment: several weeks of rest is often sufficient in slight cases. Usually a much longer period is necessary in order to achieve complete complaint regression. The cause is often insufficient shoe quality. If necessary a shock absorbent supportive sole can be fitted into the shoe.

When to refer: a periostitis should be referred after failure of conservative treatment.

Secondary care treatment: **tibial periostitis.** In a few cases a fasciotomy of the superficial flexor compartment may help.

**Fig. 11.13** Bryant traction

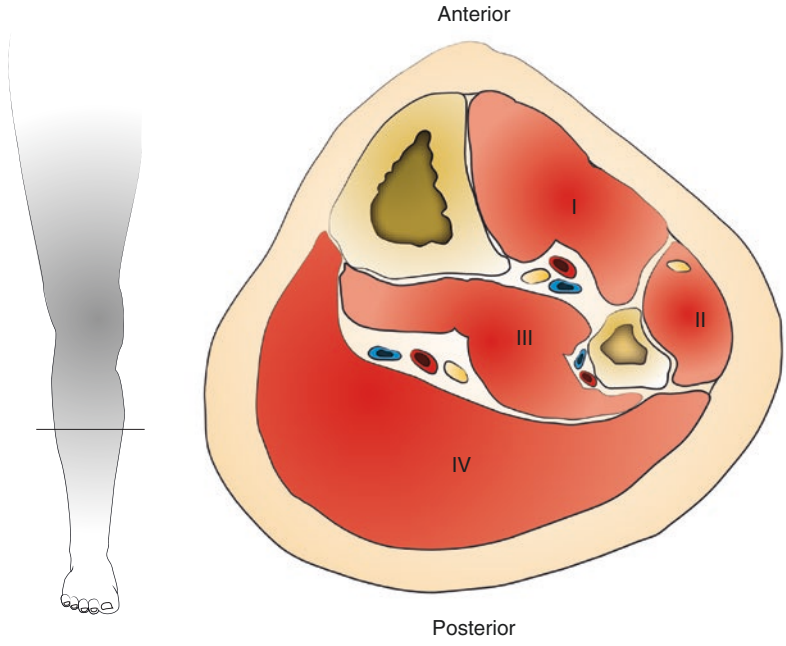



**Fig. 11.14** Transverse section of the lower leg with *I*: the anterior extensor compartment with the anterior tibial muscle, extensor digitorum longus muscle, extensor hallucis longus muscle, peroneus tertius muscle, anterior tibial artery and vein and the deep peroneal nerve. *II* The lateral peroneal compartment with the peroneus longus muscle, peroneus brevis muscle and the superficial peroneal nerve. *III* The deep posterior flexor compartment with the flexor digitorum longus muscle, posterior tibial muscle, flexor hallucis longus muscle, posterior tibial artery and vein and the peroneal artery and vein. *IV* The superficial posterior flexor compartment with the soleus muscle, gastrocnemius muscle and plantaris muscle

**Differential Diagnosis: Lower Leg Pain**

| | |
|---|---|
| **Severe lower leg pain** | |
| The foot cannot or hardly be moved | **Acute compartment syndrome** |
| **Pain in the outer side of the lower leg** | |
| Pain after repeated dorsal extension movements of the foot against resistance | **Chronic anterior tibial syndrome (chronic anterior compartment syndrome)** |
| **Pain in the inner side of the lower leg** | |
| The medial side of the tibia is pressure sensitive | **Tibial periostitis (shinsplints)** |

**Differential Diagnosis: Lower Leg Misalignment**

| | |
|---|---|
| **Forwards and outwards lower leg bowing** | |
| Anterolateral bowing of the lower leg | **Congenital anterolateral tibial and possibly the fibular bowing** |
| Bowing of both thighs and lower legs with equinus deformity or clubfeet and an abnormal face | **Camp(t)omelic dysplasia (Cumming dysplasia syndrome)** |
| Anterolateral bowing with tibial defect | **Congenital tibial pseudarthrosis** |
| Stress fracture after 5 years of age | Type I, |
| Cysts | Type II, |
| Sclerosis | Type III, |
| Dysplasia | Type IV, |
| Dysplasia with fibula pseudarthrosis | Type V, |
| Bowing occurs just before puberty | **Fibrous dysplasia of the tibia** |
| **Backwards and inwards lower leg bowing** | |
| Posteromedial lower leg bowing, hyperextension in the ankle joint | **Congenital posteromedial tibial and fibular bowing** |

# Ankle

# 12

## Ankle Misalignment

### The Heel Progressively Deviates Outwards (Valgus Deformity)

Complaint: pressure sores appear on the medial side of the ankle caused by the shoes.

Assessment: there is a valgus deformity of the ankle joint (Fig. 12.1).

Differential diagnosis:

**multiple osteochondromata (exostoses)**
**infected or fractured growth plate**
**achondroplasia**
**congenital fibular pseudarthrosis**
type I
type II
type III
type IV
**dysplasia epiphysealis hemimelica (Trevor disease)**

Explanatory note: **multiple osteochondromata.** An osteochondroma (exostosis) is a developmental disturbance in the periphery of the epiphyseal growth plate. A bony outgrowth develops usually with a cartilaginous cap. The deformity increases until growth stops. It may be solitary but may also be multiple. A slight valgus deformity may be caused by an osteochondroma in the distal part of the fibula (Fig. 12.2). As a result there is a disturbed fibula growth which acts as a brake and causes a vlagus deformity.

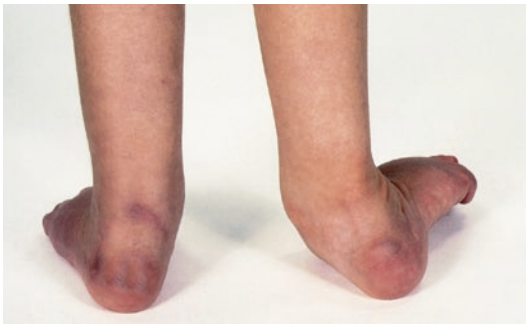


**Fig. 12.1** Valgus deformity in both ankles, right greater than left

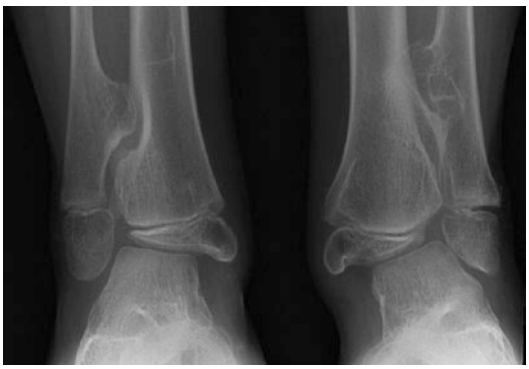


**Fig. 12.2** Anteroposterior X-ray of the ankles: osteochondroma (exostosis) can be seen in the distal part of both fibulas with a resulting valgus deformity in both talocrural joints


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_12

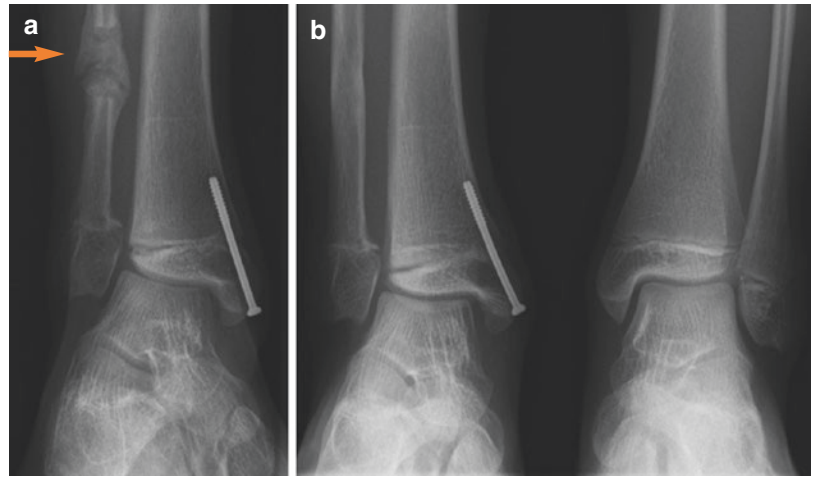



**Fig. 12.3** (**a**) Epiphyseal fracture in the distal part of the right fibula in which the growth plate has closed. Situation after resection of a part of the fibula shaft with excessive callus formation (*arrow*) and a hemi-epiphysiodesis of the growth plate at the level of the medial malleolus using a screw. (**b**) The valgus deformity in the right talocrural joint has been partly corrected

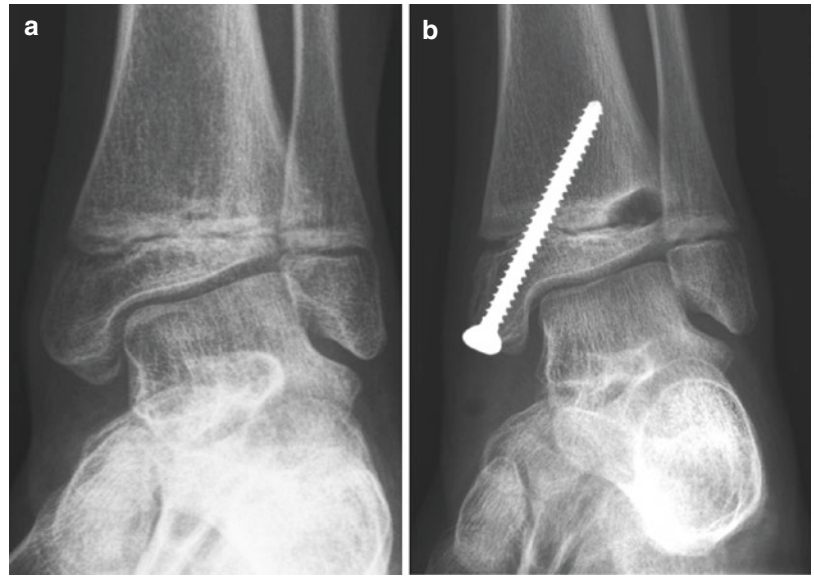



**Fig. 12.4** (**a**) There is a bony connection between the metaphysis and the epiphysis on the lateral side of the distal tibial growth plate after a growth plate fracture. (**b**) The bony bridge has been removed with a temporary hemi-epiphysiodesis of the medial malleolus

**Infected or fractured growth plate** Destruction of the epiphysis can occur due to an infection at the level of the epiphysis or an epiphyseal fracture in the distal part of the fibula. Growth in the distal fibular growth plate stops and works as a brake. However, the growth plate in the distal part of the tibia carries on growing on the medial side and this results in an ankle valgus deformity (Fig. 12.3). A valgus deformity can also be caused by a growth plate fracture as a result of damage to the lateral part of the distal tibial growth plate (Fig. 12.4).

**Achondroplasia** A valgus deformity in the ankles can be part of achondroplasia.

**Congenital fibular pseudarthrosis** This is a rare deformity. Four types can be classified (Fig. 12.5):

Type I: bowed fibula without pseudarthrosis.
Type II: congenital fibular pseudarthrosis without misalignment in the talocrural joint.
Type III: congenital fibular pseudarthrosis with a valgus misalignment in the talocrural joint.
Type IV: congenital fibular pseudarthrosis with a valgus misalignment at the ankle with a developing pseudarthrosis of the tibia later on.

**Dysplasia epiphysealis hemimelica** Dysplasia epiphysealis hemimelica is an osteochondral

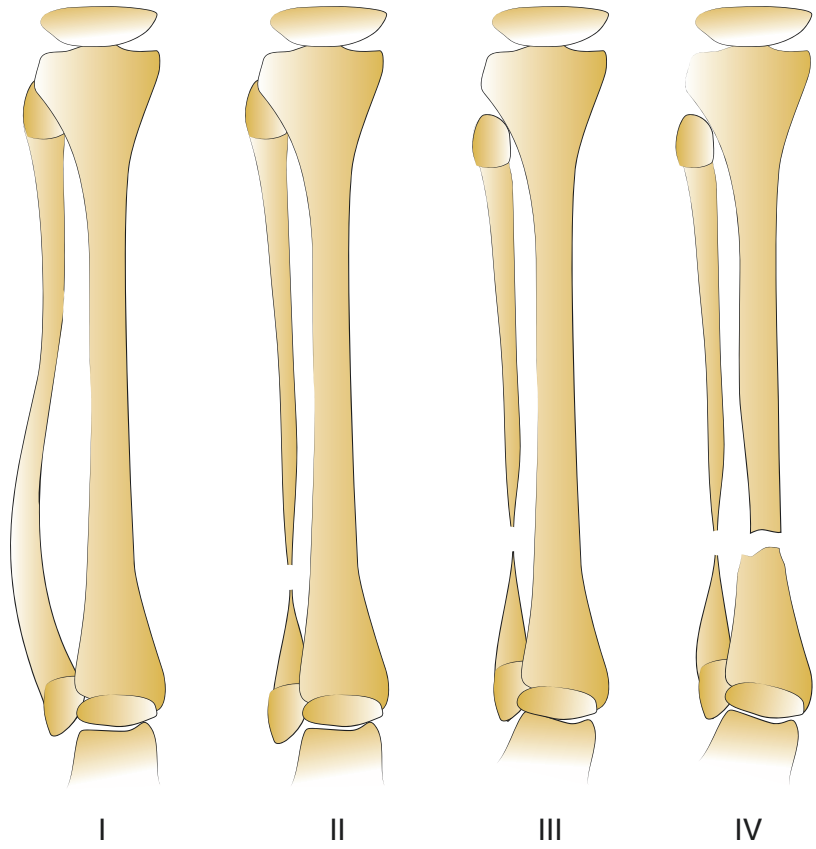



**Fig. 12.5** Classification of congenital fibular pseudarthrosis according to and (Redrawn from: Dooley BJ, Menelaus MB, Paterson DC. Congenital pseudarthrosis and bowing of the fibula. *J Bone Joint Surg Br*. 1974;56:739–43). The deformity can vary in severity: bowing of the fibula without pseudarthrosis (type *I*), a pseudarthrosis without (type *II*) or with valgus deformity (type *III*) and with latent pseudarthrosis of the tibia (type *IV*)

overgrowth of the epiphysis or articular cartilage. The condition is intra-articular and is limited to half of the extremity (hemimelia). The medial side of the tibia or talus is twice as often active than the lateral side. The valgus deformity occurs if the medial side is affected. (see also pp. 178, 179 and 183).

Supplementary assessment: anteroposterior and lateral X-rays of the ankles while standing. A CT or MRI scan of the ankles must be taken in the case of osteochondromata causing an ankle valgus deformity, growth plate damage as a result of an infection or a fracture and in the case of dysplasia epiphysealis hemimelica.

Primary care treatment: none.

When to refer: in the case of a progressive valgus deformity.

Secondary care treatment: **multiple osteochondromata**. Usually it is sufficient to remove the osteochondroma and perform a medial malleolus hemi-epiphysiodesis using 1 screw or 2 staples or a so-called eight plate. Once the ankle is straight the materials will be removed.

**Infected or fractured growth plate** There is no more growth potential due to complete closure of the growth plate in the distal part of the fibula. One can try to correct the deformity by resecting part of the fibula shaft and carry out an epiphysiodesis of the medial malleolus (Fig. 12.3). One can also consider a supramalleolar varus osteotomy with a fibulotomy if the above mentioned techniques are insufficient (Fig. 12.6). Operative excision of the bony bridge can be considered after a growth plate fracture in the lateral part of the growth plate and the distal part of the tibia if less than a third of the total growth plate is involved. After this the growth plate generally functions normally and in many cases a spontaneous correction of the valgus deformity will occur

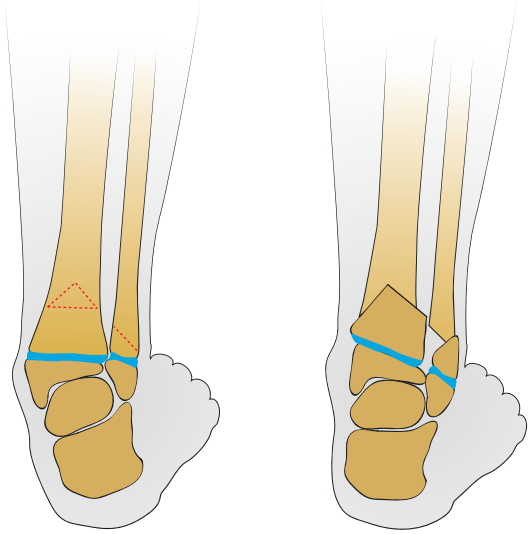


**Fig. 12.6** Supramalleolar varus osteotomy with fibulotomy on the right side

because the growth plate tries to line up vertical to the weight bearing line (Hueter-Volkmann, law 1862). Temporary medial malleolus hemi-epiphysiodesis may possibly speed up this process (Fig. 12.4).

**Achondroplasia** A temporary hemi-epiphysiodesis may be carried out by fixing the medial part of the growth plate in the distal part of the tibia with 1 screw, or 2 staples or a so-called eight-plate in the case of a minimal deformity (Fig. 12.7). The materials must be removed once the ankle is straight. These materials should not be left in position for more than 2 years because in that case a bony bridge will develop on the medial side of the growth plate in the distal part of the tibia which will result in a varus deformity. A supramalleolar valgus tibial osteotomy with a fibulotomy must be carried out in the case of more serious deformities (Fig. 12.6).

**Congenital fibular pseudarthrosis** Plate fixation with an autologous cancellous bonegraft should be carried out in the case of complaints. At the same time correction of the valgus deformity must also be carried out. If the fibula pseudarthrosis is still present after the above mentioned treatments an artificial synostosis between distal part of the tibia and fibula will be carried out (Fig. 12.8). One can consider a

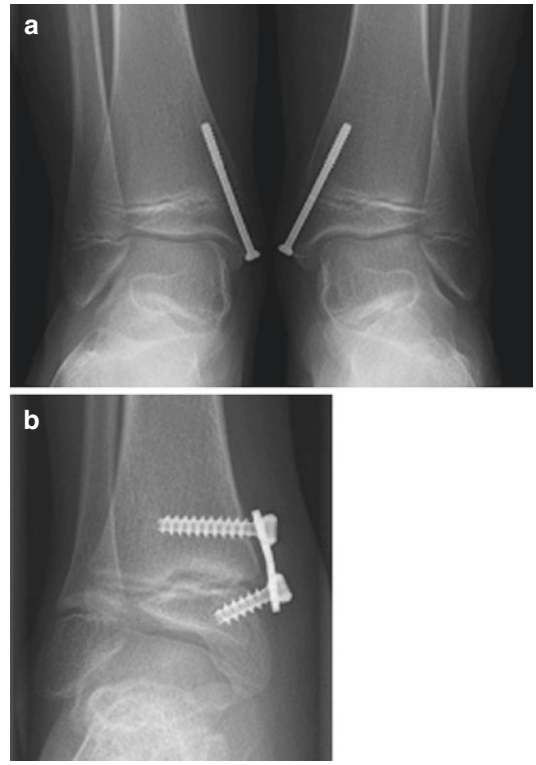



**Fig. 12.7** (**a**) Temporary medial malleolus hemi-epiphysiodesis using screws because of the valgus misalignment of both talocrural joints. (**b**) Temporary hemi-epiphysiodesis using an eight-plate at the level of the medial malleolus because of a valgus misalignment in the talocrural joint

correction of a possible valgus deformity with a temporary hemiepiphysiodesis of the medial malleolus using a screw or a superamalleolar varus osteotomy and fibulotomy once the child is grown up (for treatment of a tibia pseudarthrosis type IV congenital fibula pseudarthrosis, see p. 245).

**Dysplasia epiphysealis hemimelica** A wedge excision at the level of the deformity to correct the deformity may be carried out. There is a high risk of recurrence.

## The Heel Progressively Deviates Inwards (Varus Deformity)

Complaint: the child walks on the lateral side of the foot and frequently stumbles through the ankle.

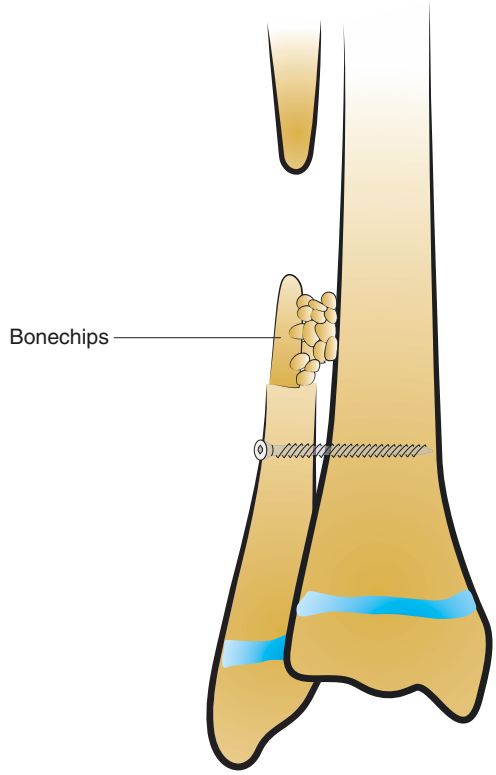



**Fig. 12.8** Artificial synostosis between the distal part of fibula and the tibia with a cancellous bone graft and screw fixation (Redrawn from: Langenskiöld A. Pseudarthrosis of the fibula and progressive valgus deformity of the ankle in children: treatment by fusion of the distal tibial and fibular metaphyses. Review in three cases. *J Bone Joint Surg Am*. 1967;49:463–70)

Assessment: there is a varus deformity in the ankle.

Differential diagnosis:

**infected or fractured growth plate dysplasia epiphysealis hemimelica (Trevor disease)**

Explanatory note: **infected or fractured growth plate**. This is the situation after an infection or an epiphyseal fracture in the distal part of the tibia in which there is destruction of a part of the epiphysis and where a bony bridge develops between the distal metaphysis and epiphysis on the medial side of the tibia.

**Dysplasia epiphysealis hemimelica** This is an osteochondral overgrowth in the epiphysis or articular cartilage. A varus deformity occurs if the abnormality is localized laterally. (see also pp. 178, 179 and 183).

Supplementary assessment: anteroposterior and lateral X-rays of both ankles while standing and also a CT-scan of the ankle.

Primary care treatment: none.

When to refer: every child with a varus deformity in the ankle should be referred

Secondary care treatment: **infected or fractured growth plate**. A CT-scan would initially be carried out to establish the extent of the damage. There is no point in removing the bony bridge if this involves more than a third of the growth plate. The remaining part of the growth plate is not sufficiently capable of correcting the deformity. One can try to operatively remove a smaller bony bridge. In that case the growth plate will carry on growing and generally one sees a spontaneous correction (Hueter-Volkmann law 1862). A supramalleolar valgus tibial osteotomy with a fibulotomy can be carried out if the bony bridge involves more than a third of the growth plate or if the removal of the bony bridge has failed or if not much more growth is expected. The wedge will be filled up with (donor) bone. The distal epiphysis of the fibula must be destroyed because this will otherwise still keep growing on and as result the lateral malleolus will become relatively too long. Leg length differences must be checked for and corrected if necessary.

**Dysplasia epiphysealis hemimelica** Wedge resection at the level of the anomaly so that the anomalous position will be corrected.

## Ankle Pain

### Pain at the Front of the Ankle

Complaint: the child complains of pain at the front of the ankle without an obvious trauma. This usually involves children at puberty who play a lot of sports.

Assessment: pain on pressure will occur on the anterolateral side of the ankle just medial to the fibula or on the anteromedial side just lateral to the medial malleolus or on the

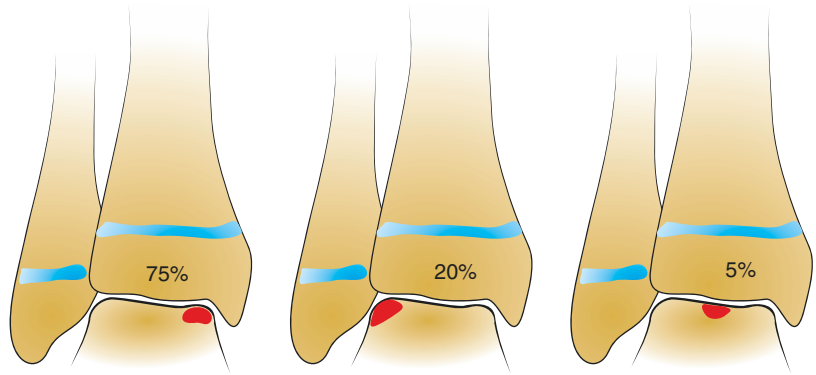



**Fig. 12.9** Localizations of osteochondritis dissecans in the talus

**Table 12.1** classification of osteochondrosis dissecans

| | |
|---|---|
| Grade I | Non displaced osteochondral fragment |
| Grade II | Non displaced loose lying osteochondral fragment |
| Grade III | Partially displaced osteochondral fragment |
| Grade IV | Fully displaced osteochondral fragment (corpus liberum) |

anterior side of the ankle. Sometimes a swelling is present in the ankle joint.

Diagnosis: **osteochondritis dissecans of the talus**.

Explanatory note: **osteochondritis dissecans of the talus**. In an osteochondritis dissecans a small part of the cartilage surface with the underlying bone with a diameter of about 0.5 cm becomes loose. With the passage of time this can become a corpus liberum (free body). In 75 % of cases the lesion is on the medial side, in 20 % of cases on the lateral side and in 5 % of cases centrally (Fig. 12.9). An osteochondritis on the lateral side is caused by earlier ankle trauma in 90 % of cases and in the case of medially located lesions 60 %. In the majority of cases osteochondral lesions of the ankle joint are associated with previous traumas.

Before the osteochondral lesion completely separates from the rest of the talus the process goes through several stages (Table 12.1).

Supplementary assessment: anteroposterior and lateral X-rays of the ankle joint do not always show the anomaly. An oblique X-ray sometimes helps, but a CT-scan or an MRI arthrogram is always positive (Fig. 12.10).

Primary care treatment: the advice is to limit weight bearing sporting activities in stage I or stage II. X-rays of the ankle will be repeated after 6 months.

When to refer: if the conservative treatment does not lead to reduction in complaints or if after 6 months no radiological improvement has occurred after stopping weight bearing sporting activities.

Secondary care treatment: **osteochondritis dissecans of the talus**. A lower leg walking plaster of Paris splint for 6 weeks can be prescribed if there is still a lot of pain in the case of stage I or II without radiological deterioration. Operative fixation of the osteochondritis dissecans fragment is recommended in stage III and when radiological deterioration has occurred in stage II. In stage IV if possible replacement and fixation of a fragment of ≥15 mm is preferred. For smaller fragments excision, curettage and bone marrow stimulation (micro-fracture technique) is the first choice of treatment. Secondary treatment may consists of osteochondral transplantation in which autologous bone with cartilage from the non weight bearing part of the knee will be implanted into the defect. Recent studies are directed at so called biological cartilage repair using cultivated or the patient's own chondrocytes, osteochondral, periostial or perichondrial

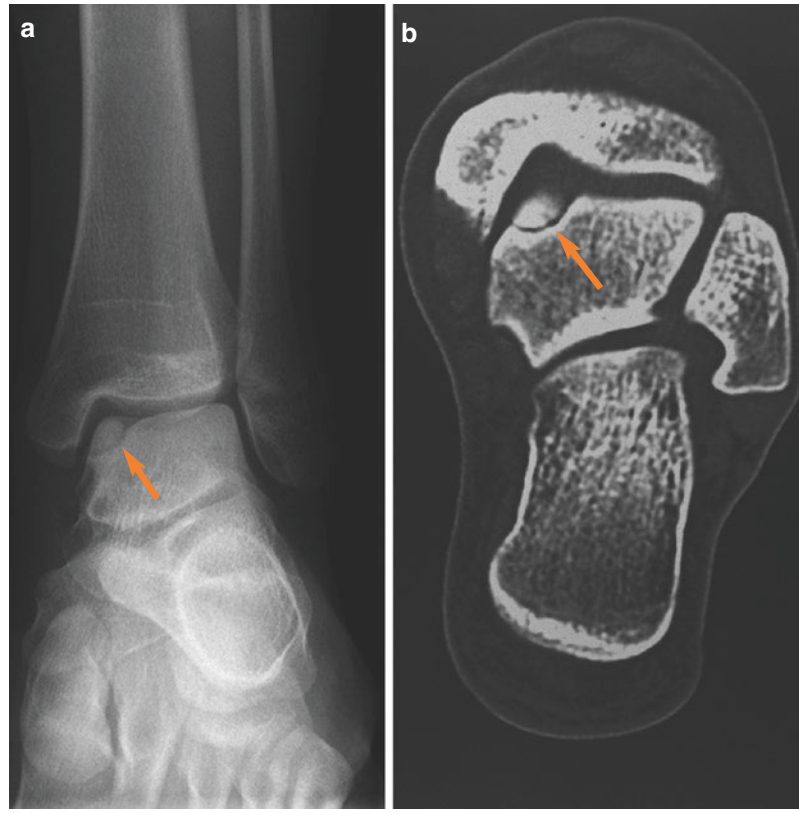



**Fig. 12.10** Osteochondritis dissecans of the medial side of the talus roll (*arrow*). (**a**) Anteroposterior X-ray of the ankle. (**b**) CT-scan (*arrow*)

bone. It is not yet clear which treatment is best.

## Pain at the Back of the Ankle

Complaint: pain on the posterior side of the ankle in children who have activities which involve toe walking, for instance in ballet dancing.

Assessment: this complaint is mostly present in maximal ankle plantar flexion.

Diagnosis: **symptomatic os trigonum**.

Explanatory note: **symptomatic os trigonum**. An os trigonum (Latin for three corners) occurs because the ossification center of the lateral tubercle of the posterior process of the talus does not fuse with the talus or because it becomes separated from the rest of the talus due to a trauma. This tubercle is lateral to the groove in which the tendon of the flexor hallucis longus muscle is situated.

On the medial side of this tendon the medial tubercle of the posterior process of the talus is situated. The ossification centre of the lateral tubercle appears between 8 and 11 years of age and in general fuses with the rest of the talus within a year. If this doesn't happen the lateral tubercle remains as a rounded off three cornered piece of bone which can be seen on the posterior side of the talus. An os trigonum is generally asymptomatic. In symptomatic cases a trauma is mentioned in 80 % of cases. As a rule the child has twisted or sprained the ankle. The lateral tuberculum contacts the posterior side of the tibia with the ankle in maximum plantar flexion. The lateral tubercle can be broken off in forced plantar flexion. On the lateral tubercle X-ray an irregular three cornered os trigonum can be seen.

Supplementary assessment: anteroposterior and lateral X-rays of the ankle (Fig. 12.11).

Primary care treatment: none.

When to refer: with persistent complaints.

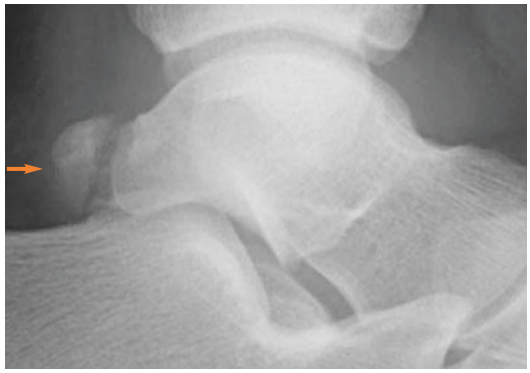


**Fig. 12.11** Lateral X-ray of the ankle: os trigonum (*arrow*)

Secondary care treatment: lower leg plaster immobilization during 6 weeks. If this does not help a corticosteroid injection may be tried but if necessary operative removal of the os trigonum should be considered. The chances of improvement are less if these complaints have been present for more than 2 years, compared to children who have had complaints for a shorter period.

## Extremely Painful Immobile Ankle

Complaint: the ankle is severely painful and cannot be moved.

Assessment: the ankle is severely painful.

Differential diagnosis:

**acute osteomyelitis**
**septic arthritis**

For explanatory note, supplementary assessment, primary care treatment, when to refer and secondary care treatment, see Chap. 18.

## Ankle Sprain

Complaint: the child complains of pain on the outer side of the ankle after a sprain. It usually involves children at puberty who regularly sprain the ankle with the foot turned inwards. There may or may not be a previous history of torn ligaments.

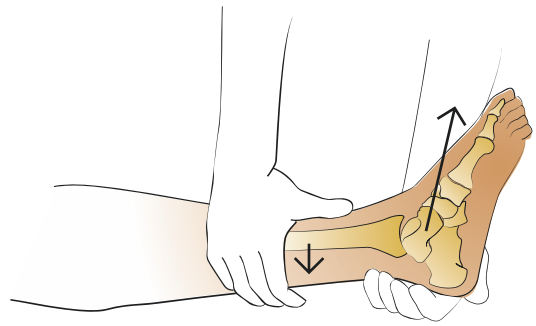


**Fig. 12.12** The anterior drawer test is carried out with the ankle in 30° of plantar flexion. As a result of this the posterior talofibular ligament and the calcaneo fibular ligament are relaxed and the peroneal tendons will not prevent anterior shifting. The examiner exerts counter pressure with one hand on the lower leg and pulls the heel anteriorly with the other hand. The test is positive if the foot shifts more than 3 mm anteriorly

Assessment: there is often hypermobility in the joints caused by inborn flaccid ligaments (see Fig. 10.26, p. 123). The anterior drawer test of the ankle (Fig. 12.12) may be positive on both sides.

Differential diagnosis:

**recurrent ankle inversion injuries with hypermobility**
**recurrent posttraumatic ankle inversion injuries**
**ball- and- socket ankle**

Explanatory note: **recurrent ankle inversion injuries with hypermobility**. Regular sprains are caused by a hypermobile ankle joint. The findings are symmetrical on both left and right side. The tendency to twist the ankle disappears when the child becomes older when there is reduction in the joint hypermobility.

**Recurrent posttraumatic ankle inversion injuries** As a rule torn ankle ligaments occur only in children 12 years of age or older. The lateral ankle ligament complex consists of 3 parts: the anterior talofibular ligament, the calcaneofibular ligament and the posterior talofibular ligament. The first 2 ligaments mentioned are the most susceptible for tears after inversion injuries to the ankle joint. Fifty percent of cases will regularly have a feeling that the ankle twists

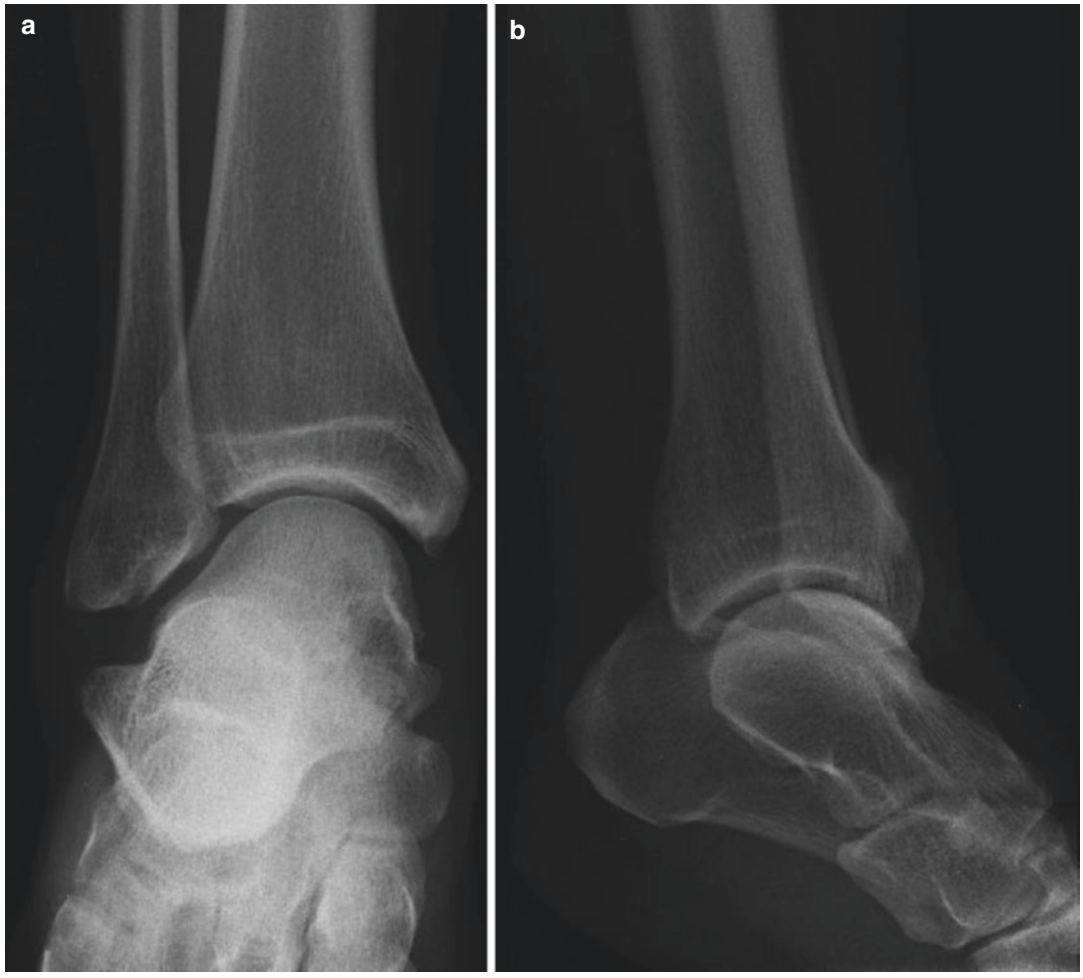



**Fig. 12.13** (**a**) anteroposterior and (**b**) lateral X-rays of the ball-and-socket ankle with a talocalcaneal coalition

after a healed tear of the lateral ankle ligament complex. Only in 1 or 2 % with an insufficient lateral ankle ligament complex have a positive anterior drawer test.

**Ball-and-socket ankle** The ankle is rounded off in the frontal plane and has more of a ball shape than a roll, so that more lateral mobility is present in the ankle (Fig. 12.13). Other accompanying deformities can be a tarsal coalition (see for tarsal coalition, pp. 279–285), a skew foot (see for a skew foot, pp. 265–267), a fibular hypoplasia or aplasia (see pp. 378–381).

Supplementary assessment: anteroposterior and lateral X-rays of the ankle and forced positional views of both ankles (Fig. 12.14).

Primary care treatment: prescribe high ready-made footwear with a strongly reinforced heel.

When to refer: if wearing the above mentioned shoes have no effect.

Secondary care treatment: **recurrent ankle inversion injuries with hypermobility**. Usually it is sufficient to wear an ankle brace for 4 months (Fig. 12.15). The ankle brace will only be worn in sports activities after 4 months until the complaints completely disappear.

**Recurrent posttraumatic ankle inversion injuries** In the first instance an ankle brace may be prescribed. If the child has stopped growing but still has problems with ankle twisting because of an insufficient lateral ankle ligament complex operative more proximal reinsertion of the lax ligaments (Fig. 12.16) or reconstruction can be carried out. The tendon of the peroneus brevis muscle may be used for reconstruction of the lateral ankle ligament complex according to Evans (Fig. 12.17a) or Watson-Jones (Fig. 12.17b) or a modification of these techniques. The ankle ligaments can also be reconstructed using the tendon of the plantaris muscle or the long extensor tendon of the second toe (Fig. 12.17c). More operation techniques are possible.

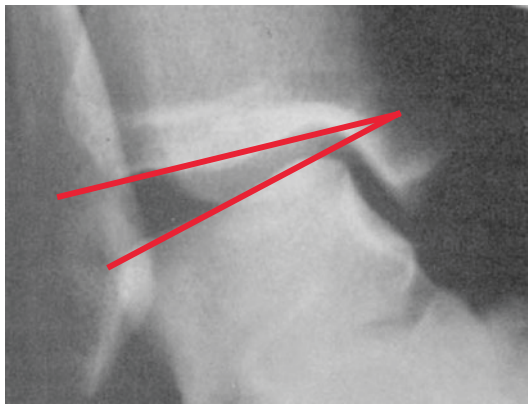


**Fig. 12.14** Stress radiograph of the ankle. Anteroposterior X-ray of the ankle in which the examiner puts counter pressure on the lower leg with one hand and inverts the foot with the other hand. This can be carried out on both the injured and uninjured side. The angle between the tibia and talus is measured on both sides (*talar tilt*). If the difference on the injured and on the uninjured side is less than 5° this is within the normal physiological situation. If the difference is between 5° and 10° there is a possible insufficient lateral ankle ligament complex and in more than 10° then there is strong evidence of an insufficient lateral ankle ligament complex

**Ball-and-socket ankle** An ankle orthosis may be prescribed in the case of complaints. An arthrodesis of the ankle joint may be considered in the case of unusually exceptionally unstable ankles.

**Fig. 12.15** Ankle orthosis (*brace*) type: Swede-O

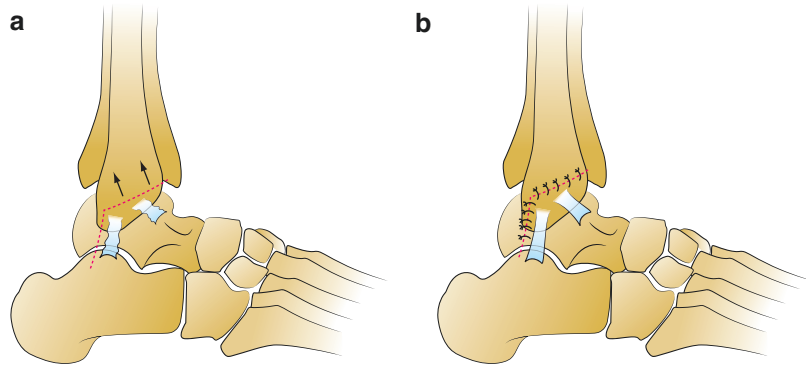



**Fig. 12.16** Proximal reinsertion of the anterior talofibular ligament and the calcaneofibular ligament according to Ahlgren and Larsson (Redrawn from: Ahlgren O, Larsson S. Reconstruction for lateral ligament injuries of the ankle. *J Bone Joint Surg Br*. 1989;71-B:300–3). (**a**) Incision periosteum. (**b**) Reinsertion of both ligaments under proximal tension with a periostial flap. Sutures using drill holes in the fibula

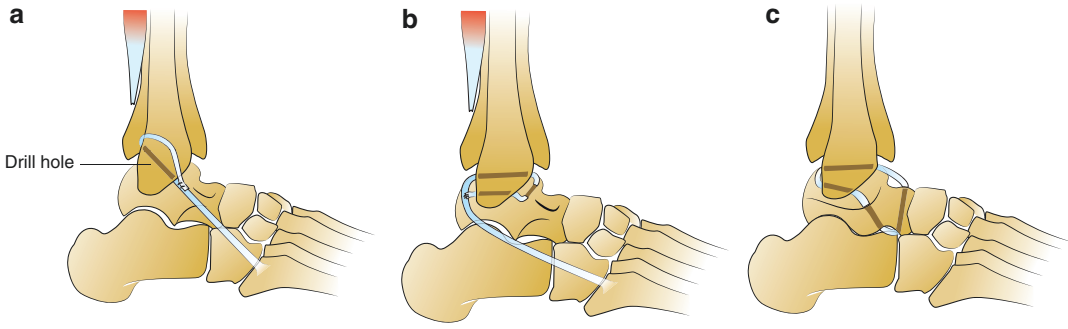



**Fig. 12.17** (**a**) Reconstruction of the lateral ankle ligament complex according to Evans. The tendon of the peroneus brevis muscle will be divided at the level of the muscle tendon transition. The proximal part will be sutured onto the tendon of the peroneus longus muscle. The distal part will be pulled through a drill hole in the distal part of the fibula and fixed to itself (Redrawn from: Evans DL. Recurrent instability of the ankle – a method of surgical treatment. *Proc R Soc Med*. 1953;46:343–4). (**b**) Reconstruction of the lateral ankle ligament complex according to Watson-Jones. The lateral ankle ligament complex will be reconstructed by pulling the tendon of the peroneus brevis muscle through 2 drill holes in the fibula and 1 drill hole in the talus and the tendon will be pulled through and fixed to itself under tension. The proximal part of the tendon of the peroneus brevis muscle will be sutured onto the tendon of the peroneus longus muscle (Redrawn from: Watson-Jones R. Recurrent forward dislocation of the ankle joint. *J Bone Joint Surg Br*. 1952;34-B:519). (**c**) Lateral ankle ligament reconstruction according to Sefton. In this case the lateral ankle ligament complex will be reconstructed using the tendon of the plantaris muscle or the long extensor tendon of the second toe. The tendon will be pulled through two drill holes in the distal part of the fibula and two drill holes in the talus and fixed to itself under tension (Redrawn from: Sefton GK, George J, Fitton JM, McMullen H. Reconstruction of the anterior talofibular ligament for the treatment of the unstable ankle. *J Bone Joint Surg Br*. 1979;61-B:352–4)

**Differential Diagnosis: Ankle Misalignment**

| | |
|---|---|
| **The heel progressively deviates outwards (valgus deformity)** | |
| Swellings around the ankle after an infection or growth plate fracture | **Multiple osteochondromata (exostoses)** |
| | **Infected or fractured growth plate** |
| Dwarf growth | **Achondroplasia** |
| Defect in the fibula | **Congenital fibular pseudarthrosis** |
| Bowed without pseudarthrosis | Type I |
| Pseudarthrosis | Type II |
| Pseudarthrosis with a valgus deformity ankle joint | Type III |
| Pseudarthrosis with a valgus deformity ankle joint together with a tibial pseudarthrosis | Type IV |
| Movement limitations and swelling on the medial side of the ankle | **Dysplasia epiphysealis hemimelica (Trevor disease)** |
| **The heel progressively deviates inwards (varus deformity)** | |
| After an infection or a growth plate fracture | **Infected or fractured growth plate** |
| Limitations of movement and swelling on the lateral side of the ankle | **Dysplasia epiphysealis hemimelica (Trevor disease)** |

**Differential Diagnosis: Ankle Pain**

| | |
|---|---|
| **Pain at the front of the ankle** | |
| Pressure pain medially, laterally or centrally at the level of the ankle joint | **Osteochondritis dissecans of the talus** |
| **Pain at the back of the ankle** | |
| Pain particularly on maximal plantar flexion in the ankle joint | **Symptomatic os trigonum** |
| **Extremely painful immobile ankle** | |
| The ankle is severly painful and no longer mobile | **Acute osteomyelitis** |
| | **Septic arthritis** |

**Differential Diagnosis: Ankle Sprain**

| | |
|---|---|
| Ankle sprain occurring as a result of hypermobility | **Recurrent ankle inversion injuries due to hypermobility** |
| Positive drawer test of the ankle joint | **Recurrent posttraumatic ankle inversion injuries** |
| The ankle joint is rounded off on anteroposterior X-rays | **Ball-and-socket ankle** |

# Foot

# 13

## Foot Deformities

### Foot Deformities Present at Birth

Complaint: the parents find that their child's foot has an abnormal appearance.

Assessment: the foot has an abnormal appearance.

Differential diagnosis:

**metatarsus primus varus**
- flexible type rigid type

**metatarsus adductus**
- flexible type
- rigid type

**metatarsus varus (one third of a clubfoot)**

**pes varus (two thirds clubfoot)**

**talipes equinovarus (clubfoot)**
- postural talipes equinovarus
- idiopathic talipes equinovarus
- Teratogenic talipes equinovarus
- neurological talipes equinovarus

**calcaneovalgus deformity**

**convex pes valgus (congenital talus verticalis)**
- isolated convex pes valgus
- teratogenic convex pes valgus
- neurological convex pes valgus

**skew foot (Z-foot, serpentine foot)**

Explanatory note: the incidence of most congenital foot deformities is 1–2 in 1000 live births, with exception in the case of convex pes valgus and the skew foot, which are rare anomalies occurring respectively 1 in 10,000 and 1 in 50,000 live births.

**Metatarsus primus varus** The outer four metatarsals are parallel with normal spaces in between. The first metatarsal is extremely adducted with respect to the rest of the foot. There is a strikingly great space between the big toe and the second toe (Fig. 13.1). Normally on an anteroposterior X-ray of the foot there is an angle of 7° between the first and second metatarsals. An angle greater than 10° is abnormal. The deformity is an inborn error, often familial, usually bilateral and appears more often in girls than in boys. There is a distinction between a flexible and a rigid metatarsus primus varus.

Flexible type. A flexible metatarsus primus varus is only present when the child stands and not in a non weight bearing foot. This is often called a searching toe and is caused by an overactive abductor hallucis muscle (Fig. 13.1). This corrects itself spontaneously.

Rigid type. In this case the deformity remains in a non weight bearing foot. A hallux valgus will develop during puberty if this deformity remains untreated.

**Metatarsus adductus** In this case the forefoot is adducted with respect to the midfoot. We see no deep creases on footsole inspection (Fig. 13.2). The metatarsus adductus can usually be passively corrected by the examiner.


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_13

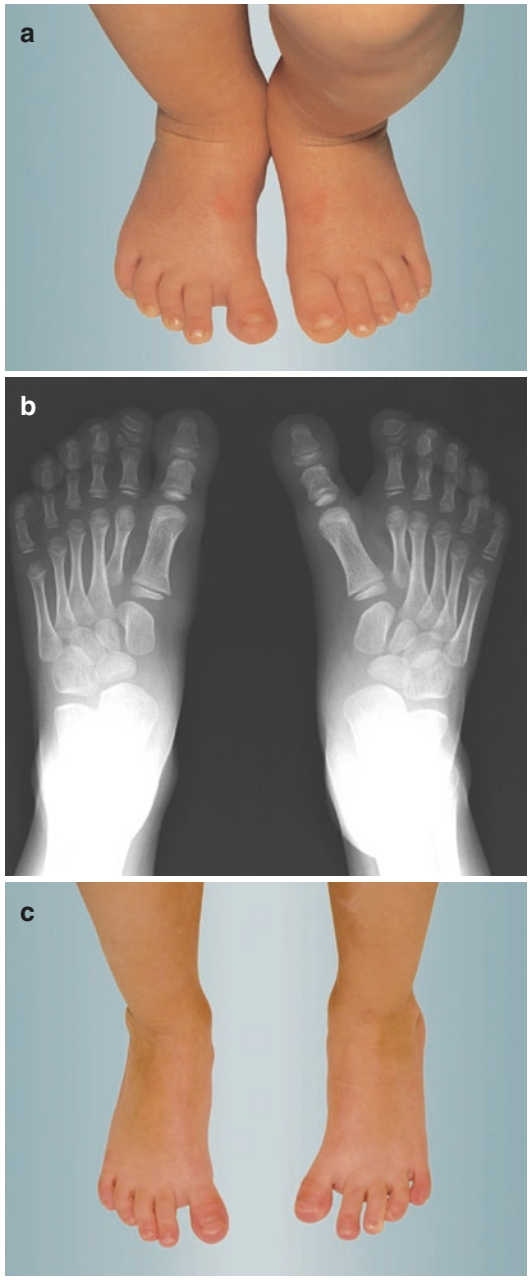



**Fig. 13.1** (**a**) A right sided rigid metatarsus primus varus. The first metatarsal together with the big toe is adducted with regard to the rest of the foot. The space between the big toe and the second toe is conspicuous. (**b**) Anteroposterior X-ray of the feet. In a metatarsus primus varus the intra-metatarsal angle between the first and second metatarsal is greater than 10°. (**c**) Bilateral flexible metatarsus primus varus. On weight bearing the big toes are medially displaced. The big toes join the rest of the toes when the feet are non weight bearing. These are often called "searching toes"

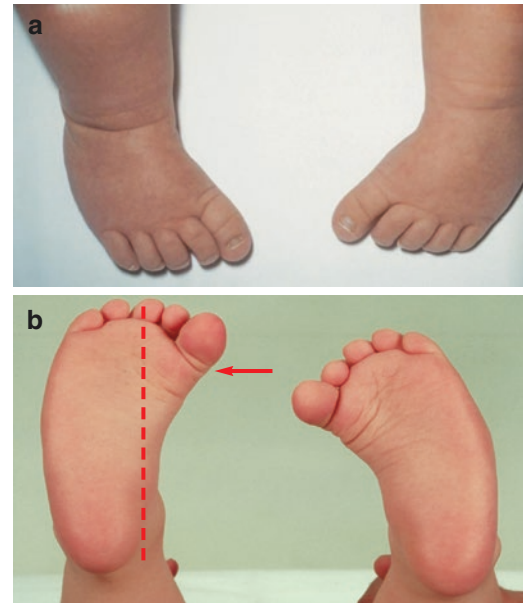



**Fig. 13.2** (**a**) Bilateral metatarsus adductus. The forefoot is in adduction. (**b**) On inspection of the footsole we do not see a deep crease. The footsole is smooth. If the forefoot can be brought into abduction relative to the hindfoot (beyond the *dotted line*) then correction of the metatarsus adductus will be spontaneous

Flexible type. The forefoot can be brought into abduction with respect to the posterior part of the foot in 90% of cases. In this case the metatarsus adductus will be corrected spontaneously.

Rigid type. In the other cases the forefoot can just or just not be brought into a neutral position. Spontaneous correction does not occur in these cases

**Metatarsus varus** This deformity is often incorrectly called a rigid type of metatarsus adductus. The difference is not only that the mid- and forefoot is in adduction with respect to the hindfoot but also because the foot is in supination (Fig. 13.3). There is a deep crease at the level of the transition between the mid- and hindfoot (Fig. 13.3). This deformity will not spontaneously correct itself. The child walks on the lateral side of the foot.

**Pes varus** The mid- and forefoot is in adduction and supination and the hindfoot has a heel varus (Fig. 13.4). Dorsal extension is complete. The

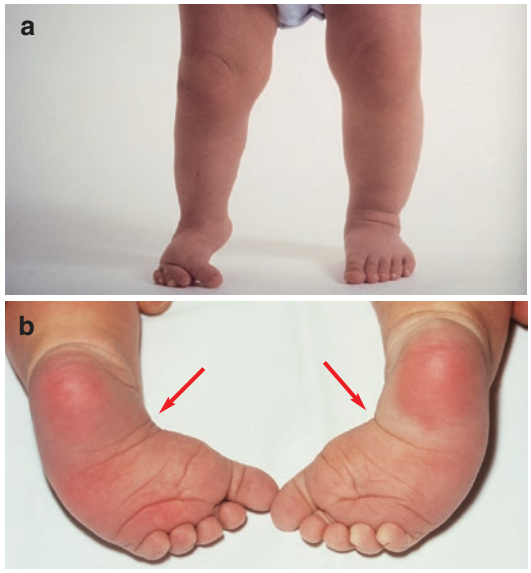



**Fig. 13.3** (**a**) Right metatarsus varus (one third clubfoot). The mid- and forefoot are in adduction and supination. (**b**) Bilateral metatarsus varus (one third clubfoot) seen from the plantar side. A deep crease can be seen in the transition between the midfoot and hindfoot (see *arrows*) (see also the difference with Fig. 13.2b)

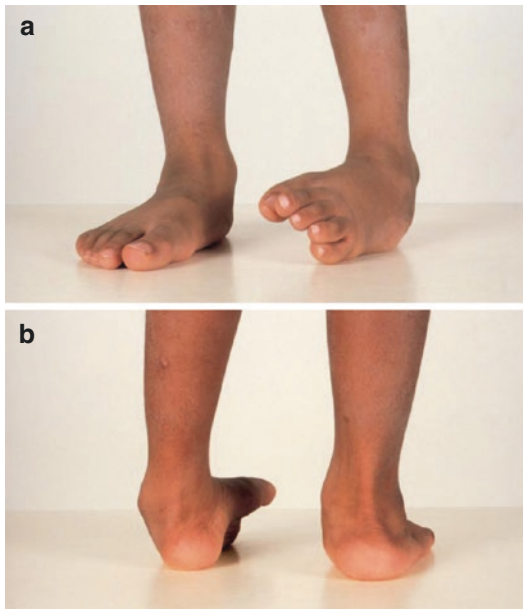



**Fig. 13.4** Left pes varus (two thirds clubfoot). The mid- and forefoot are in adduction and supination and the hindfoot has a varus angulation. Dorsiflexion is normal. (**a**) Anterior view. (**b**) Posterior view

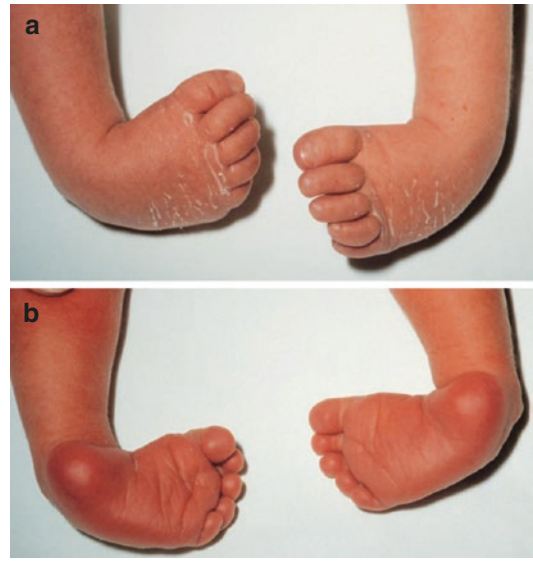



**Fig. 13.5** (**a**) Bilateral idiopathic talipes equinovarus (clubfoot) seen dorsally. The mid- and forefoot are adducted and supinated and the hindfoot has a heel equinus and varus deformity. (**b**) The same clubfeet as in Fig. 13.5. A seen from the plantar side

child walks completely on the lateral side of the foot if the problem is untreated.

**Talipes equinovarus** The mid- and forefoot is in adduction and supination and the hindfoot has a heel equinus and varus deformity (Fig. 13.5). If the foot is turned inwards in newborns it is sometimes difficult to tell if we are dealing with normal feet or if one or both feet are abnormal. A child can for instance hold the foot or feet in a preferential clubfoot position. In order to check this one can tickle the lateral foot border and if this foot is actively everting we are dealing with a normal foot (Fig. 13.6).

The child will walk continuously on the outside of the foot if the clubfoot remains untreated.

An untreated clubfoot looks like a golfclub (Fig. 13.7). A clubfoot can be divided into 4 types:

Postural talipes equinovarus. This is a flexible deformity which can be brought into a neutral position directly after birth (Fig. 13.8).

Idiopathic talipes equinovarus. This is a clubfoot in which the cause is unknown. This is the

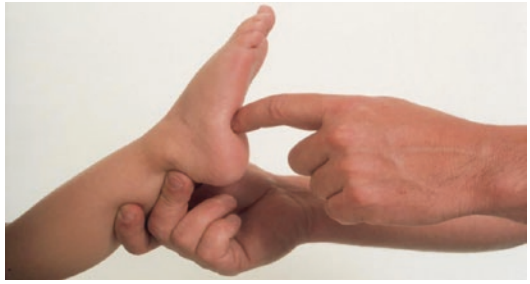


**Fig. 13.6** There is a normal foot position if the foot actively achieves a normal position after the lateral foot border has been tickled. There is a positional deformity if this is not the case

**Fig. 13.7** An untreated talipes equinovarus (clubfoot) looks like a golfclub (From: Handbuch der Orthopädische Chirurgie, prof. dr. Joachimstahl, 1905)

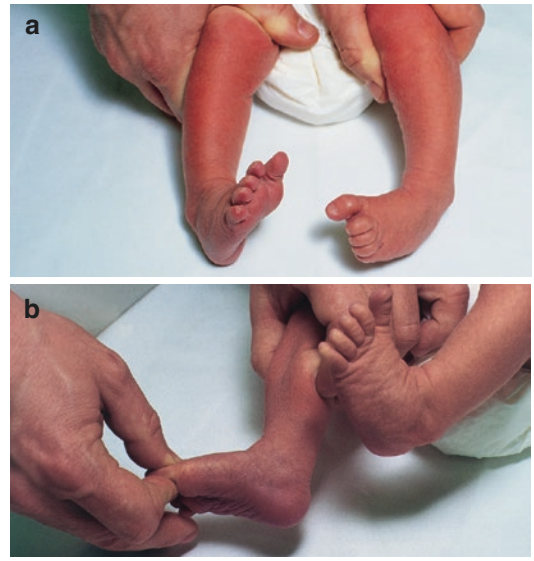



**Fig. 13.8** (**a**) A postural talipes equinovarus. (**b**) This is a flexible deformity which can be brought into a neutral position directly after birth

**Table 13.1** Syndromes with a pes equinovarus

| |
|---|
| Arthrogryposis multiplex congenita |
| Cranio-carpo-tarsal syndrome |
| Diastrophic dwarf growth |
| Larsen syndrome |
| Möbius syndrome |

See Appendix for details of these syndromes

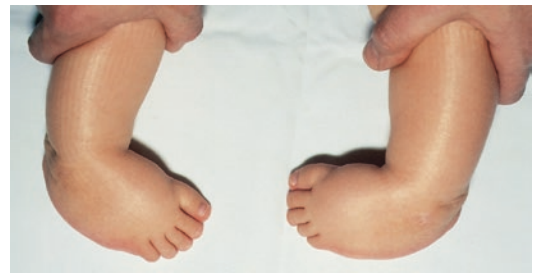


**Fig. 13.9** Teratogenic clubfoot. These feet are generally shapeless and very stiff

most common type and varies in rigidity. The degree of rigidity can be classified with a scoring system such as that of Dimeglio and Pirani.

Teratogenic talipes equinovarus. This clubfoot is associated with a syndrome (Table 13.1). These feet are generally rigid (in Greek teras means monster and genan is production) (Fig. 13.9).

Neurological talipes equinovarus. This is particularly associated with spina bifida. The clubfoot is usually rigid in these cases. An idiopathic clubfoot is seen in 1 or 2 per 1000 live births. Incidence between boys and girls is 3:1 and in 40 % of the cases bilateral. Parents without a clubfoot or feet who have a child with a clubfoot or feet have a 2.5–6.5 % chance that their next child will also have a clubfoot or feet.

If one of the parents has idiopathic clubfeet, then the chances that their child will be born with clubfeet is 10–25 %. In monozygotic

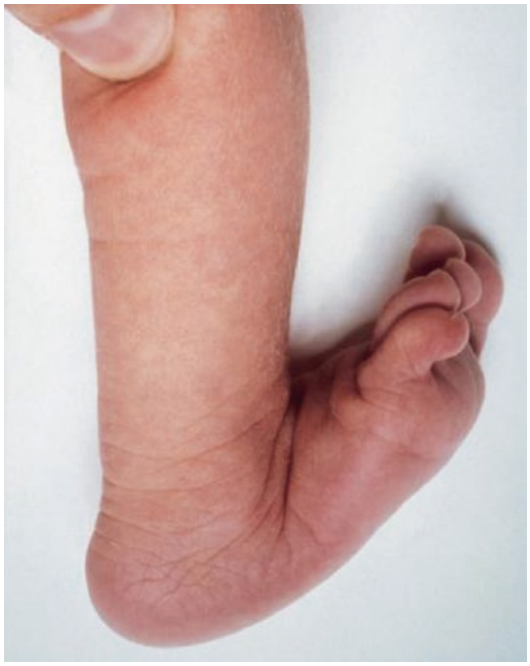


**Fig. 13.10** Calcaneovalgus deformity. The hindfoot is in extreme dorsal extension with a light valgus angulation

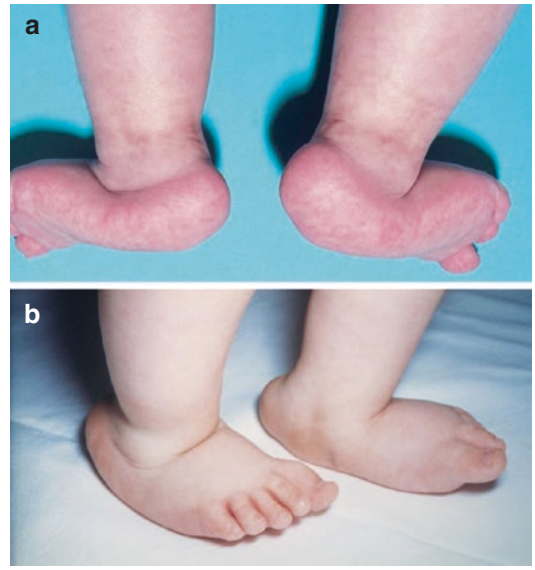



**Fig. 13.11** (**a**) A bilateral convex pes valgus in a newborn. (**b**) Bilateral convex pes valgus (untreated) when the child starts to walk. The mid- and forefoot is fixed in abduction and dorsalflexion and the hindfoot has a heel equinus and valgus deformity. The footsole is convex

(identical) twins the chance that both children will have clubfeet is 32.5 % and in dizygotic twins 3 %.

**Calcaneovalgus deformity** The forefoot has a neutral position. The hindfoot has extreme dorsiflexion, with a slight heel valgus (Fig. 13.10). If the foot can be manually manipulated into slight plantar flexion the deformity will spontaneously be corrected within 3 months. If this is not possible then the deformity will persist. In this case the deformity is part of a generalized problem such as in spina bifida.

**Convex pes valgus** The mid- and forefoot is fixed in abduction and dorsiflexion and the hindfoot has a heel equinus and valgus deformity. The footsole is convex (Fig. 13.11). In some cases this deformity is difficult to differentiate from a calcaneovalgus deformity directly after birth. One must have a good look at the footsole. These are flat in the case of a calcaneovalgus deformity and in a convex pes valgus it will be convex. The heel sticks out posteriorly and it looks as if the lower leg is implanted into the middle of the foot. A convex pes valgus is also much stiffer than in calcaneovalgus deformity. There is a dorsolateral dislocation in the talonavicular joint in convex pes valgus. There are three types:

Isolated convex pes valgus. This is unilateral and the least rigid.

Teratogenic convex pes valgus. This is always combined with other inborn deformities such as hip dislocation, hand deformities and a contralateral clubfoot. This type is often familial. The deformity is rigid and usually bilateral.

Neurological convex pes valgus. This type is associated with spinal canal abnormalities such as in spina bifida, sacral agenesis and diastematomyelia. The rigidity varies in this group.

**Skew foot** This is also referred to as a Z-foot or serpentine foot. This is a complex deformity in which the forefoot is in adduction, the midfoot is in abduction and the hindfoot is in equinus and valgus (Fig. 13.12). Sometimes there will be a tarsal coalition (see pp. 279–285).

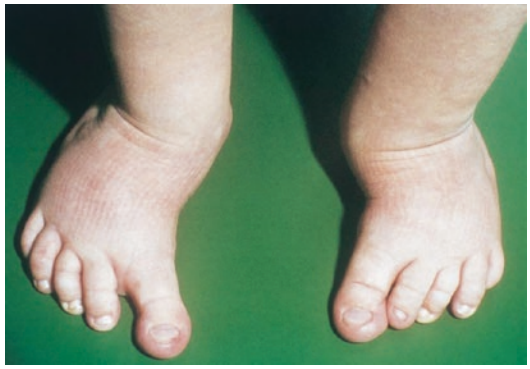


**Fig. 13.12** Skew foot. This is a complex positional deformity in which the forefoot is in adduction, the midfoot in abduction and the hindfoot in valgus and equinus

Supplementary assessment: in the first instance no further studies are necessary in the case of metatarsus primus varus, metatarsus adductus, metatarsus varus, pes varus and calcaneovalgus deformity. It is wise to carry out X-rays in two directions in case of other defomities. In a normal foot the angle between the talus and calcaneus (T.C.-angle = talocalcaneal angle) should be between 20° and 40° in both the anteroposterior and the lateral X-rays. In a clubfoot this angle is much smaller and the longitudinal axes of the talus and calcaneus are often more or less parallel to each other (Fig. 13.13). The angle between the talus and calcaneus in a convex pes valgus is much greater and the longitudinal axes of talus and calcaneus are more or less perpendicular to each other and that is why it is also called congenital talus verticalis (Fig. 13.14). In a skew foot the longitudinal axis of the hind-, mid- and forefoot on the anteroposterior X-ray show a Z-shape (Fig. 13.15). That's why the deformity is also known as a Z-foot.

It is wise to do an ultrasound or an anteroposterior X-ray of the pelvis because these foot deformities have a high incidence of developmental dysplasia of the hip except in the case of metatarsus primus varus.

Primary care treatment: a wait and see approach should be taken in the case of a flexible metatarsus primus varus or metatarsus adductus in which the forefoot can be manually brought into abduction with respect to the posterior part of the foot and in the case of a postural talipes equinovarus. These deformities will be spontaneously corrected before the fourth year of age. A pes calcaneovalgus corrects itself within 3–6 months of age.

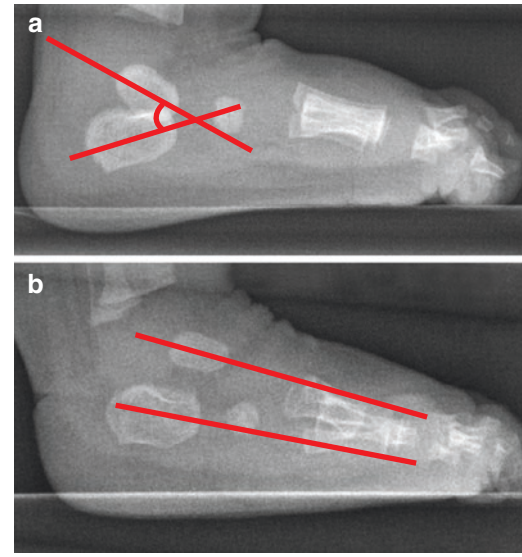



**Fig. 13.13** Lateral X-ray. (**a**) This is a normal foot with a normal angle between the talus and calcaneus. (**b**) Clubfoot, the longitudinal axis of the talus and calcaneus on the right side are almost parallel

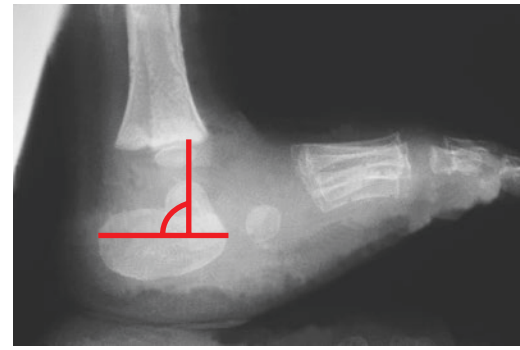


**Fig. 13.14** Lateral X-ray of a convex pes valgus. The longitudinal axis of the talus is more or less perpendicular to the axis of the calcaneus and that is why it is also called a talus verticalis

When to refer: all other foot deformities must be referred.

Secondary care treatment: **metatarsus primus varus.** A rigid metatarsus primus varus is treated with manual reduction and lower

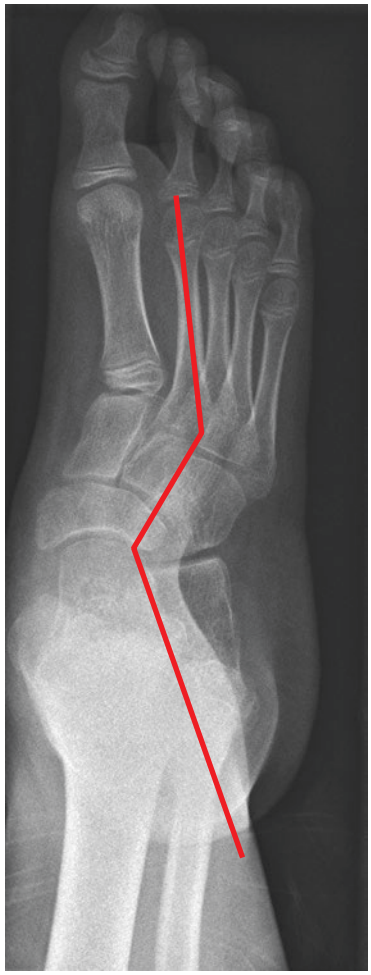


**Fig. 13.15** Anteroposterior X-ray of a skew foot. The longitudinal axes of the hind-, mid- and forefoot have a Z-shape

leg plaster immobilization. If a normal position is achieved then the child will be treated with a ankle-foot orthosis during the night for 1 year (Fig. 13.16).

**Metatarsus adductus** In a rigid metatarsus adductus in which the forefoot can be brought into a neutral position, an ankle-foot orthosis should be worn at night for 1–2 years (with the foot in the corrected position). The foot will be manually corrected and immobilized with a long-leg plaster cast if the forefoot cannot be manipulated into a neutral position. Once the medial foot border is straight, an ankle-foot orthosis will be applied for 1–2 years at night (Fig. 13.17).

**Fig. 13.16** Ankle-foot orthosis

A release of the abductor hallucis muscle and a capsulotomy of the first tarsometatarsal joint will be carried out if operative treatment is necessary.

In children up to the age of 5 a capsule release of the tarsometatarsal and intermetatarsal joints should be considered if the above mentioned treatment is insufficient. (Heyman-Herndon procedure) (Fig. 13.18). A dorsal subluxation of the Lisfranc joint is a risk after this treatment. A crescentic shaped osteotomy at the level of the base of the metatarsal bones may be carried out in children older than 5 years who have a persistent rigid metatarsus adductus (Fig. 13.19).

**Metatarsus varus and pes varus** The treatment is more or less the same as in talipes equinovarus. However, in a metatarsus varus, an equinus and varus deformity of the hindfoot need not be operatively corrected and the same applies to the equinus deformity of the hindfoot in a pes varus. Operative treatment is generally not necessary and one can treat these with serial manual corrections followed by a short-leg plaster cast.

**Talipes equinovarus** The initial treatment of clubfeet is conservative (not operative) and consists of manual correction and long-leg plaster

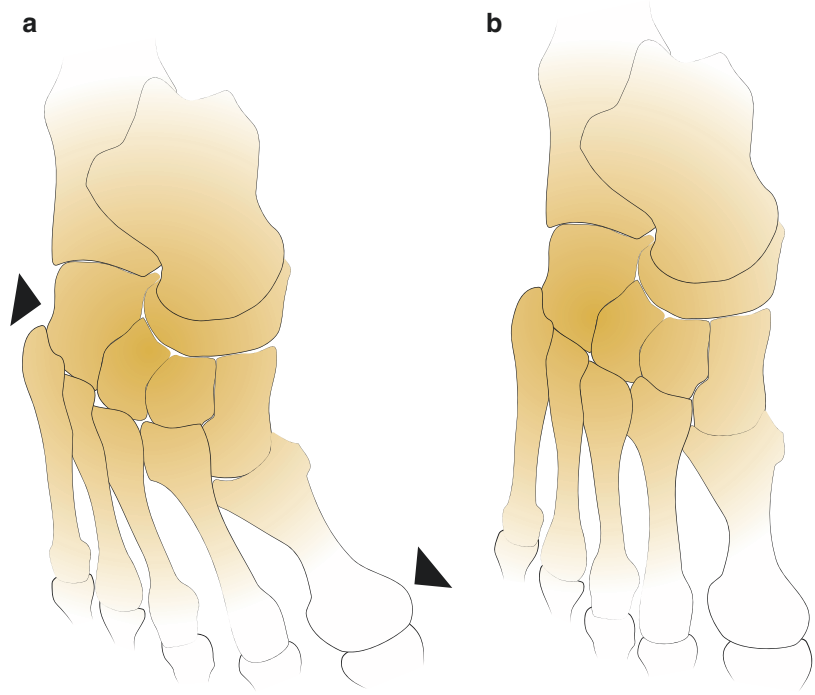



**Fig. 13.17** Manual correction of a metatarsus adductus. (**a**) Sideways pressure on the cuboid with the thumb of one hand and sideways pressure on the head of the first metatarsal with the other. (**b**) The final position achieved

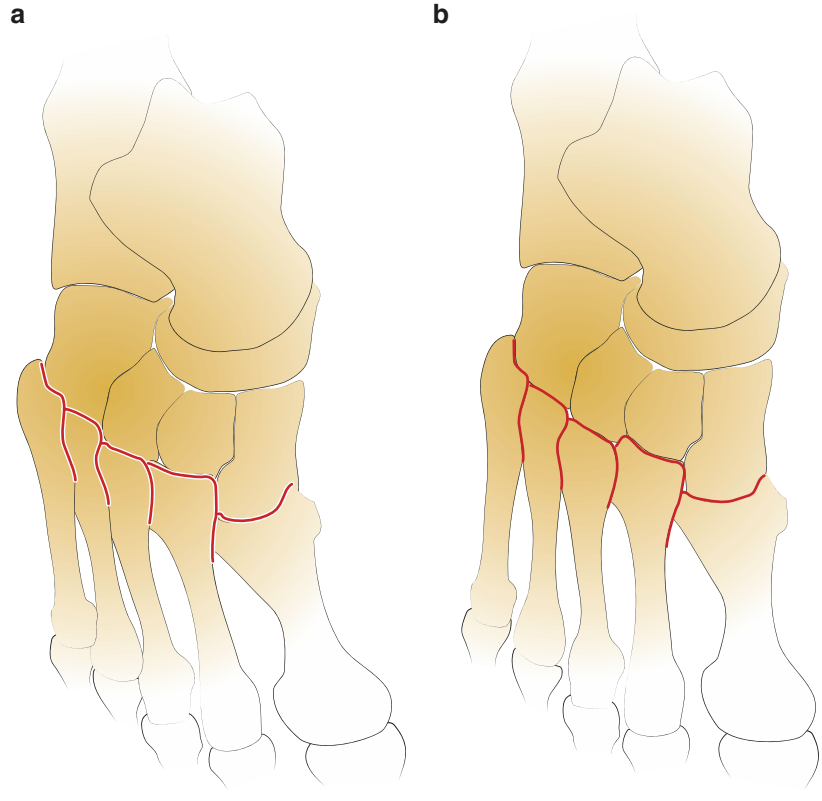



**Fig. 13.18** Heyman-Herndon procedure for metatarsus adductus. (**a**) Release of the joint capsules of the tarsometatarsal and intermetatarsal joints (*red*). (**b**) Corrected position

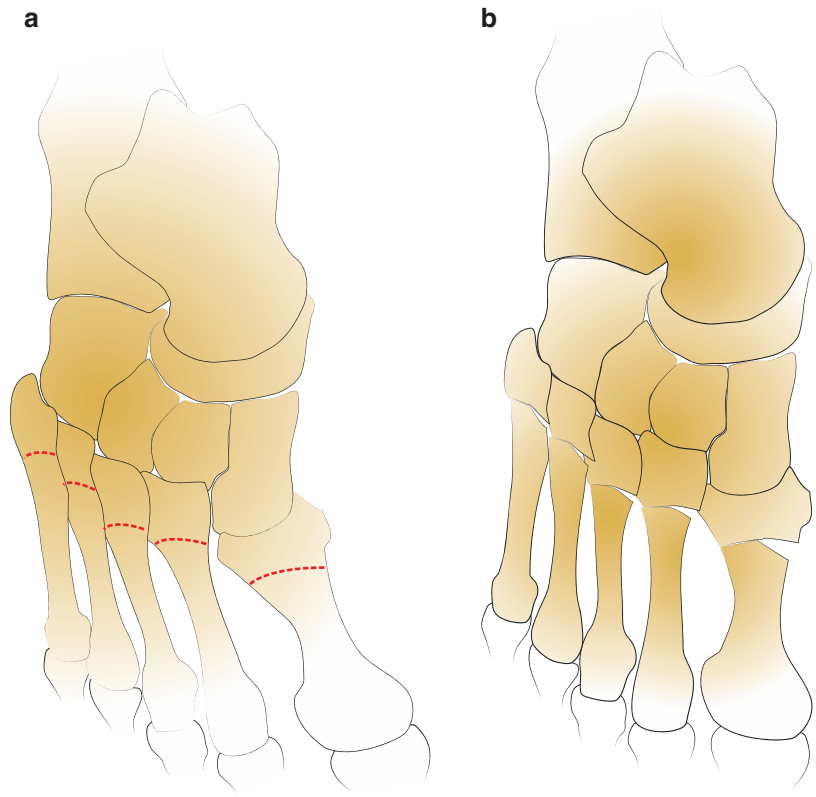



**Fig. 13.19** (**a**) Crescentic osteotomies (*red dotted lines*) at the level of the base of the metatarsals for rigid metatarsus adductus. (**b**) Corrected position

cast immobilization. There are two methods for treating clubfeet.

**Kite method** In the Kite method the center of rotation in the midfoot is at the level of the joint between the calcaneus and cuboid. In the first instance the forefoot adduction and supination is treated with manipulations and long-leg plaster cast immobilization, followed by correction of the varus deformity of the hindfoot and finally correction of the equinus position of the hindfoot. When correcting the equinus deformity one must take care that correction is carried out in the hindfoot and not in the midfoot, otherwise the footsole will develop a convex shape, the so-called "rocker bottom foot" (Fig. 13.20). The chances of success using this technique varies from 15 to 50 %. Between the ages of 4 and 12 months evaluation will establish whether the conservative treatment has been sufficient. If this is not the case then a so-called "posteromedial release à la carte" will be carried out. This is an operation in five stages in which the following procedures will be carried out (Fig. 13.21):

1. The hindfoot equinus,
2. The hindfoot internal rotation,
3. The hindfoot varus,
4. The adduction deformity at the level of the Chopart joint,
5. The adduction position at the level of the tarsometarsal joint (Lisfranc joint).

In a posteromedial release the posterial tibial nerve and vessels must be localized and held on one side. After that the following structures can be lengthened or divided:

1. The equinus position of the posterior foot is corrected by Z-shaped achilles tendon lengthening and division of the posterior capsule of the talocrural and subtalar joints. This is also called a posterior release.

2. The internal rotation of the calcaneus can be corrected by division of the calcaneofibular ligament and the posterior talofibular ligament.
3. The varus position of the hindfoot can be corrected by dividing the superficial deltoid ligament on the medial side of the subtalar joint.
4. After that the adduction and supination at the level of the Chopart joint is corrected by lengthening the posterior tibial tendon, division of the talonavicular joint capsule on the medial and plantar side and if necessary the joint capsule between the calcaneus and the cuboid on the plantar side.
5. Finally the metatarsus adductus can be corrected by lengthening the abductor hallucis and releasing the first tarsometatarsal joint on the medial side.

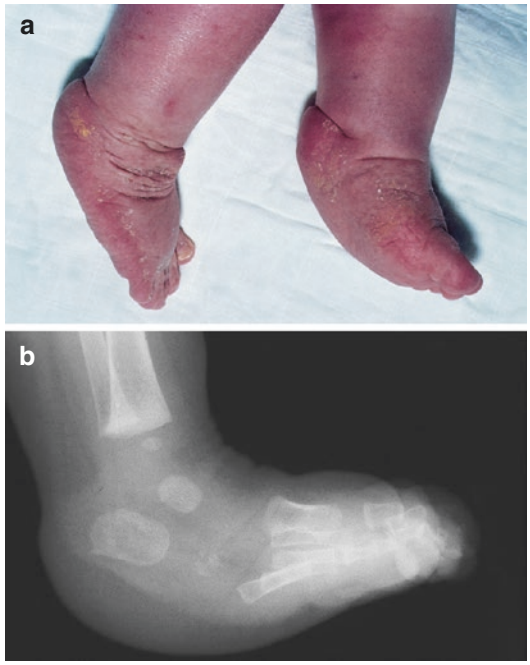



**Fig. 13.20** (**a**) Bilateral rocker bottom foot. (**b**) Lateral X-ray of a rocker bottom foot

After each of these five procedures one can assess the necessity for the following procedure or if a procedure can be omitted. Postoperative treatment of a posterior release is 6 weeks immobilization with a long-leg plaster cast and after a posteromedial or a medial release 12 weeks long-leg plaster immobilization. After this the child will wear an ankle-foot orthosis day and night until the child starts to walk, after which it will be worn only at night and depending on the tendency for recurrence from 2 to 5 years if necessary. One can stop the post-operative treatment with the ankle-foot orthosis if the child can actively dorsiflex and evert the foot and if there is no forefoot adduction. 

Overcorrection is a risk after this operation as a result of a too extensive release. As a result a flatfoot may develop which can be difficult to

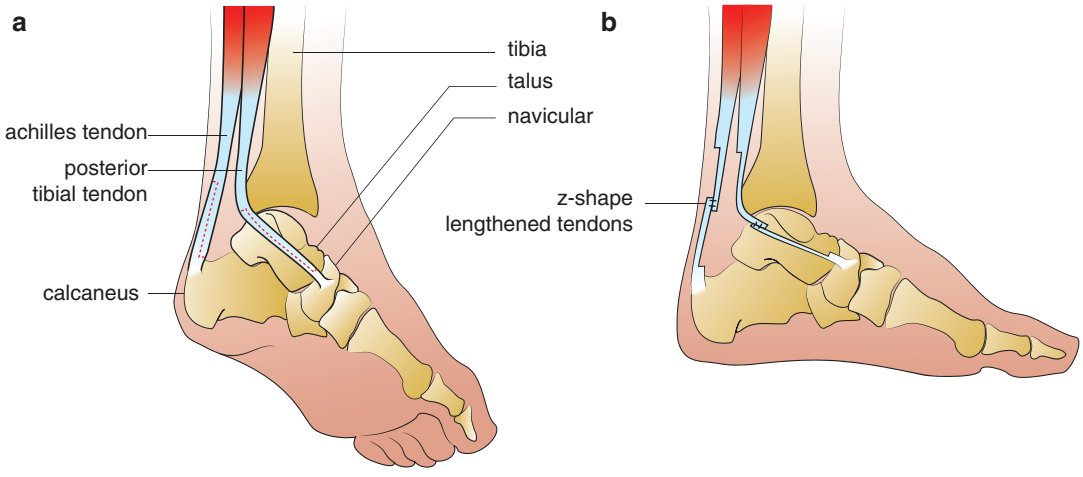



**Fig. 13.21** (**a**) In a posteromedial release the achilles tendon and tibialis posterior tendon are lengthened. (**b**) See the above text regarding the extensive operative procedures

treat, in which case a triple arthrodesis must be carried out (Fig. 13.22). Another complication is dorsiflexion of the first metatarsal bone as a result of overactivity of the anterior tibial muscle with respect to the peroneus longus muscle with plantar flexion of the first metatarsophalangeal joint as a result with hyperextension in the interphalangeal joint. A hallux flexus is the result. It looks as if there is a swelling on the dorsal side of the head of the first metatarsal (Fig. 13.23). Wearing shoes can be painful.

**Ponseti method** Correction of the midfoot using the Kite technique involves rotation around the joint between the calcaneus and cuboid. In the Ponseti technique the foot is rotated around the talus. In the first instance the first ray is brought into dorsiflexion so that the plantar aponeurosis will be stretched and the talonavicular joint will be more mobile making further correction possible.

Next, the foot is abducted in supination with one hand, whilst the talus is stabilized with the thumb

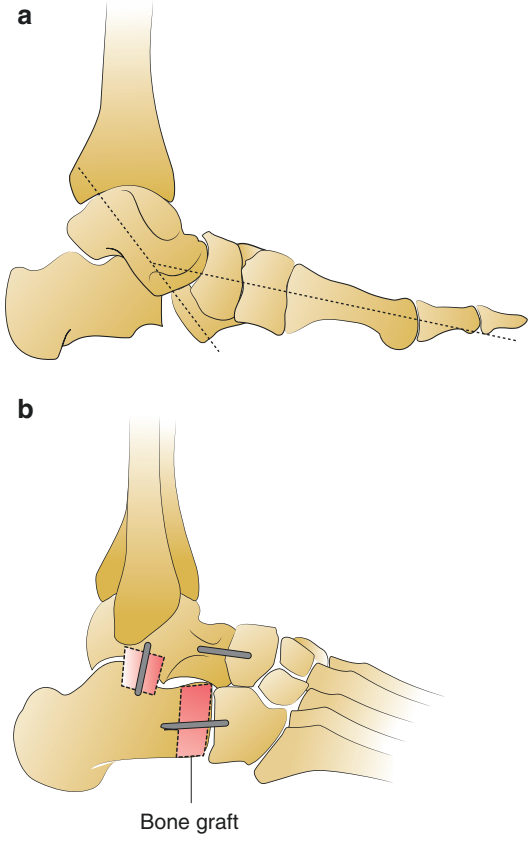



**Fig. 13.22** (**a**) Overcorrection in carrying out the posteromedial release may result in a flatfoot which is difficult to treat. (**b**) If really necessary a triple arthrodesis may be carried out in which the lateral foot border will be lengthened and the subtalar joint raised up and brought into a neutral position

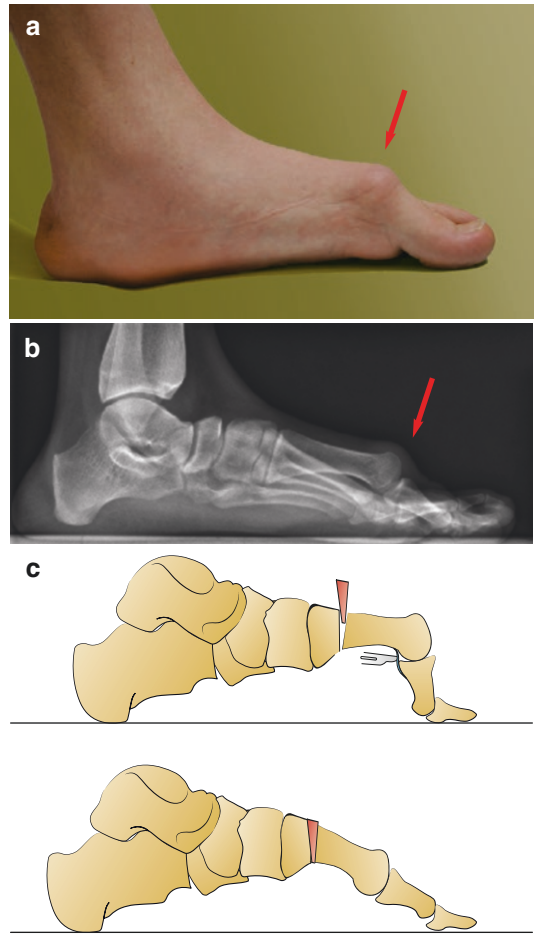



**Fig. 13.23** (**a**) The hallux flexus (flexed big toe) as a result of domination of the anterior tibial muscle activity compared to that of the peroneus longus. As a result the distal part of the first metatarsal is pulled up dorsally with compensatory plantar flexion of the first metatarsophalangeal joint and hyperextension in the interphalangeal joint. It looks as if the distal part of the first metatarsal has a swelling (*arrow*). (**b**) Lateral X-ray of a hallux flexus. (**c**) Schematic representation of the operative correction of a hallux flexus. The treatment consists of a flexion osteotomy in the proximal part of the first metatarsal bone, division of the joint capsule of the metatarsophalangeal joint on the plantar side, lengthening of the tendon of the flexor hallucis longus and cleavage or dorsal transposition of the flexor hallucis brevis tendon. The dominant anterior tibial tendon will be lengthened or transposed to the second ray

using the other hand. The supination and adduction of the forefoot and the varus position of the hindfoot is corrected in this way. If these components have been corrected then an effort will be made to correct the equinus position of the hindfoot (Fig. 13.24). In 90% of cases a percutanious achilles tendon tenotomy is necessary at 6 weeks of age (Fig. 13.25). After serial manual corrections and longleg plaster cast immobilization and a possible tenotomy of the achilles tendon a Dennis Brown bar or a similar bar will be applied that will be worn day and night until the child starts to walk after which it will be worn up to 3–4 years of age only at night (Fig. 13.26). In 30% of cases an adduction/supination deformity persists in the forefoot in which case lateral reinsertion of the tendon of the anterior tibial muscle will be carried out between 3 and 4 years of age in order to correct the deformity (Fig. 13.27). In case of a idiopathic talipes equinovarus most surgeons prefer the Ponseti technique to the Kite method.

A teratogenic and neurological talipes equinovarus are usually corrected operatively.

A correction can be carried out by repeating the lengthening of the achilles tendon and capsulotomies on the posterior side of the talocrural and subtalar joints if there are still elements of the equinus foot at a later age. The varus position in the hindfoot can be corrected between the 3rd and 10th year of life by carrying out a lateral closed wedge osteotomy of the calcaneus according to Dwyer (Fig. 13.28). An extra long lateral foot border in which the foot has the shape of a bean

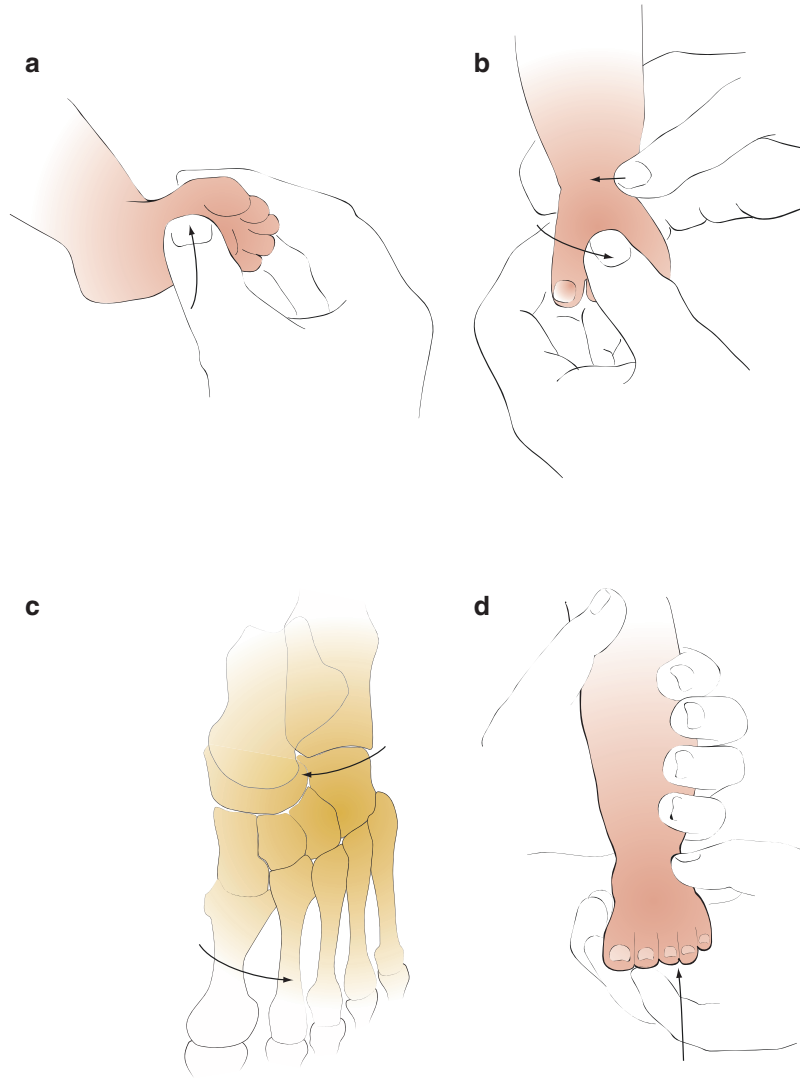



**Fig. 13.24** Ponseti clubfoot correction technique. Hereby we have a mnemonic namely CAVE in which the C stands for cavus, the A for adduction, the V for varus and the E for equinus. (**a**) In the first instance the cavus position must be corrected by bringing the first metatarsal bone into dorsiflexion. (**b**, **c**) The foot in supination will be abducted while the surgeon stabilizes the talus with the thumb of the other hand. As a result the foot rotates around the talus by which the adduction/supination in the mid- and forefoot and the varus deformity in the hindfoot will be corrected. (**d**) The equinus deformity of the hindfoot will be finally corrected

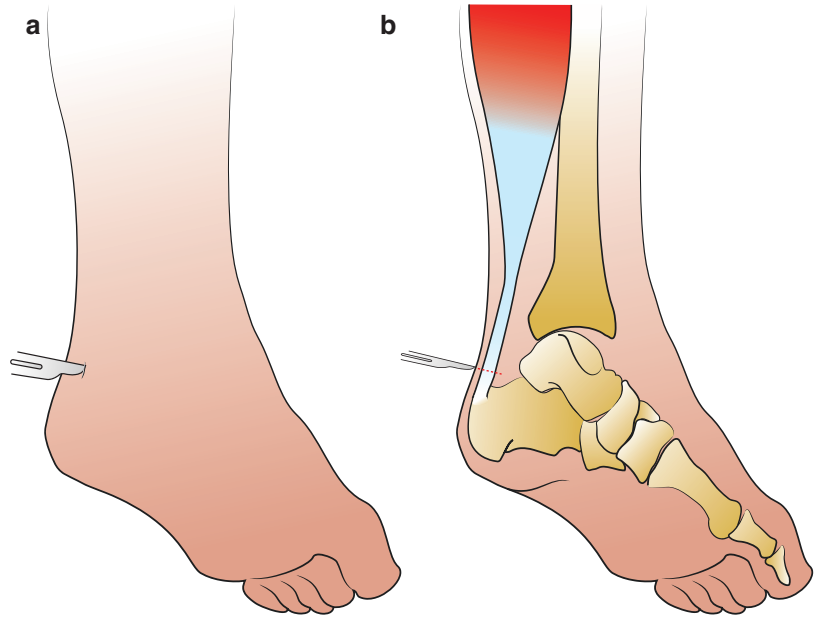



**Fig. 13.25** Percutaneous achilles tendon tenotomy

**Fig. 13.26** Modified Dennis Brown bar

can be corrected between 4 and 8 years of age by shortening the lateral border according to the Evans technique (Fig. 13.29). If a medial release has not been carried out, then the above mentioned procedure must be combined with the Evans operation. A possible remaining metatarsus adductus can be treated from the age of 5 years by carrying out osteotomies at the level of the metatarsal bases (Fig. 13.19). If all of the components of the clubfoot are still present at an older age a corrective triple arthrodesis may be carried out after the age of 12. In this case an arthrodesis is carried out in the subtalar joint, the calcaneocuboid joint and in the talonavicular joint.

**Convex pes valgus** In convex pes valgus there are also two available techniques just as in clubfoot treatment.

First method. In the first method one begins with serial manual corrections and immobilization in a long-leg plaster cast. In the first instance one tries to correct the forefoot by fixing the talus and calcaneus with one hand and with the other hand bringing the forefoot into adduction and plantar flexion. Secondarily one tries to correct the equinus in the hindfoot. Very occasionally a convex pes valgus can be corrected using serial manual corrections and plaster cast immobilization, but this is exceptional.

In most cases operative correction is required. This will preferably not be carried out during the first year of life because the extensor

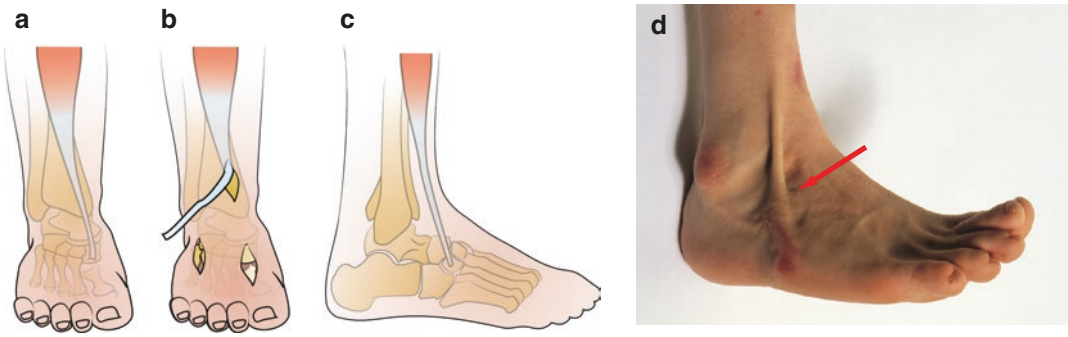



**Fig. 13.27** (**a**–**c**) The complete insertion of the anterior tibial muscle tendon will be transposed laterally in the case of a persistent adduction/supination deformity. (**d**) The arrow points to the transposed tibialis anterior tendon

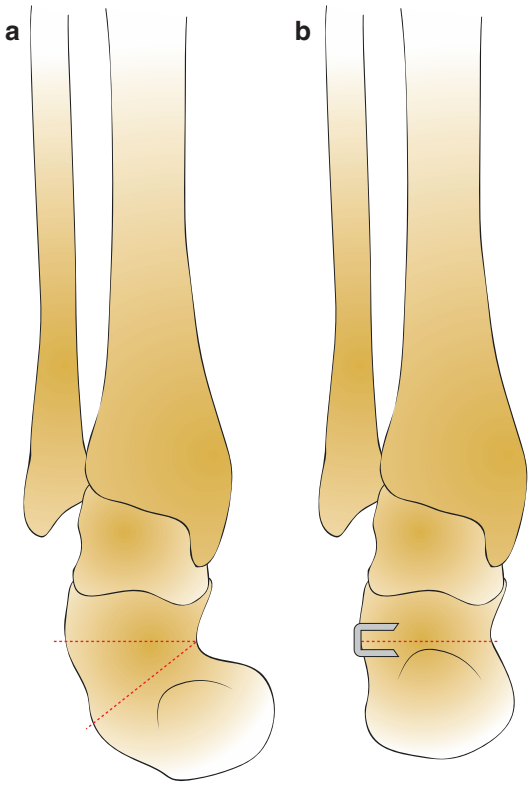



**Fig. 13.28** Lateral closed wedge osteotomy of the calcaneus according to Dwyer for a persistent varus position of the hindfoot. (**a**) Planned osteotomy for wedge excision (*dotted line*). (**b**) Closed wedge osteotomy fixed with a staple

tendons to the toes are so thin that a Z-shaped lengthening is almost impossible. A soft tissue operation can be carried out between the 1st and 3rd year of age. First of all the origin of the extensor digitorum brevis muscle must be divided, after which a capsulotomy is carried out on the dorsal side of calcaneocuboid joint and on the dorsal side of the talonavicular joint through an incision in the tarsal sinus. The tendon of the anterior tibial muscle is divided through an incision on the medial side of the foot. After this the Chopart joint will be repositioned and fixed with a Kirschner wire through the talonavicular joint.

The anterior tibial muscle tendon will be reinserted at the level of the talus. If necessary the interosseous talocalcaneal ligament may be divided in order to be able to transpose the subtalar joint. Furthermore, the achilles tendon will be lengthened and a capsulotomy will be carried out on the posterior side of the talocrural and subtalar joints. If the peroneus longus and brevis muscles, the extensor hallucis longus tendon and the extensor digitorum longus tendons are too tight, they will be lengthened using the Z-technique. Aftercare involves 12 weeks of long-leg plaster cast immobilization, followed by a anklefoot orthosis for the night for 2–5 years. If the hindfoot remains in valgus in children from 4 to 8 years of age an extraarticular subtalar arthrodesis can be carried out. Children of 12 years of age and older who still have a persistent deformity are best treated with a corrective triple arthrodesis.

If treatment begins in children older than 3 years of age it is often necessary to remove the navicular bone in order to reposition the Chopart joint.

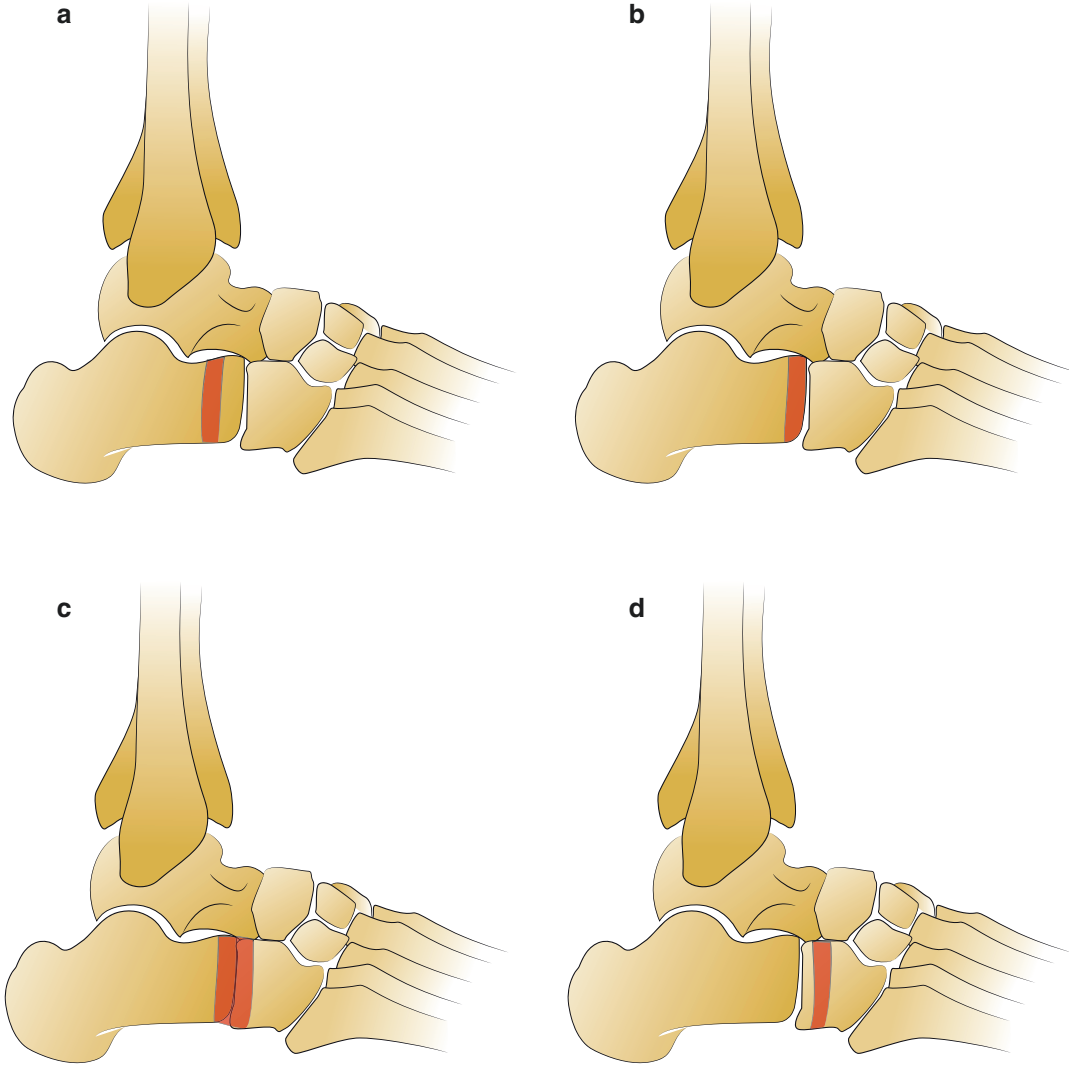



**Fig. 13.29** Shortening of the lateral border of the foot according to Evans with a closed wedge shaped resection with the base of the wedge on the lateral side. (**a**) At the level of the anterior part of the calcaneus. (**b**) At the level of the anterior part of the calcaneus including removal of the cartilage surface from the cuboid. (**c**) At the level of calcaneocuboid joint. (**d**) At the level of the cuboid

Second method. This is a sort of reversed Ponseti technique. The foot will be brought into plantar flexion and inversion using one hand and by using the thumb of the other hand to give counter pressure to the head of the talus at the level of the footsole. The foot then assumes a clubfoot appearance and is immobilized in a long-leg plaster cast. This will be repeated every week for 6 weeks. After 6 weeks a lateral X-ray of the foot will be taken to see if the talus is in line with the navicular bone. If the navicular bone is not ossified the longitudinal axis of the talus should be in line with the axis of the first metatarsal bone. Ossification of the navicular bone occurs in girls between 1 and 3 years of age and in boys between 3 and 5 years of age. If the talus is in line with the navicular bone the talonavicular joint will be operatively immobilized using a Kirschner wire. If this is not the case one tries to improve the position under anesthetic before immobilizing the talonavicular joint with a Kirschner wire. If this is still not possible then a

small medial incision is made and a dorsal capsulotomy of the talonavicular joint is carried out. Occasionally the peroneal muscles and/or the posterior tibial tendon and/or the anterior tibial tendon should be fractionally lengthened at the level of the transition between tendon and muscle. Once the talonavicular joint has been stabilized with a Kirschner wire then a percutaneous tenotomy is carried out 1 cm proximal to the insertion of the achilles tendon. The foot is immobilized with a long-leg plaster cast postoperatively with the foot in 5° of dorsiflexion. After 3 weeks a new plaster is applied with the foot in 10° of dorsiflexion. After 6 weeks the plaster cast and the Kirschner wire are removed. During this period an ankle-foot orthosis has been designed in which the foot is in 15° of plantar flexion at the level of the Chopart joint. The orthosis will be worn for 24 h a day until the child can stand and walk. After this the orthosis will be worn for some time only at night depending on the tendency for recurrence.

**Skew foot** Non operative treatment for a skew foot is very difficult. The valgus position of the hindfoot cannot be corrected with manipulation and cast immobilization. The forefoot adduction can be corrected but a disadvantage is that after correction of the forefoot adduction the valgus position in the hindfoot increases so that finally the child ends up with a severe flatfoot. After 5 years of age a lateral lengthening of the calcaneus can be carried out (Fig. 13.30) or an osteotomy to medialize the calcaneal tubercle (Fig. 13.31). Metatarsal osteotomies may be considered in the case of persistent adduction of the forefoot (Fig. 13.19).

## Toe Walking

Complaint: the child walks on the toes (Fig. 13.32).

Assessment: apart from bilateral limitation in dorsiflexion in the talocrural joints no other orthopedic or neurological anomalies are found. The ankle can generally be just brought passively into a neutral position.

Diagnosis: **habitual toe walker (idiopathic toe walking, equinus gait, congenital short achilles tendons)**

Explanatory note: **habitual toe walker**. Toe walking is a normal variation when a child starts to walk. A normal heel-toe-gait should develop 3 and 6 months after the child starts walking. However, due to shortening of the calf muscles, some children still continue to walk on the toes. Most of them can bring the heel onto the ground if requested by the examiner or parent. A normal heel-toe-gait can usually be demonstrated in children who do their best. However, they walk on their toes again if their attention is distracted. The deformity is always bilateral. The term habitual toewalker suggests that the children habitually walk on their toes which is not the case. The cause is unknown, with the exception of an accessory soleus. Toe walking is caused by shortening in the calf muscles. In 40 % of cases there is a positive family history. Older children are often plagued by other children because of toe walking. A broad forefoot will occur in children who have been toe walking for a long time and a painful callus may develop in adults at the level of the ball of the foot.

In most cases there is a contracture in the gastrocnemius muscle. Ankle dorsiflexion is limited with the knee extended and normal with the knee flexed.

In rare cases there is a contracture of the triceps muscles. It is not possible to bring the foot into a neutral position from plantar flexion. Dorsiflexion is limited regardless of the knee position. Operative correction by heel-cord lengthening may be necessary.

A rare congenital deformity is an accessory soleus in which the muscle body of the soleus extends to the ankle. There is a fullness on the medial side of the ankle, dorsiflexion is limited, regardless of the knee position. Operative lengthening may be necessary.

Neurological examination is completely normal. However, if there are lively reflexes and a

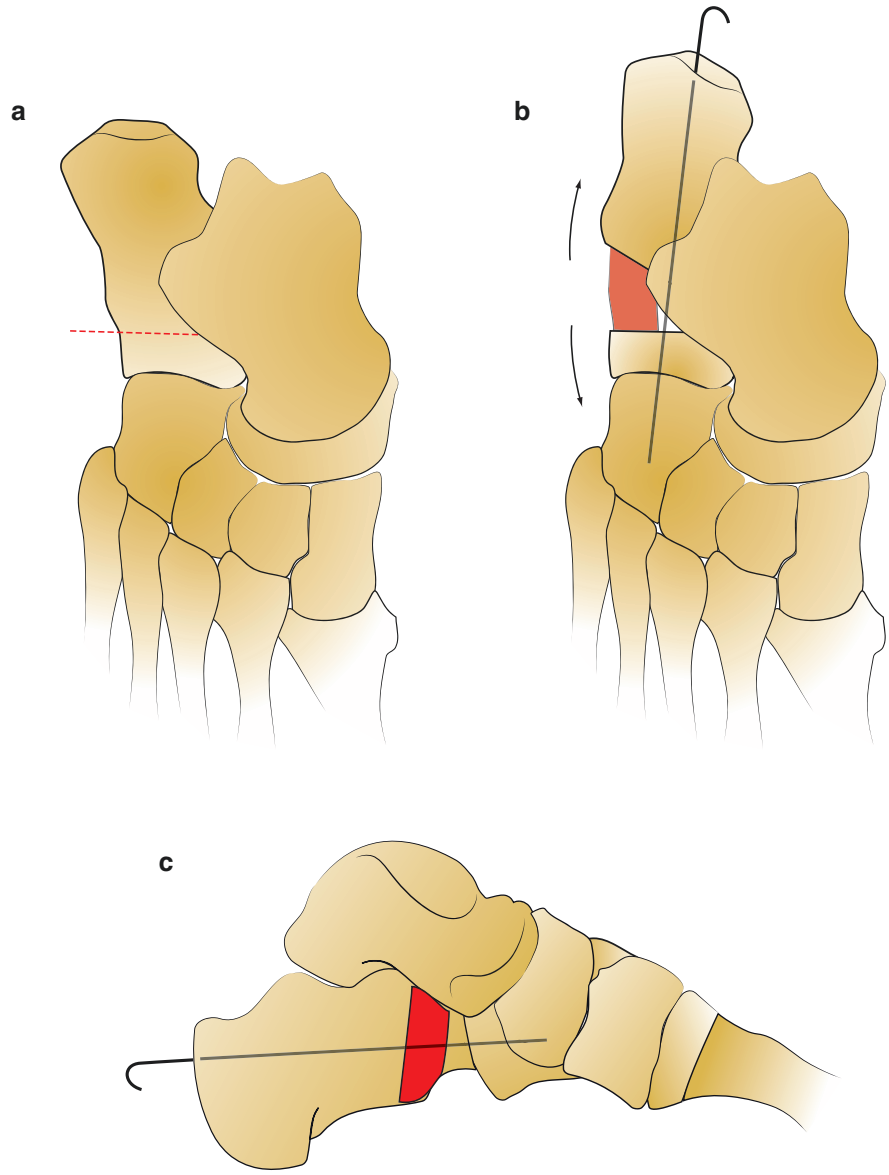



**Fig. 13.30** Lateral column lengthening of the calcaneus. (**a**) In this case a calcaneal osteotomy is performed 1 to 1.5 cm posterior to the calcaneocuboid joint. The osteotomy is performed between the anterior and middle part of the calcaneus. Then the osteotomy is opened up and a free cortical block of bone will be interposed and fixed with a Steinmann pin. (**b**) View from above. (**c**) View from the lateral side. The joint capsule on the medial side of the talonavicular and the naviculocuneiform joint may be reefed if necessary

clonus in the calf musculature can be provoked one should be aware of an encephalopathy (spasticity).

Supplementary assessment: none.

Primary care treatment: a wait-and-see policy in the first instance. Toe walking spontaneously improves in most children with time. This leads to a normal heel-toe gait.

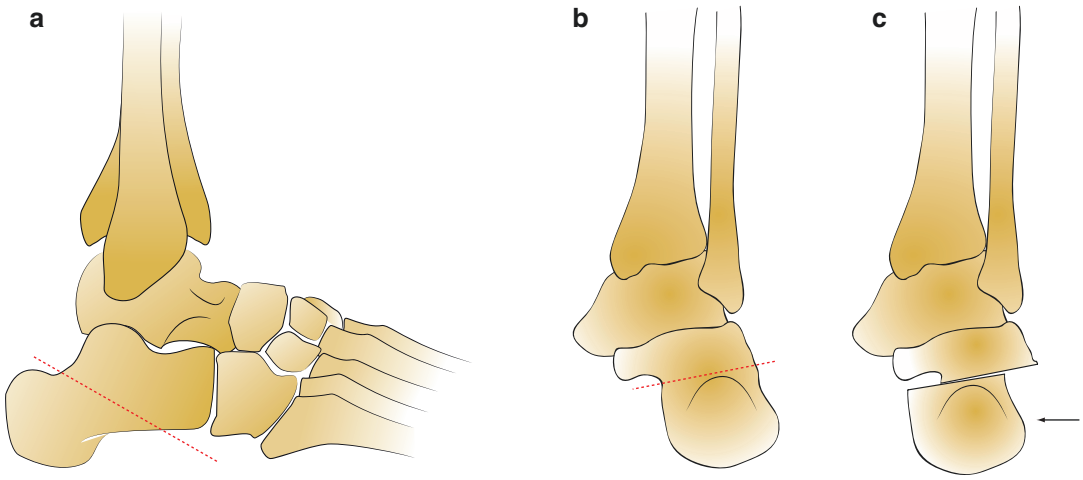



**Fig. 13.31** (**a**, **b**) Planned medial displacement calcaneal tubercle osteotomy (*dotted line*). (**c**) Medial displacement of the posterior part of the calcaneus

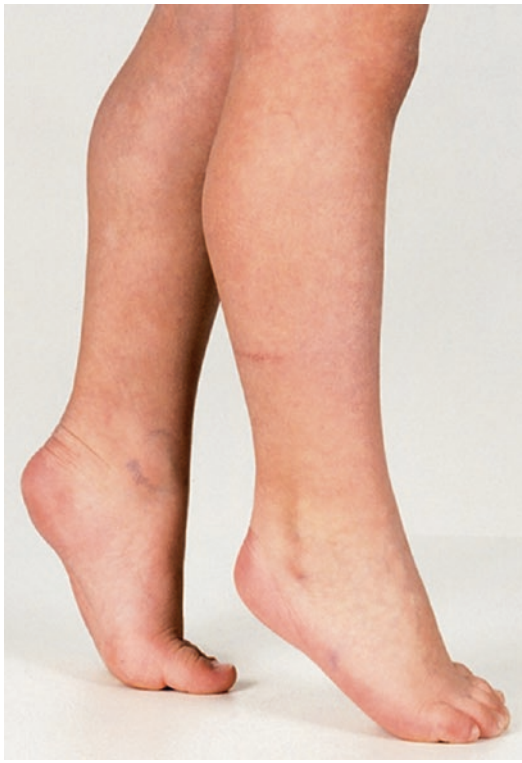


**Fig. 13.32** Toe walker

When to refer: if the child still walks on the toes at the age of 3.

Secondary care treatment: **habitual toe walker**. Treatment starts from the third year of life. There is a great chance of spontaneous correction prior to this age. If there is a neutral dorsiflexion/plantar flexion position or a slight dorsiflexion in the ankle joint on passive examination these feet can be treated with ankle-foot orthosis during the night with the foot in neutral position or slight dorsiflexion (Fig. 13.16). We normally sleep with the feet in plantar flexion. The calf muscles, will be stretched by keeping the feet in a neutral position with the ankle-foot orthosis at night. The ankle-foot orthosis treatment must be continued for 1–2 years. They will not toe walk anymore in 90 % of cases. However, after this treatment there may well be a flatfoot.

A structural equinus position in the foot will be treated with serial manual corrections and plaster cast immobilization until the foot can be brought into a neutral position. An achilles tendon lengthening may be considered in only exceptional cases in which a structural equinus is still present after serial manual correction and plaster cast immobilization. In order to prevent a recurrence it is advisable to carry out an achilles tendon lengthening in children older than 5 years of age followed by cast immobilisation for 6 weeks. After serial manual corrections with a short leg cast immobilisation or after an achilles

tendon lengthening further care is carried out with an ankle-foot orthosis during the night for 1 year.

## Flattened Longitudinal Medial Foot Arch

Complaint: the parents complain that their child's feet turn inwards.

Assessment: there is a flattening of the longitudinal medial foot arch and a slight heel valgus on examination when standing (Fig. 13.33).

Differential diagnosis:

**flat foot**
flexible pes planovalgus (flexible flatfoot)
masked equinus (hypermobile flatfoot and heel cord contracture)
tarsal coalition (rigid flatfoot, rigid pes planovalgus,
tarsal synostosis, peroneal spastic flatfoot)

Explanatory note: **flat foot**. There is a flattened longitudinal medial foot arch.

**Flexible pes planovalgus** A flexible flatfoot is generally caused by slack joint ligaments. A subluxation can occur in the talonavicular joint or the naviculocuneiform joint or both on weight bearing. The calcaneus pronates with respect to the talus and (or) the medial cuneiform subluxates in a plantar direction. This results in a heel valgus and the normal concavity of the foot arch flattens. When the child lies down or sits on the lap of the parent or carer whereby the feet hang down (in plantar flexion) the longitudinal medial foot arch becomes normal. The same situation occurs if the child lies with the feet in plantar flexion or if the child stands on the toes (Fig. 13.33). In some children the concavity of the medial foot arch will be seen in a weight bearing foot in which the big toe has been passively brought into extension, the so-called Hübscher test (Fig. 13.34). If there is a subluxation in the naviculocuneiform joint, then the Hübscher test will be positive. In many of these cases this will also be the case in the midfoot at the level of a subluxation of the naviculocuneiform as well as the talonavicular joint. The arch of the foot will be corrected by contracture of the flexor hallucis longus muscle. However, on passive extension of the big toe when the midfoot at the level of the talonavicular joint is not counterbalanced correction of the medial foot arch does not occur. The leverage of the flexor hallucis longus muscle is not sufficient to correct the subluxated talonavicular joint.

Children with flatfeet generally seldom have complaints.

The parents or carers sometimes think that something should be done to treat the deformity. One should avoid unnecessary treatment.

Most children up to the age of 2 years have flatfeet. A flexible flatfoot is present in 30–40 % of children between the ages of 2 and 5 years. Almost all flexible flatfeet will be spontaneously corrected before 7 years of age. Only 3–4 % of adults have a flexible flatfoot. A spontaneous correction occurs in more than 95 % of cases.

**Masked equinus** Some flatfeet are caused by a short achilles tendon in combination with hypermobility. A subluxation occurs at the level of the talonavicular and/or naviculocuneiform joint when the heel is grounded (Fig. 13.35). Apart from this subluxation the foot pronates which in time causes development of a hallux valgus. The foot must be brought into supination before being brought into dorsal extension in order to properly assess the degree of equinus.

**Tarsal coalition** In this type of flatfoot the longitudinal medial arch is also flattened and there is a heel valgus.

The longitudinal medial arch does not correct itself when the child is sitting, lying or standing on the toes in contrast to the flexible flatfoot. The longitudinal medial foot arch is not hollow and remains flat (Fig. 13.36). Inversion and eversion of the foot is not possible. The anomaly is caused by an abnormal complete or partial bony bridge connection between two or more bones. These are found usually between the calcaneus and navicular at the level of the tarsal sinus (50 %), on the medial side between the talus and calcaneus (40 %) and in the other 10 % of cases between

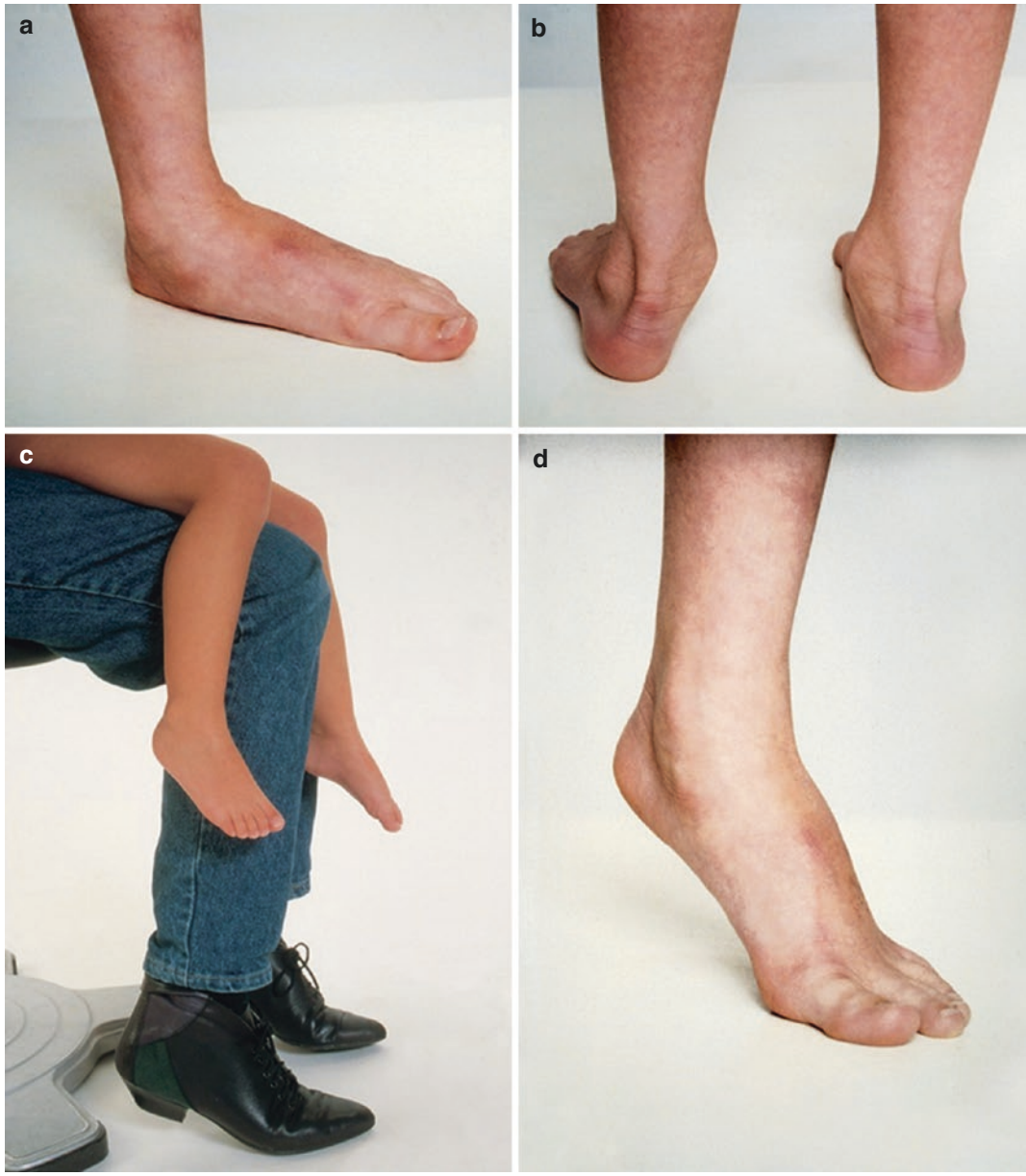



**Fig. 13.33** (**a**) Flatfoot. The normal longitudinal medial foot arch in the left foot is flattened. (**b**) There is a heel valgus. (**c**) The same foot as in Fig. 13.33a. A flexible flatfoot will be spontaneously corrected while sitting, in which case the foot is in plantar flexion at rest. The longitudinal medial foot arch becomes concave. (**d**) The same patient as in Fig. 13.33a. In a flexible flatfoot the concavity of the medial foot arch is restored when standing on the toes (plantar flexion)

the talus and navicular, talus and cuboid, navicular and cuneiform and the cuboid and navicular bones. The bony bridging is not necessarily complete. There may be fibrous or cartilaginous areas in the area where the bony bridging approaches two different pieces of bone. The deformity may give rise to pain at the level of the tarsal sinus between calcaneus and navicular. Pain occurs after ossification of the abnormal bony bridging. Bony bridging between the calcaneus and

navicular occurs between 8 and 12 years of age, between the talus and calcaneus between 12 and 16 years of age. The incidence of a tarsal coalition varies in various studies from 0.03 % to 2 %. The abnormality is seen in 39 % of first degree consan-

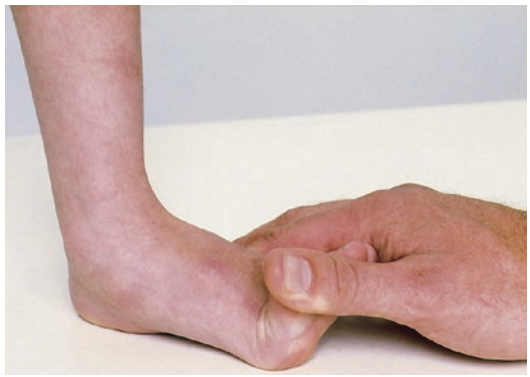


**Fig. 13.34** Hübscher test

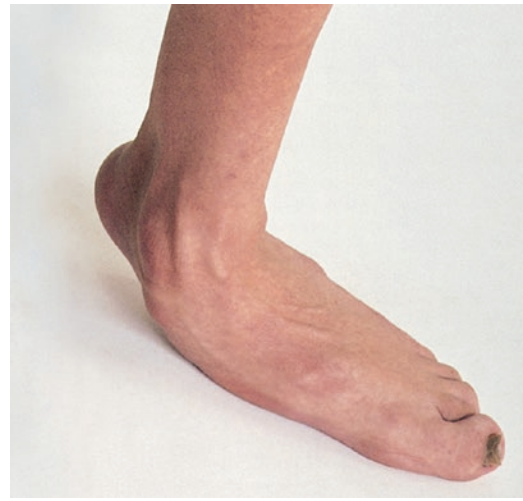


**Fig. 13.35** Masked equinus. In order to put the heel on the ground a subluxation occurs at the level of the talonavicular and/or the naviculocuneiform joint. There is foot pronation which causes a hallux valgus

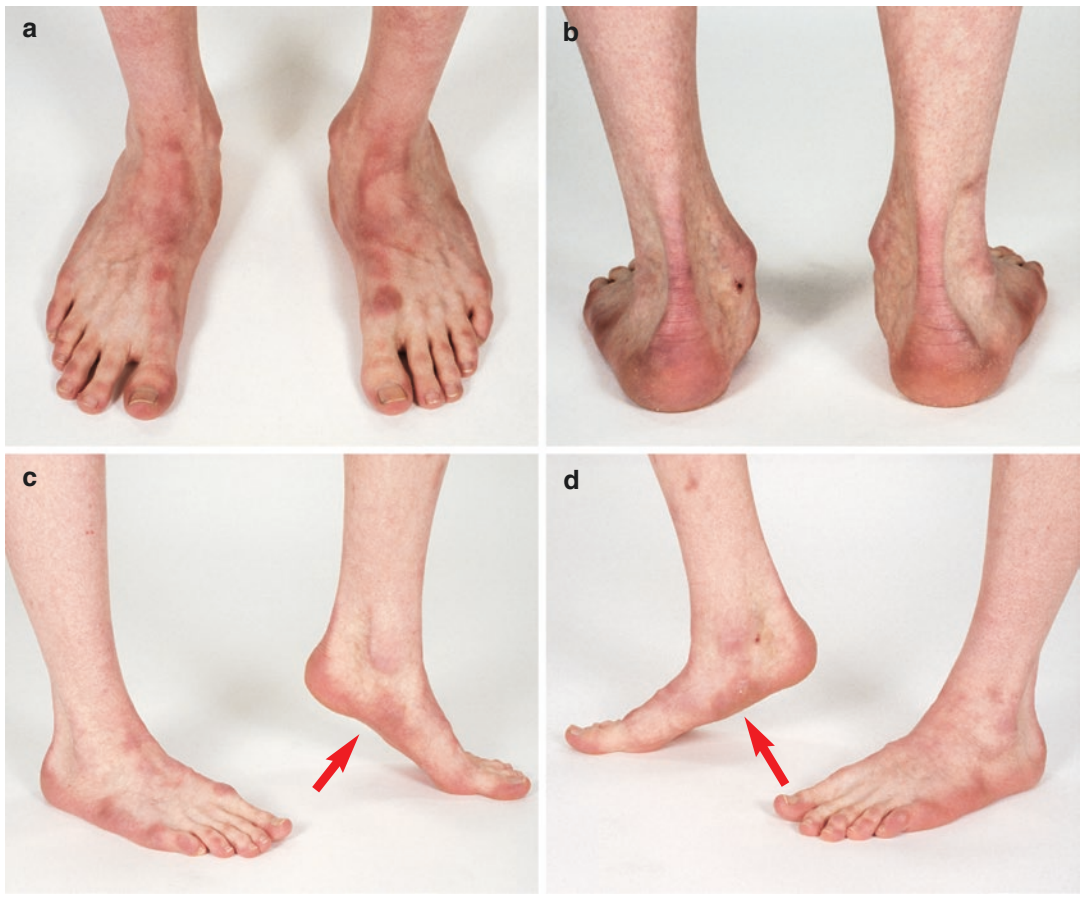



**Fig. 13.36** (**a**) Flattened foot arches. (**b**) Heel valgus. (**c**) The longitudinal medial foot arch of the left foot is corrected when standing on the toes: flexible flatfoot. (**d**) The longitudinal medial foot arch of the right foot remains flattened when standing on the toes: tarsal coalition

guinity. There is a bilateral deformity in 40–70 % of cases and in some cases there are more bony coalitions in the same foot. Sometimes there is an accompanying ball-and socket ankle (see p. 257).

Supplementary assessment: this is not necessary in a flexible flatfoot. Operative correction will only be considered in exceptional cases in which case the tarsometatarsal angle of Méary will be measured on a lateral standing X-ray of the foot (Fig. 13.37), this is normally 0°. In a flatfoot there is a plantar directed angle.

Apart from anteroposterior and lateral X-rays in case of a tarsal coalition an oblique (three quarters) X-ray must be taken. A possible abnormal bony bridge between the talus and navicular can be better observed on an oblique X-ray (Fig. 13.38). A CT-scan or MRI is necessary in order to see a bony bridge between the talus and calcaneus (Fig. 13.39). A technetium scan can be considered if there is any doubt. An enhanced activity at the level of the coalition shows that this is responsible for the complaints.

Primary care treatment: children with flexible flatfeet, with no complaints, do not need to be treated. The natural course of flexible flatfeet is not influenced by arch supports, shoe adjustment or exercises. Investigation of the muscles of the foot and lower leg have shown that the position of the foot is not influenced by these in a normal standing person. Above all in a flexible flatfoot the medial foot arch will be spontaneously normally established in plantar flexion during a large part of the day when lying down, sitting

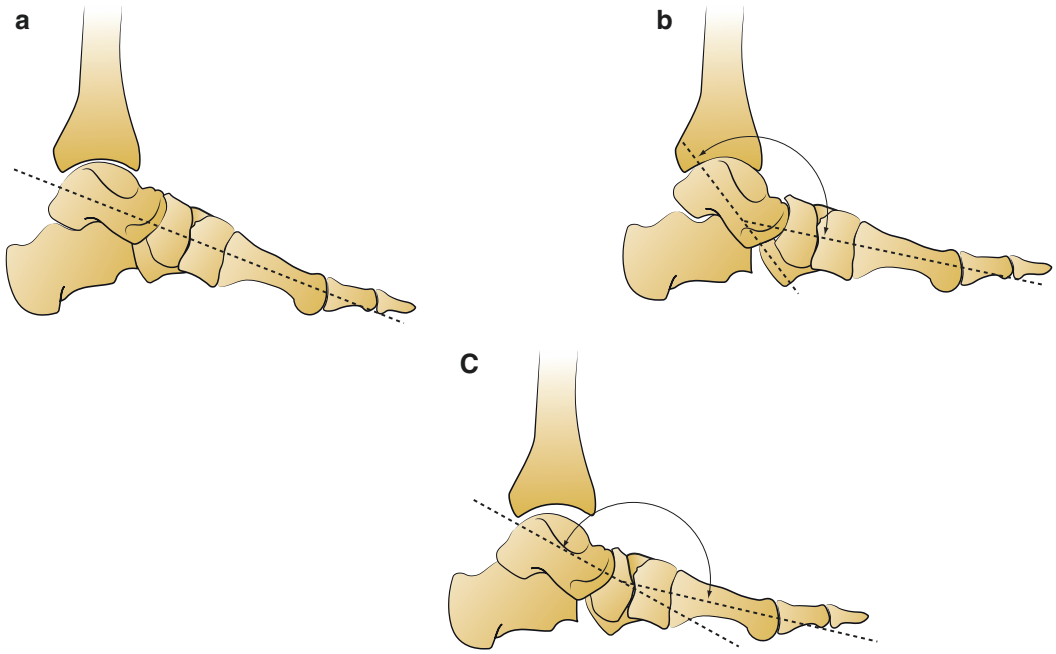



**Fig. 13.37** (**a**) The angle between the longitudinal axis of the talus and first metatarsal (Méary tarsometatarsal angle) must be normally 0°. Méary angle will show a plantar directed angle in a flatfoot. (**b**) Flatfoot with insufficiency at the level of the talonavicular joint. (**c**) Flatfoot with insufficiency at the level of the naviculocuneiform joint

with unburdened feet, standing on the toes and toe off during walking. It is often said, jokingly, that the child only has flatfeet when he or she has to stand neatly in front of the doctor. The parents may complain that the medial side of the shoes are worn out.

This is caused because the weight bearing area is on the medial side in a severe flatfoot. In these cases it is advisable to buy the child shoes with a solid strong heel (Fig. 13.40). As a result the heel valgus is counteracted and the weight bearing area is not on the medial side of the foot anymore. Occasionally a child will complain about flexible flatfeet. There is pain in the medial side of the foot, rapid fatigue and cramp in the lower legs. In these cases it is wise to consider correction of the flexible flatfeet with arch supports. Parents or carers must realize that the arch supports do not influence the final shape of the feet, but are only meant to reduce the complaints.

One in every four children with a tarsal coalition have no complaints. Treatment is not required if there are no complaints. Treatment of a symptomatic tarsal coalition is carried out in the first instance with the help of an arch support. One in four children are free of pain with this treatment.

» When to refer: in the case of a masked equinus and a tarsal coalition if the child still has complaints notwithstanding adequate arch supports.

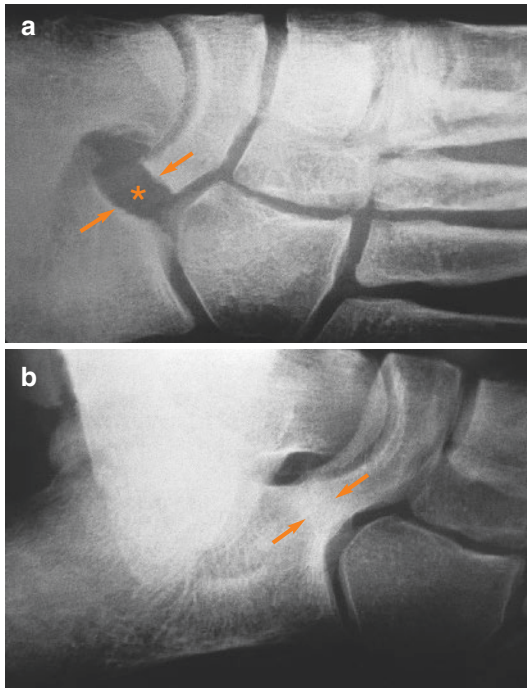



**Fig. 13.38** Oblique X-ray of the foot. (**a**) No connection between the calcaneus and navicular bones (*tarsal sinus). (**b**) Tarsal coalition (bony connection) between the calcaneus and navicular bones (between the *arrows*)

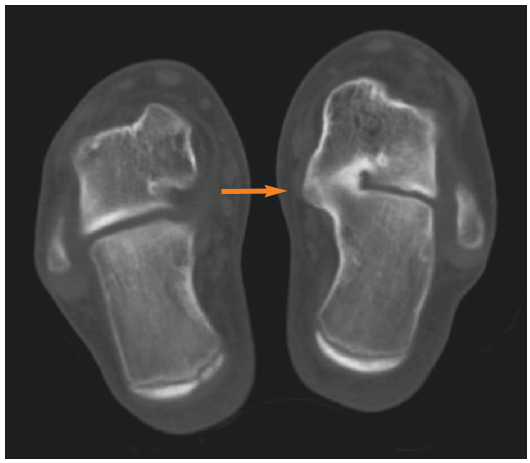


**Fig. 13.39** CT-scan foot: tarsal coalition on the medial side of the right foot between the calcaneus and talus (*arrow*)

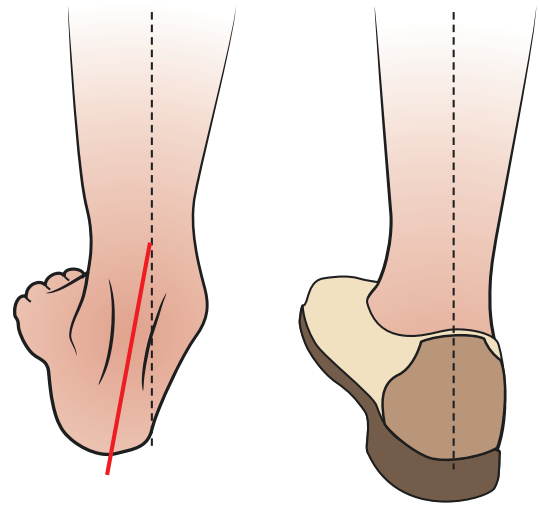


**Fig. 13.40** Heel valgus can be counteracted with a stiff strong heel shoe

Secondary care treatment: **flexible pes planovalgus**. Operative treatment is almost never necessary. In extreme cases an operative correction may be considered. Many operative procedures have been described, such as arthrodesis of the talonavicular joint or the joint between the navicular and cuneiform bones and also a subtalar extra articular arthrodesis. Nowadays the most techniques used are the lateral column lengthening of the calcaneal tubercle (Fig. 13.30) and an osteotomy to medialize the calcaneus (Fig. 13.31). After these procedures a short-leg cast is applied during 12 weeks and in the last 4 weeks a short-leg walking cast. An alternative is the insertion of an implant into the tarsal sinus (Kalix II). After care involves 4 weeks immobilization with a short-leg cast. This implant must be removed at the end of the growth period (Fig. 13.41). The long term results after this operation are not yet known.

**Masked equinus** Children with a masked equinus must be treated. Generally this can be done using serial manual corrections and cast immobilization followed by an ankle-foot orthosis at night for 1 year. In a few cases lengthening of the achilles tendon may be considered.

**Tarsal coalition** A short-leg plaster for 4 weeks can be prescribed in children who still have complaints after wearing arch supports. The complaints disappear in a few of these children. If the

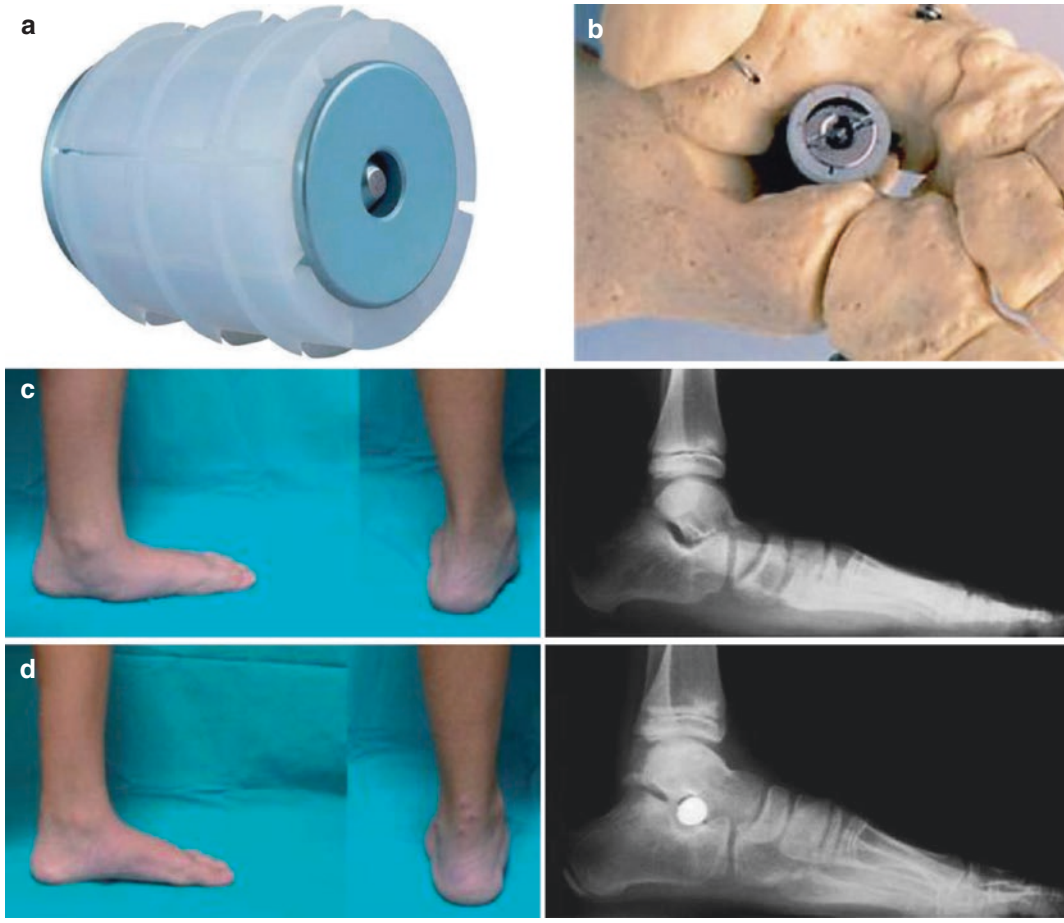



**Fig. 13.41** (**a**) Kalix II flatfoot implant. (**b**) Implant in the tarsal sinus. (**c**) Situation preoperatively. (**d**): situation postoperatively

complaints reoccur after short-leg plaster treatment an operation is necessary. The bony bridge is removed and in order to avoid a recurrence subcutaneous fat or, in the case of a coalition between the calcaneus and navicular bones, the muscle belly of the extensor hallucis brevis muscle can be interposed. Complaints remain in 10 % of cases after removal of the coalition. If secondary degenerative changes have already occurred, which is often the case in adults, a triple arthrodesis will be carried out when there is a coalition between the calcaneus and navicular bone and a subtalar arthrodesis will be chosen in the case of a coalition between the talus and calcaneus. There is no point in doing an excision if the bony bridge between the talus and calcaneus is more than 50 % of the total articular surface of the talocalcaneal joint. The complaints will remain. In that case there is an indication for a subtalar arthrodesis.

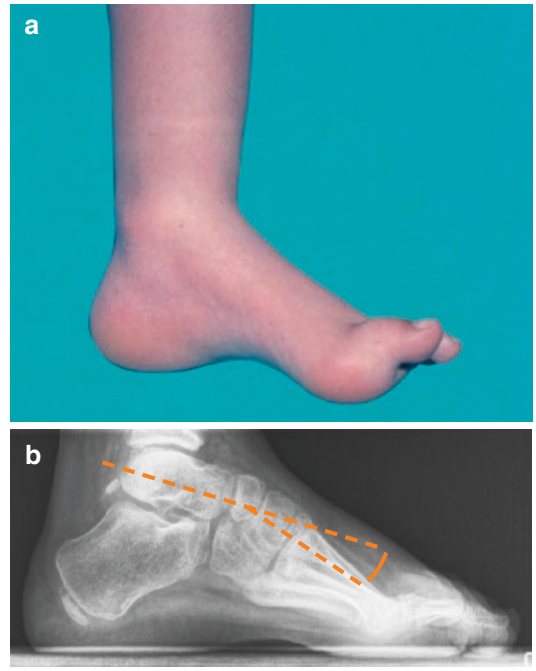



**Fig. 13.42** (**a**) Pes cavus. (**b**) Lateral X-ray of a pes cavus (see also Fig. 13.45)

## Abnormal High Longitudinal Medial Foot Arch

Complaint: the foot arch is too hollow.

Assessment: the longitudinal medial foot arch is higher than normal. There may be also be claw toes and a heel varus.

Differential diagnosis:

**hollow foot**
pes cavus (talipes cavus, high arch foot, cavoid foot)
pes calcaneocavus
pes cavovarus

Explanatory note: **hollow foot**. There is a abnormal high longitudinal medial foot arch.

Pes cavus. In this case the longitudinal medial foot arch is raised up. There is no varus deformity in the hindfoot and usually there are no claw toes. Children with a raised medial longitudinal foot arch usually go to the general practitioner with their parents at 6–7 years of age. In the first instance the abnormal shape of the foot was not conspicuous because the parents found that their child had a nice foot arch compared to other children, because young children usually have flatfeet. After complaints with extra callus formation on the ball of the foot they see that the arch of the foot is much higher than in children of the same age (Fig. 13.42). The deformity is usually bilateral. One of the parents often has hollow feet. Generally there are no neurological problems.

Pes calcaneocavus. The hindfoot is in dorsiflexion in the ankle joint which is compensated by plantar flexion, adduction and supination in the mid- and forefoot (Fig. 13.43). The deformity is caused by weakness of the calf muscles in combination with an overactive tibialis anterior muscle. It is often seen in poliomyelitis, spina bifida and after the achilles tendon has been lengthened too much.

Pes cavovarus. Apart from an increased longitudinal medial foot arch there is also a

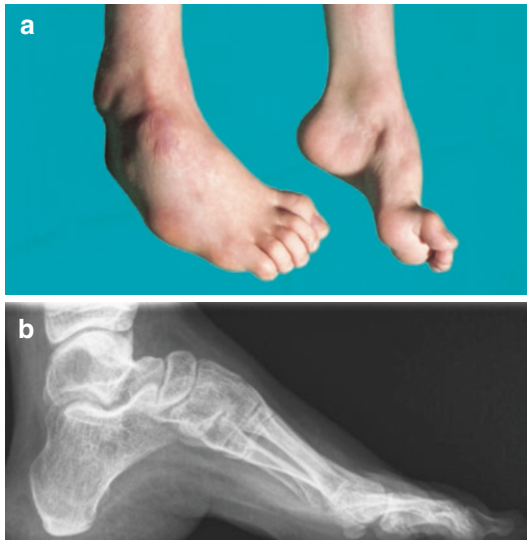



**Fig. 13.43** (**a**) Pes calcaneocavus. The hindfoot is in dorsiflexion which is compensated by plantar flexion in the midfoot. There is also an adduction and supination deformity. (**b**) Lateral X-ray of the left foot

heel varus, plantar flexion of the first metatarsal bone and a claw toe in all the toes (Fig. 13.44). Charcot-Marie-Tooth disease or hereditary motor and sensory neuropathy, shortened to HMSN, are the most common neurological causes of this deformity (Fig. 13.44). There is a weakness in the peroneus brevis muscle and tibialis anterior muscle in the Charcot-Marie-Tooth disease. The peroneus longus muscle keeps its normal strength in which case there is inversion[1] of the forefoot and a plantar flexion in the first ray. The strength of the tibialis posterior muscle remains normal whereby a varus deformity develops in the hindfoot. The extensors of the toes try to compensate the loss of function of the anterior tibial muscle. As a result there is hyperextension in the metatarsophalangeal joints and flexion in the proximal and distal interphalangeal joints due to passive tightening of the toe flexors.

A pes cavovarus also occurs in Friedreich ataxia and in an encephalopathy (spasticity).

[1]Inversion: hindfoot is in plantar flexion and adduction; the forefoot is in adduction and supination.

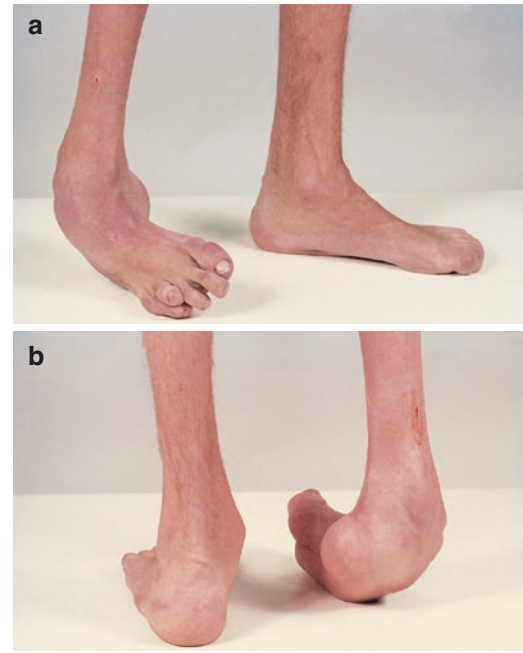



**Fig. 13.44** (**a**–**b**) Right pes cavovarus in Charcot-Marie-Tooth disease. Left side has already been corrected with a triple arthrodesis

In a spasticity case there is a dominating influence of the tibialis anterior muscle, the tibialis posterior muscle and the calf muscles.

Supplementary assessment: anteroposterior and lateral standing X-rays of both feet. The lateral X-ray is most important. On these X-rays the first metatarsal longitudinal axis must lie in continuity with the longitudinal axis of the talus.

In a hollow foot there is an angle between the two axes with the angle situated dorsally, the so-called Méary angle (Fig. 13.45). An alternative is to measure the Hibb angle. This is the angle between the longitudinal axis of the first metatarsal and the longitudinal axis of the calcaneus. This is more than 150° in a normal foot (Fig. 13.45).[1]

Primary care treatment: arch supports will be prescribed in order to redistribute pressure, reduce the pressure on the heads of the metatarsals and provide support for the medial foot arch in cases of mild hollow feet that are just not quite normal with complaints

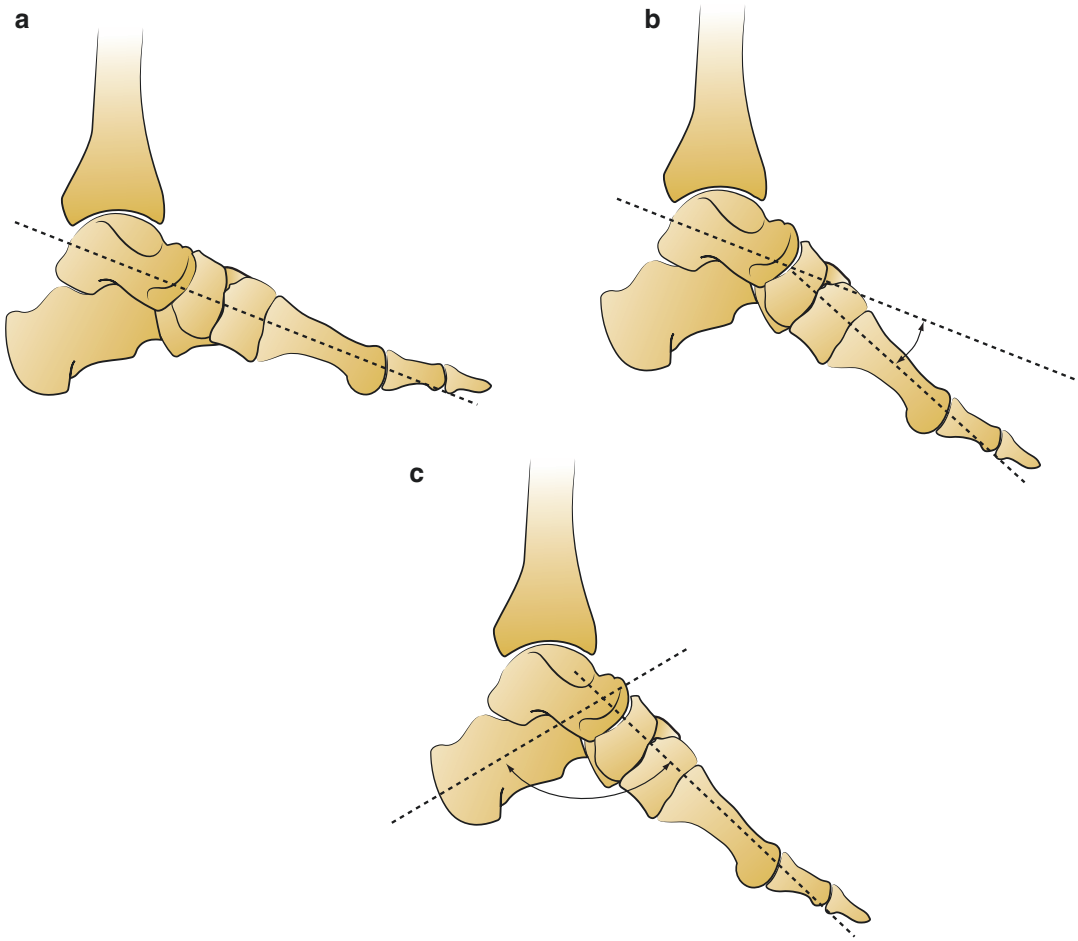



**Fig. 13.45** (**a**) Lateral view of the foot. The longitudinal axis of the first metatarsal should lie in line with the axis of the talus. (**b**) In a hollow foot the angle between these 2 length axes, Méary angle, will show a dorsally directed angle. (**c**) An alternative is to measure in the Hibb angle. This is the angle between the longitudinal axis of the first metatarsal and the longitudinal axis of the calcaneus. This angle is greater than 150° in a normal foot

at the level of the forefoot or the medial foot arch.

When to refer: if the arch supports do not help and if there is suspicion of a neurological problem. In this case referral to an orthopedic surgeon should be made and referral to a pediatric neurologist is necessary. About 2 of every 3 children with hollow feet have a neurological problem especially in the case of pes calcaneocavus and pes cavovarus.

Secondary care treatment: **pes cavus**. A soft tissue correction can be carried out in children younger than 8 years of age. A Steindler release is sufficient if there is only a hollow foot (Fig. 13.46). A medial incision is made just in front of the calcaneus and the plantar aponeurosis is divided transversely and the origins of the abductor hallucis brevis muscle, the flexor digitorum brevis muscle and abductor digiti quinti muscle are released. In the case of an extensive pes cavus a wedge osteotomy of the midfoot will be carried out in children older than 12 years of age (Fig. 13.47).

**Pes calcaneocavus** In a pes calcaneocavus an oblique osteotomy is carried out in the calcaneal tubercle after which the posterior fragment is cranially transposed (Fig. 13.48). A Steindler release is also necessary during this operation. An alternative is a triple arthrodesis, to correct the deformity (Fig. 13.49).

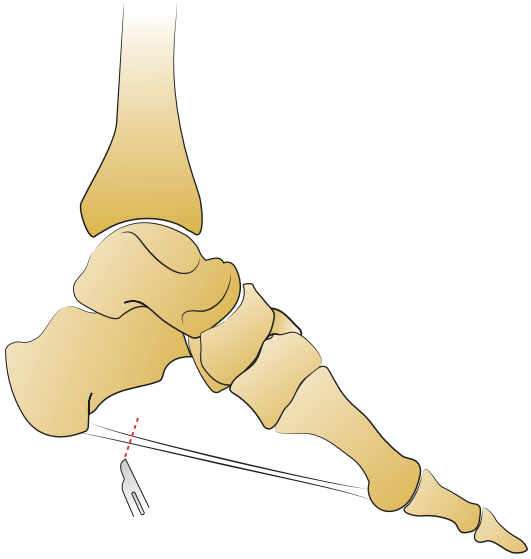


**Fig. 13.46** Steindler release. The plantar aponeurosis is divided and the origins of the abductor hallucis, flexor digitorum brevis and abductor digiti minimi muscles are released

**Pes cavovarus** In the case of a deeply plantar flexed first metatarsal bone and a varus deformity in the hindfoot one should first determine if the positional deformities are flexible or rigid. The heel varus can be best tested by placing the lateral side of the child's foot onto a plank (Fig. 13.50). If correction occurs then this is a flexible deformity. In Charcot-Marie-Tooth disease a flexible deformity can be treated with a Steindler release and the function of the peroneus brevis muscle can be restored by transposing the peroneus longus tendon to that of the

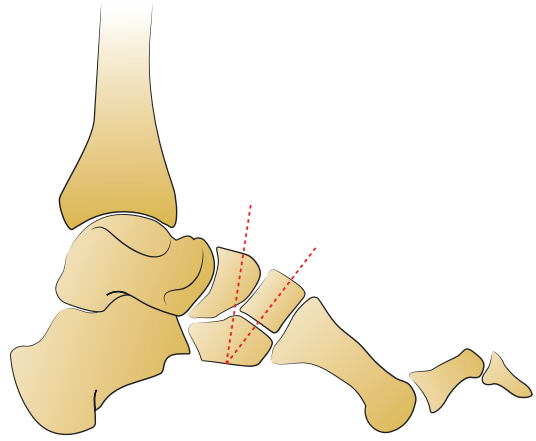


**Fig. 13.47** Closing wedge osteotomy of the midfoot

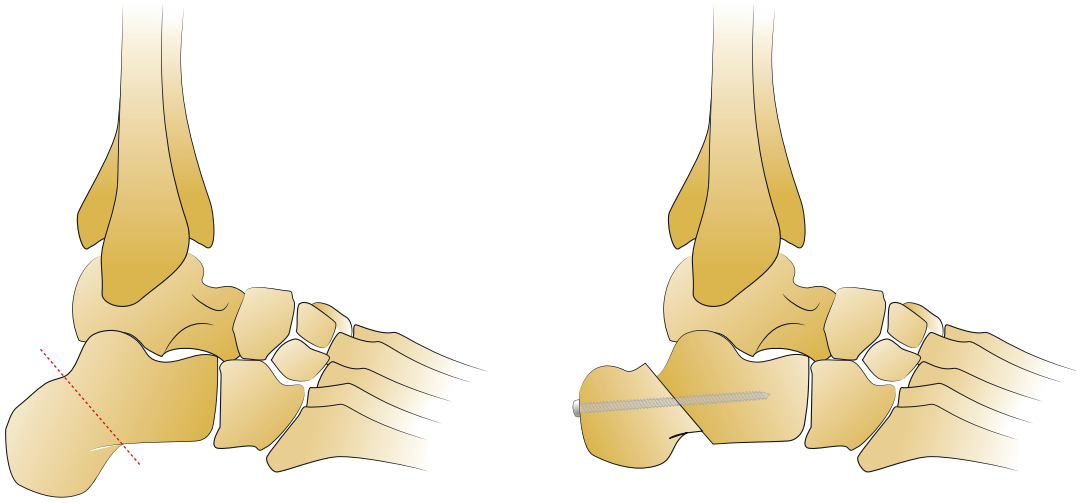


**Fig. 13.48** Osteotomy of the calcaneal tubercle in which the posterior part of the calcaneus is transposed dorsally. A Steindler release is also necessary when carrying out this operation

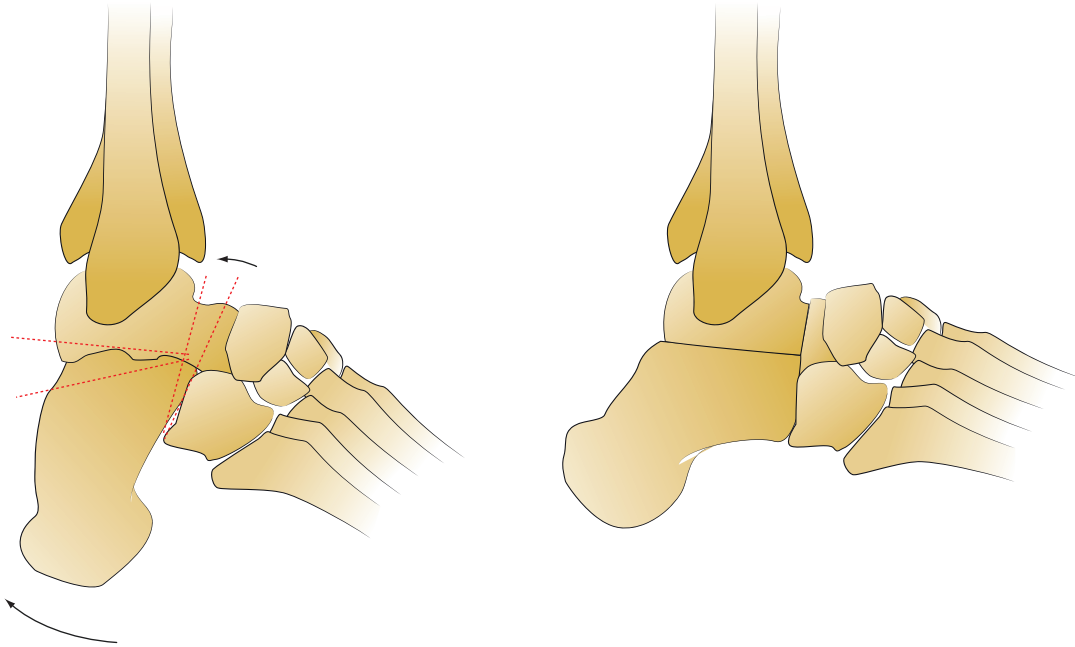


**Fig. 13.49** Pes calcaneocavus correction with a triple arthrodesis

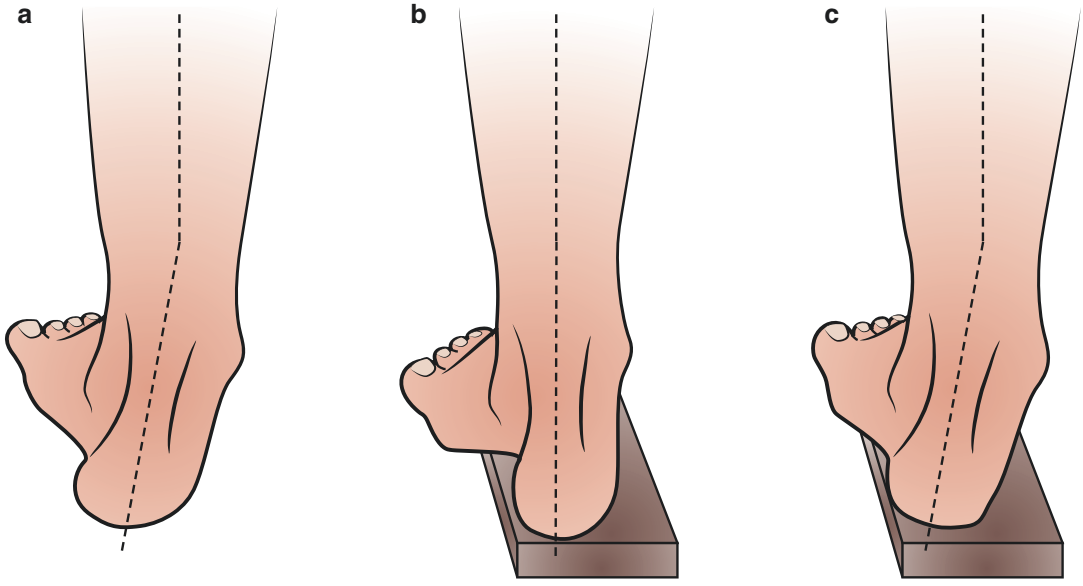



**Fig. 13.50** (**a**) A varus deformity of the hindfoot can best be tested by placing the child's lateral foot edge onto a plank whereby the head of the first metatarsal lies medial to it. (**b**) If correction occurs then we are dealing with a flexible heel varus. (**c**) In a rigid heel varus correction will not occur ("Coleman block test")

peroneus brevis. The posterior tibial tendon can be transposed to the dorsum of the foot in order to lessen the heel varus and to improve the strength of dorsiflexion in the ankle. A deeply plantar flexed first metatarsal bone can be corrected using the Jones procedure. The extensor hallucis longus tendon will be divided in the middle of the proximal phalanx and this will be tunneled through a borehole in the head of the first metatarsal bone and sutured to itself. By suturing the distal part of the tendon onto the proximal phalanx a tenodesis is achieved in the interphalangeal joint (Fig. 13.51). The forced abnormal position of the other toes can be solved by dividing the insertion of the extensor tendons and suturing these onto the heads of the metatarsals. These tendon insertions can also be fixed at the level of the cuneiform bones.

A Steindler release and tendon transpositions are basically insufficient in children between the ages of 8 and 12.

In the case of a rigid (structural) deeply flexed first metatarsal bone a dorsiflexion osteotomy of the first ray can be carried out apart from the Jones procedure (Fig. 13.52). A structural heel varus will be corrected with a so-called Dwyer osteotomy (Fig. 13.28), in which the varus deformity is corrected by removing a wedge with the base on the lateral side of the calcaneus. In the case of a serious cavovarus in children older than 12 years of age, apart from a correction of the first metatarsal bone, a triple arthrodesis will be carried out (Fig. 13.53). In this case wedge shaped osteotomies at the level of the subtalar joint and the Chopart joint are carried out with the base of the wedge on the lateral side.

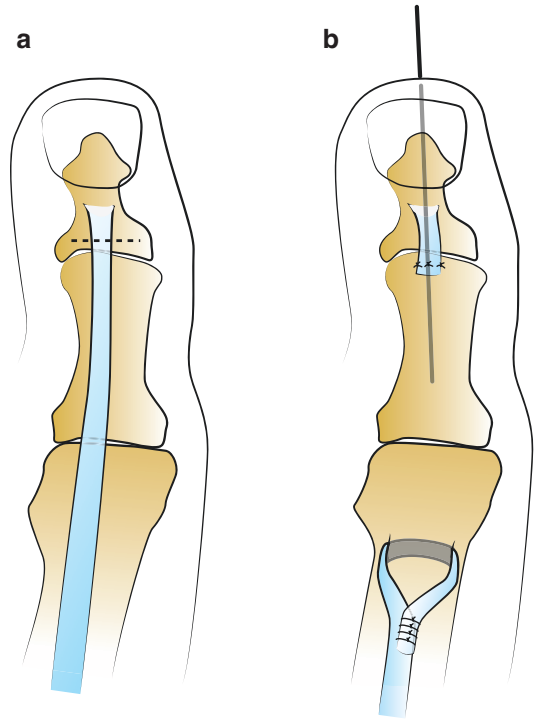



**Fig. 13.51** Jones procedure. (**a**) The tendon of the extensor hallucis longus muscle will be divided at the level of the middle of the proximal phalanx and (**b**) will be tunneled through a bore hole in the head of the first metatarsal bone after which it will be sutured to itself. A tenodesis of the interphalangeal joint of the big toe is achieved by suturing the distal part of the tendon to the proximal phalanx

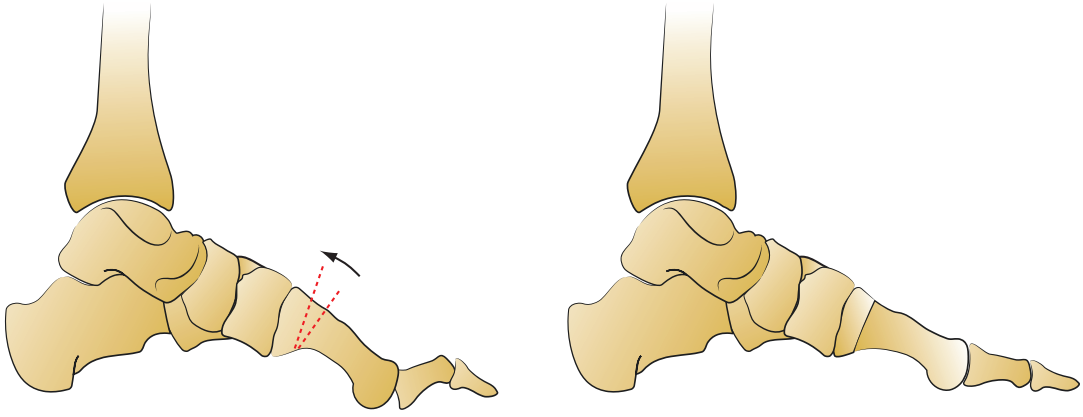


**Fig. 13.52** Dorsiflexion osteotomy first metatarsal

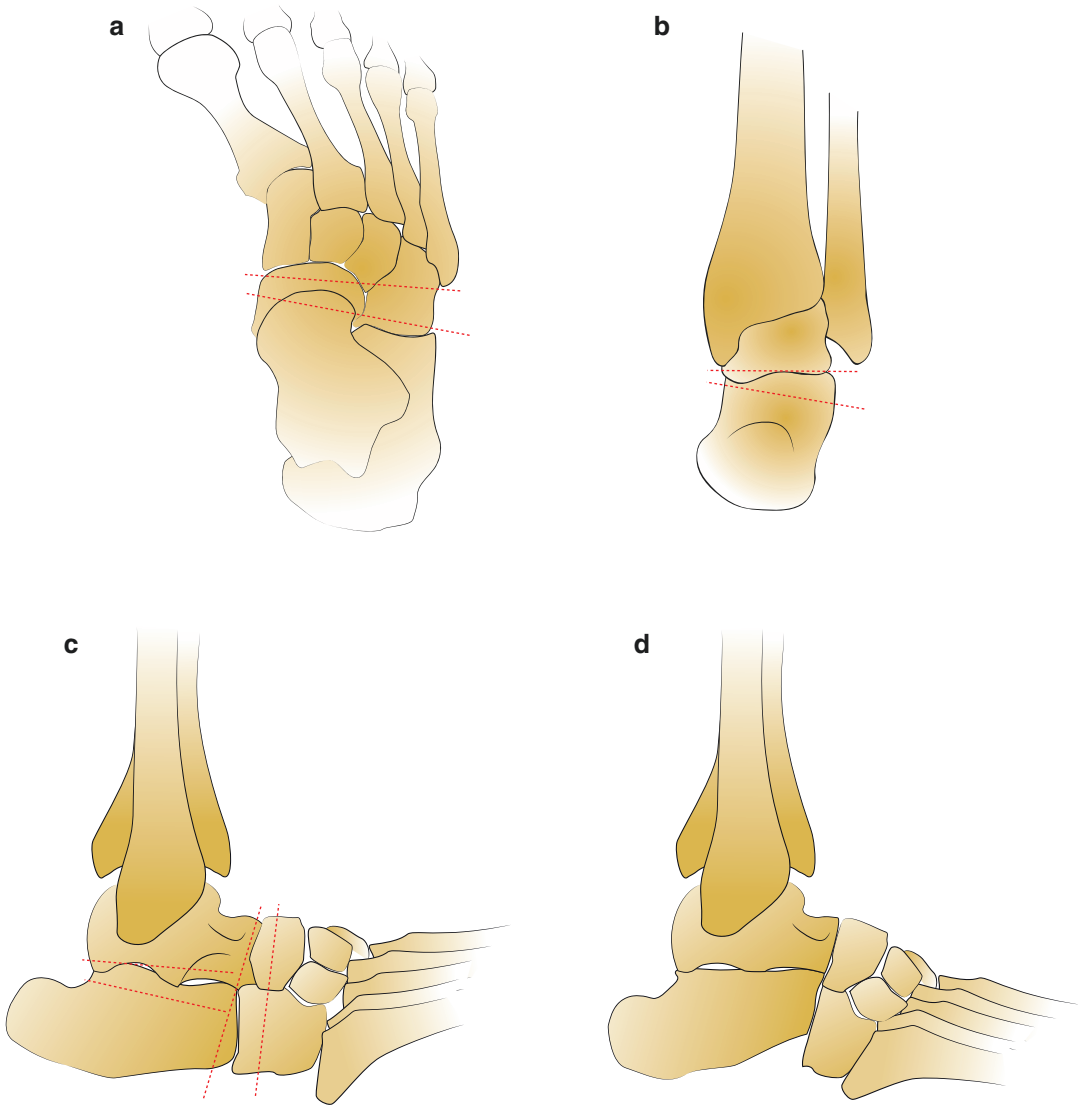



**Fig. 13.53** Triple arthrodesis in a pes cavovarus. (**a**) A closing wedge osteotomy of the Chopart joint with the base of the wedge on the lateral side. (**b**) Closed wedge osteotomy of the subtalar joint with the base of the wedge on the lateral side. (**c**) A possible equinus stand in the hindfoot will be corrected with a closed wedge osteotomy at the level of the subtalar joint in which the wedge on the anterior side is made broader than on the posterior side. (**d**) Corrected position

## Foot Pain

If a child complains about a painful foot, it is usually older than 5 years of age. The child can usually quite accurately point to the localization.

### Pain in the Outer Side of the Hindfoot

Complaint: the child complains of pain at the level of the tarsal sinus usually after 8 years of age.

Assessment: there is a flattened longitudinal medial foot arch and generally a slight heel valgus. The arch is not hollow, but remains flattened when the children stand on their toes. Inversion and eversion are impossible. Forced inversion and eversion is painful.

Diagnosis: **tarsal coalition** (**rigid flatfoot**, **rigid pes planovalgus**, **tarsal synostosis**, **peroneal spastic flatfoot**)

**Differential Diagnosis: Foot Deformities**

| | |
|---|---|
| **Foot deformities present at birth** | |
| First metatarsal bone and big toe in adduction | **Metatarsus primus varus** |
| Mid- and forefoot in adduction | **Metatarsus adductus** |
| Mid- and forefoot in adduction and supination | **Metatarsus varus** (**one third clubfoot**) |
| Mid- and forefoot in adduction and supination, heel varus | **Pes varus** (**two thirds clubfoot**) |
| Mid- and forefoot in adduction and supination, heel in equinus and varus | **Talipes equinovarus** (**clubfoot**) |
| Flexible clubfoot | Postural talipes equinovarus |
| Clubfoot with an unknown cause | Idiopathic talipes equinovarus |
| Clubfoot associated with a syndrome | Teratogenic talipes equinovarus |
| Clubfoot associated with spinal cannel abnormalities, such as spina bifida sacral agenesis | Neurological talipes equinovarus |
| Extreme heel dorsiflexion and slight heel valgus | **Calcaneovalgus deformity** |
| Mid- and forefoot in abduction and dorsiflexion. Heel equinus and valgus | **Convex pes valgus** (**congenital talus verticalis**) |
| Unilateral | Isolated convex pes valgus |
| Associated with a symdrome | Teratogenic convex pes valgus |
| Associated with spinal cannel abnormalities, such as spina bifida sacral agenesis | Neurological convex pes valgus |
| Forefoot in adduction, midfoot in abduction, heel equinus and valgus | **Skewfoot** (**Z-foot**, **serpentine foot**) |
| **Toewalking** | |
| The child walks on the toes, there is limited dorsiflexion in the ankle | **Habitual toe walker** (**idiopathic toe walking**, **equinus gait**, **congenital short achilles tendon**) |
| **Flattened longitudinal medial foot arch** | **Flatfoot** |
| The longitudinal medial foot arch is flattened when standing | |
| The longitudinal medial foot arch becomes normal with the ankle in plantar flexion at rest | |
| | Flexible pes planovalgus (flexible flatfoot) |
| Flattened longitudinal medial foot arch with heel equinus | Masked equinus (hyper mobile flatfoot and heel-cord contracture) |
| Flattened longitudinal medial foot arch which is not corrected with the ankle in plantar flexion | Tarsal coalition (rigid flatfoot, rigid pes planovalgus, tarsal synostosis, peroneal spastic flatfoot) |
| **Abnormal high longitudinal medial foot arch** | **Hollow foot** |
| High longitudinal medial foot arch | Pes cavus (talipes cavus, high arch foot, cavoid foot) |
| The hindfoot is in dorsiflexion, the mid- and forefoot in plantar flexion, adduction and supination | Pes calcaneocavus |
| Heel varus, and extensive plantar flexion of the first metatarsal bone, claw toes | Pes cavovarus |

Explanatory note: **tarsal coalition** (for explanatory note, supplementary assessment, primary care treatment, when to refer and secondary care treatment, see pp. 279–285).

## Heel Pain

Complaint: this usually involves boys between 8 and 13 years of age who complain of heel pain.

Assessment: there is pressure pain at the level of the insertion of the achilles tendon.

Differential diagnosis:
**Sever-Schintz disease** (**apophyseal osteochondrosis**, **calcaneal apophysitis**)
**unicameral bone cyst** (**juvenile**-, **solitary**-, **simple bone cyst**)
**calcaneal stress fracture**
**plantar fasciitis**

Explanatory note: **Sever-Schintz disease**. This involves an overloading at the level of the insertion of the achilles tendon on the calcaneal apophysis. The incidence is about 6 %. The complaints are bilateral in 50 % of cases. It particularly involves boys between 8 and 13 year of age who are quite active. On examination squeezing on both sides of the heel causes pain (squeeze test). The complaints usually disappear spontaneously after some years.

**Unicameral bone cyst** A solitary bone cyst is a cavity which is filled with yellow fluid and in which the wall is covered with a thin fibrous membrane. The condition is found in 90 % of cases in patients younger than 20 years of age. A spontaneous fracture occurs in more than 80 % of cases. Previously there were no complaints and this often is the first symptom of the anomaly. 70 % of cases are males. The cyst is localized in the proximal part of the humerus and the proximal part of the femur in 90 % of cases. The third most common localization is in the calcaneus.

**Calcaneal stress fracture** Most stress fractures occur in the proximal part of the tibia and the distal part of the fibula. The third most common occurrence is in one of the metatarsals. The calcaneus, navicular and cuboid bone are the rarest locations.

**Plantar fasciitis** Pain at the level of the attachment of the plantar fascia on the calcaneus. The cause is unknown. There is pressure pain at the front of the calcaneus on the plantar side.

Supplementary assessment: an X-ray of the calcaneus should only be considered in the case of persistent complaints to exclude a calcaneal bone cyst or a stress fracture (Fig. 13.54). Sever-Schintz disease cannot be diagnosed on X-ray. There is a big radiological variation in the structure of the calcaneal apophysis (Fig. 13.55). An X-ray in plantar fasciitis will not show any anomaly.

Primary care treatment: the complaints caused by Sever-Schintz disease usually disappear after some years, particularly if the child limits his or her activities. A visco elastic heel pad may be prescribed. This heel pad is shock absorbent and at the same time the foot is fixed in a slight equinus so that the

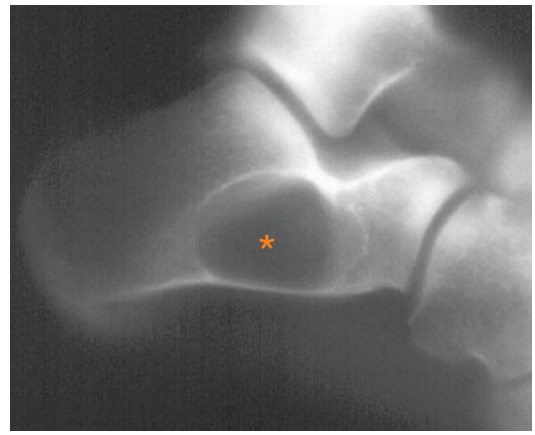



**Fig. 13.54** A unicameral bone cyst in the calcaneus (*)

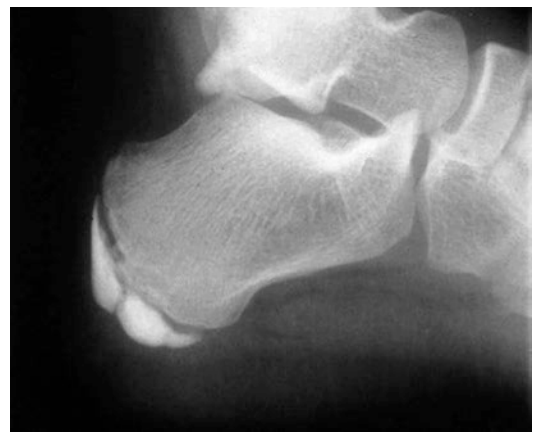


**Fig. 13.55** Lateral X-ray of the heel. There is great radiological variation in the structure of the calcaneal tuberosity. Sever-Schintz disease cannot be diagnosed on X-ray

**Fig. 13.56** Visco-elastic-heel pads

**Fig. 13.57** Strassburg sock

insertion of the achilles tendon will be less burdened (Fig. 13.56). A plantar fasciitis can be treated during the day with a visco elastic heel pad and/or a Strassburg sock at night (Fig. 13.57).

When to refer: if there is a solitary bone cyst or a stress fracture and children with Sever-Schintz disease who have no pain reduction after wearing visco elastic heel pads.

Secondary care treatment: **Sever-Schintz disease**. Secondary care treatment cannot achieve much more than primary care. A short-leg cast may be prescribed for a period of 3–6 weeks in the case of severe pain.

**Unicameral bone cyst** A possible fracture caused by a solitary bone cyst at the level of the calcaneus can be treated with a lower leg cast for a period of 6 weeks. After the fracture the bone cyst heals spontaneously in 25 % of the cases. If the bone cyst does not disappear then corticosteroids can be injected after fracture consolidation. In the case of a bone cyst localized in the proximal part of the humerus and the proximal part of the femur this treatment is more effective than in bone cysts in the calcaneus. A bone cyst at the level of the calcaneus will often be curetted and the cyst is filled up with an autologous or donor bone graft or with calcium phosphate chips.

**Calcaneal stress fracture** Six weeks cast immobilisation.

## Pain in the Inner Side of the Midfoot

Complaint: this involves children usually between 2 and 7 years of age that have pain in the medial foot arch.

Assessment: pressure pain at the level of navicular bone.

Diagnosis: **Köhler disease**

Explanatory note: **Köhler disease**. This anomaly occurs in 75 % of cases in boys between 2 and 7 year of age. It involves an ischemic necrosis of the navicular bone that spontaneously recovers after 1.5–3 years without remaining symptoms.

Supplementary assessment: anteroposterior and lateral X-rays of the foot reveal a sclerotic navicular bone (Fig. 13.58).

Primary care treatment: supportive insoles may be prescribed to diminish pain.

When to refer: if the children still have a lot of pain notwithstanding the supportive insoles.

Secondary care treatment: **Köhler disease**. A plaster of Paris lower leg splint during 8 weeks may be considered if there is still a lot of pain notwithstanding supportive insole treatment. The chance of recurrence is greater if the shortleg cast is applied for a shorter period.

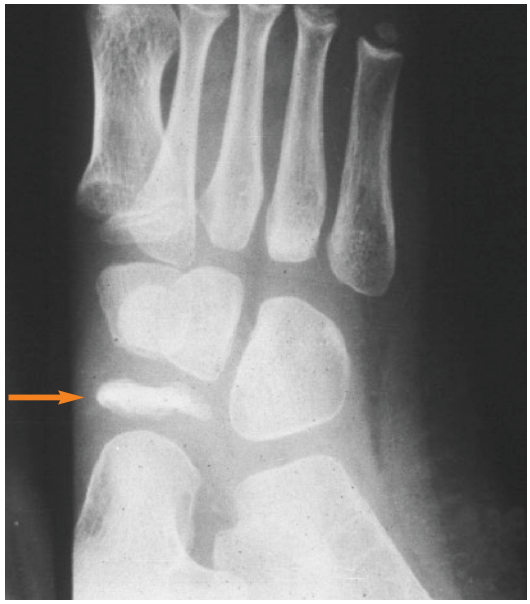


**Fig. 13.58** Köhler disease (sclerosis of the navicular bone)

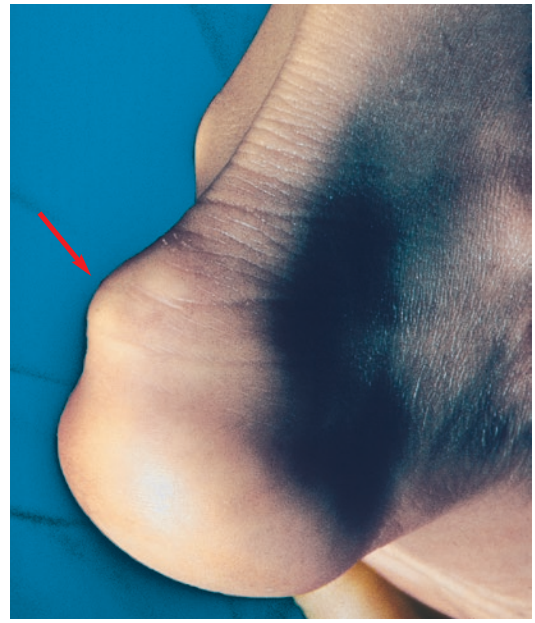


**Fig. 13.59** Haglund deformity. The swelling is caused by a bursitis of the subcutaneous calcaneal bursa

## Painful Swelling on the Upper Outer Side of the Heel

Complaint: there is a painful swelling on pressure on the upper outer side of the calcaneus usually involving girls between 10 and 15 years of age. The anomaly is generally bilateral.

Assessment: there is a painful swelling on pressure on the upper outer side of the ankle.

Diagnosis: **Haglund deformity** (**calcaneal prominence**)

Explanatory note: **Haglund deformity**. A Haglund deformity is not a specific deformity but a bony prominence of the posterior upper and outer part of the calcaneal tubercle. An irritation between this bony prominence and the achilles tendon occurs as a result of pressure from the shoes particularly shoes with a low heel support. In 50% of cases there is a bursitis in the bursa between the bony prominence and the achilles tendon, the retrocalcaneal bursa and/or the achilles tendon and the skin, the subcutaneous calcaneal bursa (Fig. 13.59).

Supplementary assessment: unnecessary.

Primary care treatment: the patient is advised to wear shoes with a high supportive heel, provided with a piece of felt on the inside of the heel of the shoe.

When to refer: with persistent complaints notwithstanding adequate footwear which have been fitted with a felt pad on the inside.

Secondary care treatment: **Haglund deformity**. Removal of the irritated bursa between the achilles tendon and the skin and/or the irritated bursa between the calcaneal tubercle and the achilles tendon. The bony prominent part of the posterior upper part of the calcaneal tubercle can be removed to prevent a recurrence of complaints in a child who has finished growing.

## Painful Swelling on the Inner Side of the Midfoot

Complaint: this involves children at puberty who have a painful swelling on the medial side of the midfoot.

Assessment: the anomaly looks like an abnormal bony protuberance just in front and under the medial malleolus.

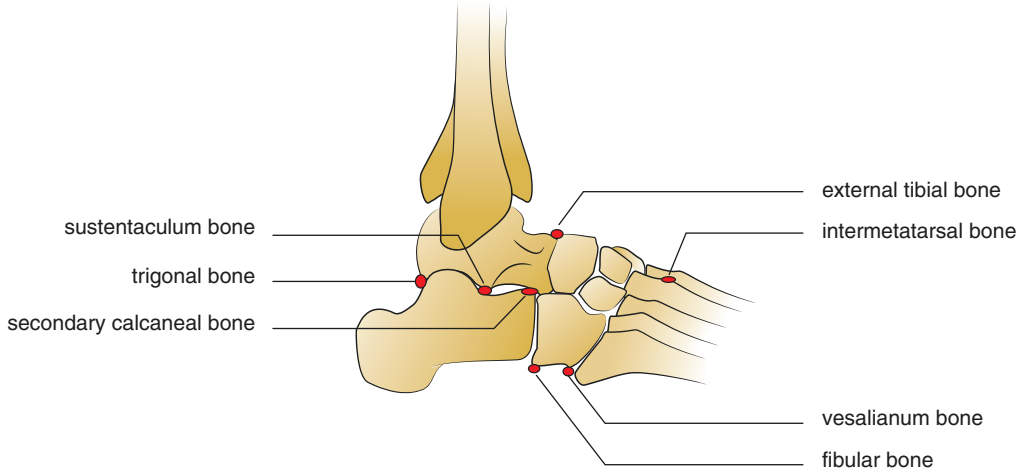



**Fig. 13.60** The most frequent extra bones. These are called sesamoid bones

Differential diagnosis:
**symptomatic external tibial bone** (**accessory navicular bone**, **secondary navicular bone**)
type I
type II
type III

Explanatory note: **symptomatic external tibial bone**. An external tibial bone is an accessory bone (sesamoid bone) that occurs in 10 % of all individuals. A sesamoid bone also can appear in other localizations in the foot (Fig. 13.60 for the most frequent localizations).

The navicular bone is the last bone to ossify. Ossification occurs in girls between 1 and 3 years of age and in boys between 3 and 5 years of age. Ossification of the accessory navicular bone occurs later. Usually there are no complaints.

The external tibial bone can be divided into 3 types.

Type I. Type I is a small accessory bone in the posterior tibial tendon.

Type II. Type II is an 8–12 mm large bone medial and plantar to the navicular bone.

Type III. This is a bony bridge between the external tibial bone and the navicular bone which appears as a bony projection on the medial and plantar side of the navicular bone. This can cause complaints at puberty when the body weight quickly increases and this is particularly the case in children who are active in sports. Tight fitting shoes can also have influence.

Supplementary assessment: the diagnosis can be determined on an anteroposterior X-ray of the foot (Fig. 13.61).

Primary care treatment: supportive insoles give relief in a large number of cases.

When to refer: if supportive insoles give no relief.

Secondary care treatment: **symptomatic external tibial bone**. The external tibial bone can be operatively removed in cases with persistent complaints notwithstanding wearing supportive insoles. If there is a prominent medial part of the navicular bone this can be operatively flattened off. In that case the posterior tibial tendon has to be reinserted. Aftercare with a short-leg cast for 6 weeks.

## Painless Bilateral Swelling on the Medial Side of the Footsole

Complaint: painless swelling on the footsole.

Assessment: nodule on the medial side of the footsole.

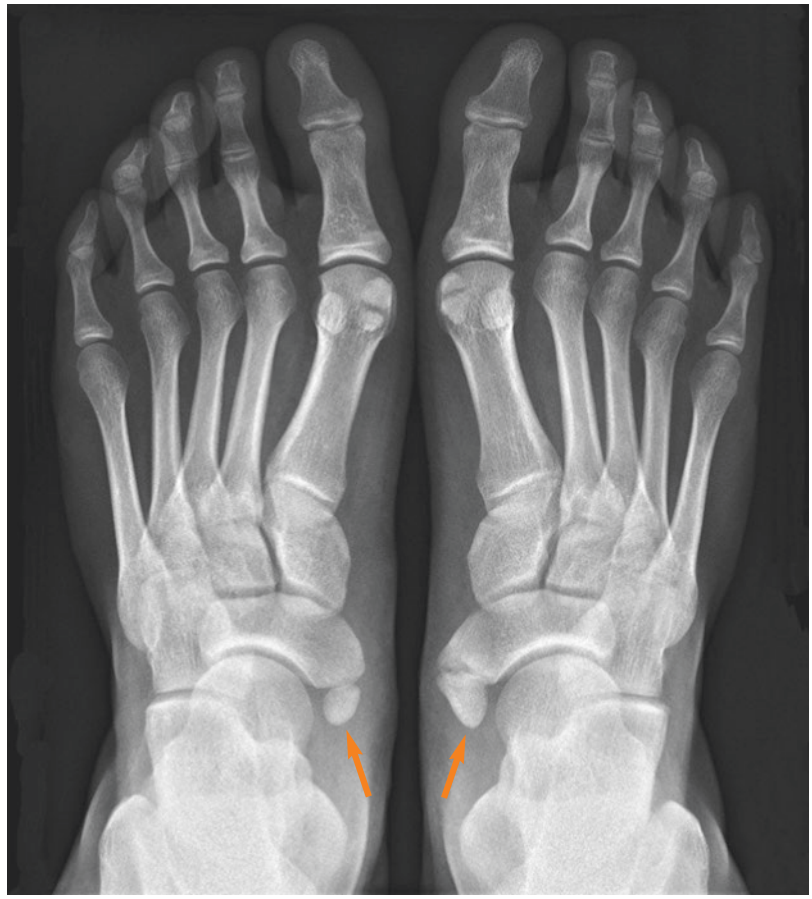


**Fig. 13.61** Bilateral external tibial bone (*arrows*)

Differential diagnosis:
**precalcaneal congenital fibrolipomatous hamartoma**
**plantar fibromatosis** (**Ledderhose disease**).

Explanatory note: **precalcaneal congenital fibrolipomatous hamartoma**. Bilateral painless fibrolipomatous swelling on the anteromedial side of the heel, 1.5 cm in diameter, present from birth. Sometimes this swelling disappears spontaneously (Fig. 13.62).

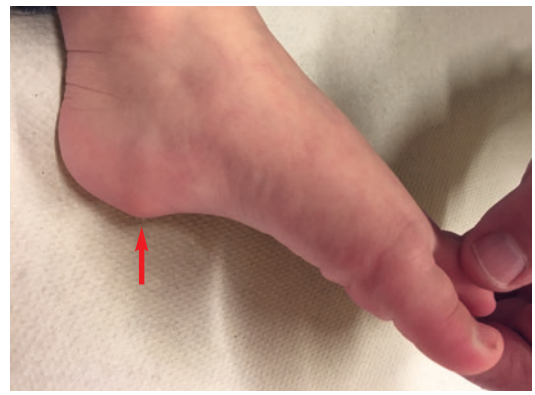


**Fig. 13.62** Precalcaneal congenital fibrolipomatous hamartoma (*arrow*)

**Plantar fibromatosis** This rare swelling is mostly present on the medial border of the foot near the highest point of the medial longitudinal arch. The lump is a nodular thickening of the plantar fascia, in 25 % bilateral and usually painless. This swelling may occur at any age. The only pain experienced is when the nodule rubs on the shoe or floor. The overlying skin is freely mobile and contracture of the toes does not occur in the initial stages.

Supplementary assessment: ultrasound and optionally an MRI.

Primary care treatment: anteromedial plantar nodules of the heel in childhood need no treatment because plantar fibromatosis mostly resolves spontaneously and recurrence following resection is common. A wait-and-see policy is indicated. Soft inner soles may be helpful in the case of pain when walking.

When to refer: only if discomfort hinders walking or there is the possibility of a fibrosarcoma.

Secondary care treatment: refer to a specialized medical centre if there is the possibility of a fibrosarcoma.

**Differential Diagnosis: Foot Pain**

| | |
|---|---|
| **Pain in the outer side of the hindfoot** | |
| Flattened longitudinal medial foot arch which does not recover with plantar flexion in the ankle | **Tarsal coalition** (**rigid flatfoot**, **rigid pes planovalgus**, **peroneal spastic flatfoot**) |
| **Heelpain** | |
| Positive squeeze test | **Sever-Schintz disease** (**apophyseal osteochondrosis**, **calcaneal apophysitis**) |
| Pressure pain at the level of the calcaneus | **Unicameral bone cyst** (**juvenile-**, **solitary-**, **simple bone cyst**) **Calcaneal stress fracture** |
| Pressure pain at the front of the calcaneus on the plantar medial side | **Plantar fasciïtis** |
| **Pain in the inner side of the midfoot** | |
| Pressure pain at the level of the navicular bone | **Köhler disease** |
| **Painful swelling on the upper outer side of the heel** | |
| Painful swelling on pressure on the upper outer side of the heel | **Haglund deformity** (**calcaneal prominence**) |
| **Painful swelling on the inner side of the midfoot** | |
| Painful swelling just in front and under the medial malleolus | **Symptomatic external tibial bone** (**accessory navicular bone**, **secondary navicular bone**) |
| | Type I |
| | Type II |
| | Type III |
| **Painless swelling on the medial side of the footsole** | |
| Painless swelling on the anteromedial side of the heel | **Precalcaneal congenital fibrolipomatous hamartoma** |
| Painless swelling on the medial border of the foot near the highest point of the arch | **Plantar fibromatosis** (**Ledderhose disease**) |

# Toes

# 14

## Abnormal Toe

### Extra Toe

Complaint: extra toe.

Assessment: there are more than 5 toes.

Differential diagnosis:

**polydactyly**
- tibial polydactyly (preaxial polydactyly, extra big toe)
- fibular polydactyly (postaxial polydactyly, extra little toe)
- central polydactyly (extra middle toe)

**fibular dimelia (mirror foot)**

Explanatory note: **polydactyly.** This is a relatively frequent congenital abnormality. The incidence is 1.7 in a 1000 births. In 50 % of the cases the abnormality is bilateral, but does not need to be symmetrical. In 34 % of the cases there is also polydactyly in the hands. Toe polydactyly may occur in combination with syndactyly or other congenital deformities such as aplasia of the tibia and can be part of a syndrome (Table 14.1).

Tibial polydactyly. An extra big toe is less frequent than an extra little toe and there also may be a hallux varus (Fig. 14.1).

Fibular polydactyly. In 80 % of the cases the extra toe is on the lateral side (Fig. 14.2).

Central polydactyly. An extra middle toe is rare.

**Fibular dimelia** Fibular dimelia is also called mirror foot because the fibular part of the lower leg and the foot are mirrored. This is a very rare deformity occurring probably in 1 in 1,000,000 births. There is a duplication of the fibula and the fibular side of the foot, whilst the tibia and tibial part of the foot are absent (Fig. 14.3). The big toe is always absent. The parts of the mirror foot do not need to be completely symmetrical. The knee and the ankle are absent and the lower leg is usually shortened and the foot has an equinovarus deformity. The upper leg may also be too short.

There may also be an ulnar dimelia and a proximal focal femoral deficiency. Apart from these other congenital abnormalities of the skeleton may be present such as a scoliosis, a synostosis between the radius and ulna, hand abnormalities such as a thumb with 3 phalanges (triphalangeal thumb). Other congenital abnormalities may frequently be present such as a ventricular septum defect, an absent kidney and undescended testicles.

Supplementary assessment: X-rays of both lower extremities.

Primary care treatment: none.

**Table 14.1** Syndromes with polydactyly

| |
|---|
| Ellis-Van Creveld syndrome |
| Jeune syndrome |
| Trisomy 13 |
| Trisomy 18 |

See appendix for details of these syndromes


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_14

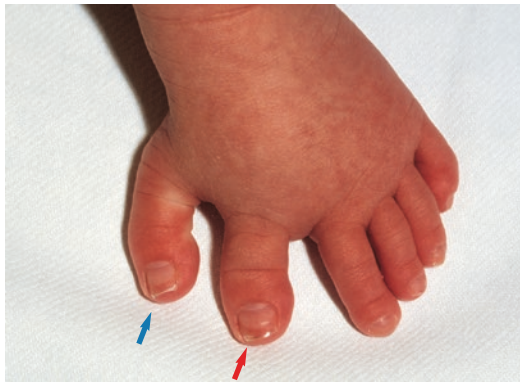


**Fig. 14.1** Tibial polydactyly (extra big toe). Next to the extra big toe (*blue arrow*) there is also a hallux varus (*red arrow*)

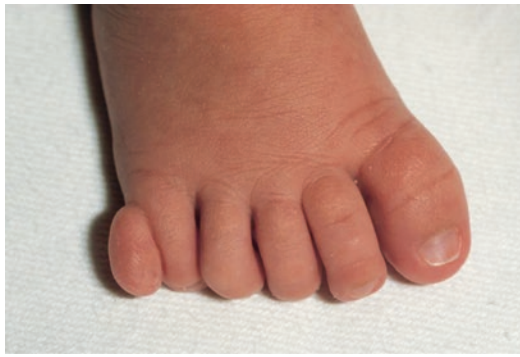


**Fig. 14.2** fibular polydactyly (*extra little toe*)

When to refer: the most optimal time for operative treatment is between 9 and 12 months of age. Therefore a child should be referred earlier.

Secondary care treatment: **polydactyly.** Surgery is recommended between 9 and 15 months of age. The most medial big toe or most lateral small toe will generally be removed notwithstanding the fact that these may look more normal than the adjacent toe. In a central polydactyly ligaments may need to be transfered in to different positions in order to prevent the forefoot from becoming deformed or widened. Following central polydactyly surgery a cast will be left in place for about 6 weeks.

**Fibular dimelia** An exarticulation through the knee joint will be carried out if there is a normal femur. A fusion will be carried out between the femur and one of the fibulae if the femur is shorter than normal as in a possible present proximal focal femoral deficiency. A Syme amputation can then be carried out at the level of the talocrural joint.

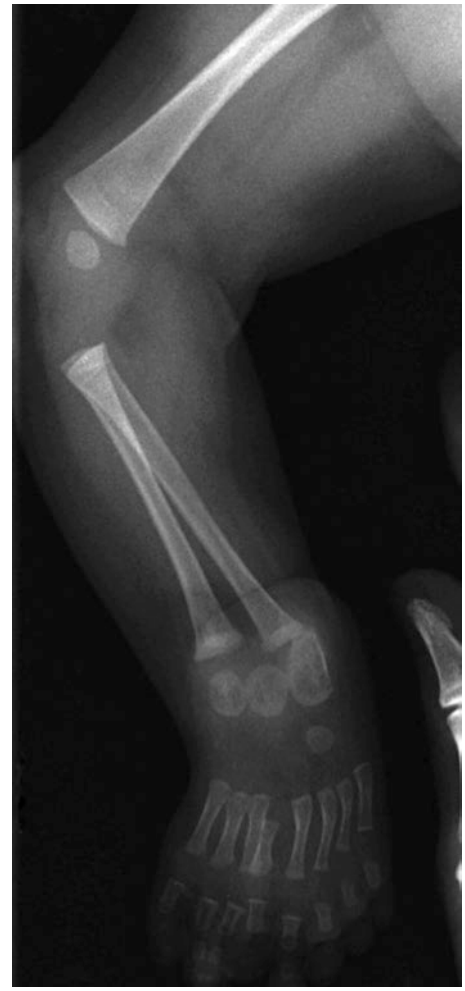


**Fig. 14.3** fibular dimelia (mirror foot). Absent tibia and medial part of the foot. Duplication of the fibula and lateral part of the foot. In this case 8 toes

## Giant Toe Growth

Complaint: one or more toes are obviously much bigger than normal at birth.

Assessment: hypertrophy of one or more toes.

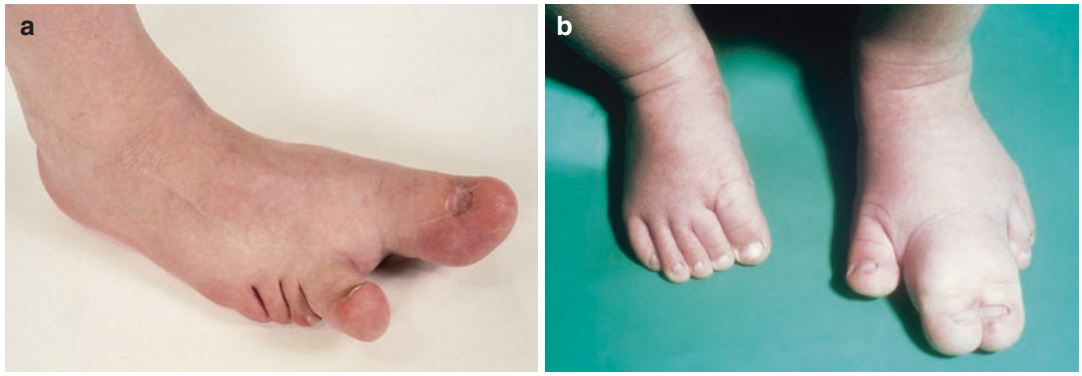



**Fig. 14.4** (**a**) Macrodactyly of the right big and second toes. (**b**) Macrodactyly of the left big, second and third toes, with a syndactyly of the second and third toes

Differential diagnosis:

**Macrodactyly**
Static type
Progressive type

Explanatory note: **macrodactyly.** The involved toes are about 1.5 times longer and broader than normal at birth. Giant growth appears mostly in the second toe followed by the big toe. The hypertrophy involves not only the bony structures but also the soft tissues including the nerves. The nerves may sometimes be 10 times as thick as normal. There is also a syndactyly in 10 % of the cases (Fig. 14.4).

Initially the joints of the involved toe (toes) move normally. At the end of the growth period they are generally stiff and early arthritis particularly in the interphalangeal joints can occur. Macrodactyly is usually unilateral. There are 2 types: a static and a progressive type.

Static Type. The size of the involved toe (toes) remains constant during growth with regard to the rest of the foot.

Progressive Type. The involved toe (toes) will become disproportionally larger than the rest of the foot during growth. The deformity usually appears as an isolated condition. The cause is unknown. However, macrodactyly can be a part of a syndrome such as: neurofibromatosis[1] (Von Recklinghausen disease), Klippel-Trenaunay-Weber syndrome[1] and the Proteus syndrome[1]. This is almost always part of a syndrome if the macrodactyly is present in more than one extremity or in the case of proximal extension with the passage of time.

Supplementary assessment: X-rays of the foot.

Primary care treatment: none.

When to refer: the parents usually want early advice from a specialist.

Secondary care treatment: **macrodactyly.** Epiphysiodesis of the involved metatarsal and proximal phalanx with debulking of the extra soft tissues in girls when the shoe size reaches 38 and in boys 40 (European shoe size). Part of the end of the toe can be amputated if necessary or the metatarsal bone can be shortened. A ray resection is the best treatment if one metatarsal bone is involved in this process but not if the first ray is involved. A first ray resection is contraindicated because the first ray has an important function when walking. In that case more operations are often necessary in order to remove the hypertrophic soft tissues. In extreme cases a Syme amputation at the level of the talocrural joint may be necessary.

## Bent Toe Deformity

Complaint: the parents complain about the child's bent toe.

Assessment: flexed toe.

[1] See Appendix.

Differential diagnosis:

**curly toe**
**hammer toe**
**mallet toe**
**claw toe**

Explanatory note: **curly toe.** The curly toe is the most frequent inborn error of the little toes. Usually it is a familial problem which appears bilaterally and in general gives no complaints. The most involved toes are the fourth and fifth toe. The distal phalanx of the toe is medially deviated, flexed and externally rotated. The involved toe lies under the medial toe next to it (Fig. 14.5). The deformity is most obvious when the child stands. This anomaly improves spontaneously in 25 % of cases. The children have no complaints in most cases, and a flexor tenotomy is rarely required for a persistant deformity.

**Hammer toe** A congenital hammer toe is characterized by extension in the metatarsophalangeal joint, flexion in the proximal interphalangeal joint and extension in the distal interphalangeal joint (Fig. 14.6). The hammer toe has no rotational component in contrast to the congenital curly toe. The deformity is often bilateral and is a familial problem. It usually involves the second toe.

**Mallet toe** In a congenital mallet toe there is a flexion deformity in the distal interphalangeal joint usually involving one toe (Fig. 14.6). Normally there are no complaints in young children. Complaints generally occur in young adults.

**Claw toe** A claw toe is characterized by hyperextension at the level of the metatarsophalangeal joint that is often subluxated dorsally with flexion at the level of the proximal and distal interphalangeal joints (Fig. 14.7).

Usually all of the toes are involved in contrast to the mallet toe often including the big toe with a flexion contracture at the level of the interphalangeal joint. Claw toes are often seen in pes cavus (hollow feet) and neurological conditions such as Charcot-Marie-Tooth disease, encephalopathy and after poliomyelitis.

Supplementary assessment: none.

Primary care treatment: treatment is not necessary if there are no complaints.

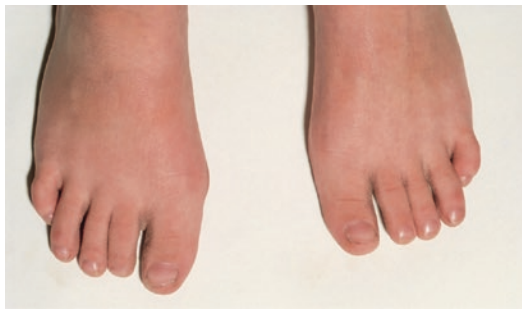


**Fig. 14.5** Curly toes of the right and left little toe

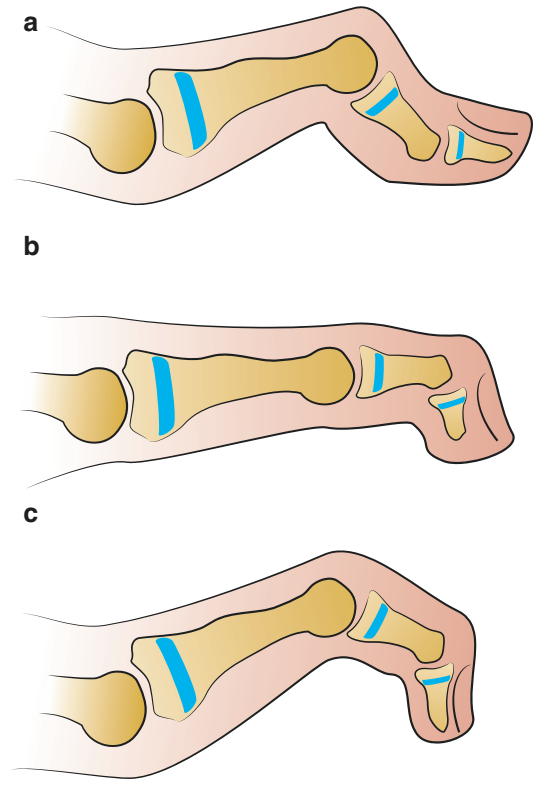



**Fig. 14.6** (**a**) Hammer toe. (**b**) Mallet toe. (**c**) Claw toe

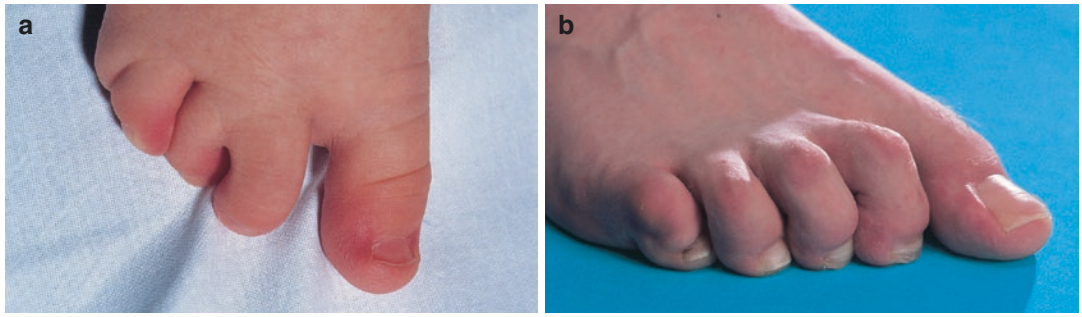



**Fig. 14.7** Claw toes. (**a**) During childhood and (**b**) In adulthood

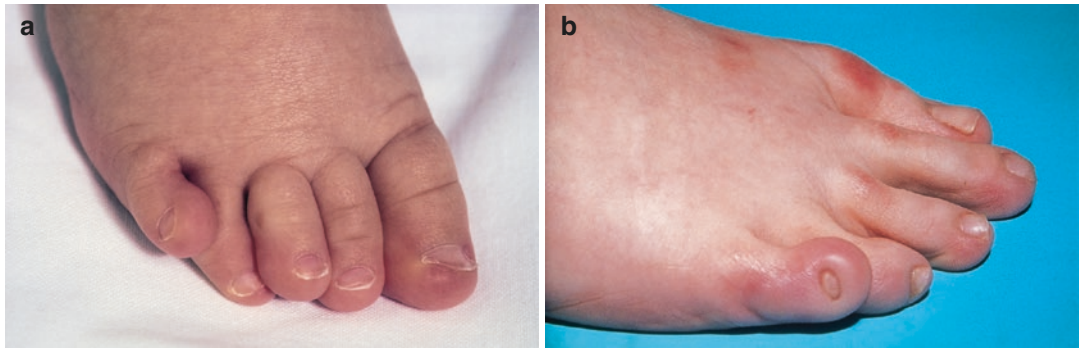



**Fig. 14.8** Overlapping of the little toe over the fourth toe (digitus quintus superductus). (**a**) In childhood. (**b**) In adulthood

When to refer: only with complaints. The complaints are usually caused by pressure areas as a result of the abnormal position of the toes.

Secondary care treatment: **curly toe.** Tenotomy of the flexor digitorum longus tendon and possibly flexor digitorum brevis, in children older than 6 years of age with complaints.

**Hammer toe and claw toe** Tenotomy of the flexor digitorum longus tendon with lengthening of the extensor tendon if necessary. A resection arthrodesis of the proximal interphalangeal joint and fixation with a Kirschner wire or a toe fixation nail during 3–4 weeks in young adulthood.

**Mallet toe** Tenotomy of the flexor digitorum longus. A resection arthrodesis of the distal interphalangeal joint in young adulthood.

## Overlapping Toe

Complaint: one toe lies above another toe.

Assessment: the little toe usually lies on top of the fourth.

Diagnosis: **digitus superductus (overlapping toe)**

Explanatory note: **digitus superductus.** Overlapping toes is an inborn error generally involving the little toe which lies over the fourth toe, so-called digitus quintus superductus (Fig. 14.8). The deformity is usually familial and is generally bilateral. The digitus quintus superductus gives problems at puberty when wearing shoes in about half of the cases. The toes can also lie above or under the toe next to it, usually the second toe crossed over or under the third toe (Fig. 14.9). The other toes usually do not cause complaints in children in contrast to the little toe.

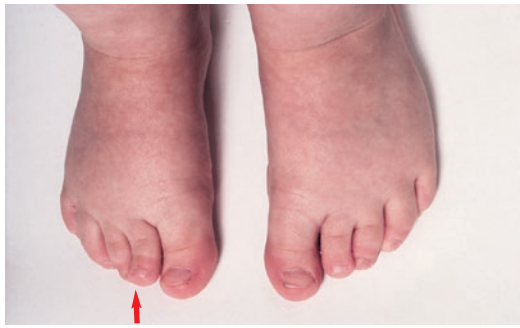


**Fig. 14.9** Overlapping of the right second toe on the third toe (digitus superductus)

Supplementary assessment: none.

Primary care treatment: conservative methods, such as orthoses and tape, are ineffective.

When to refer: only if there are complaints.

Secondary care treatment: **digitus superductus.** Many operations have been thought of to deal with this problem. The Butler operation is most frequently performed. A racket incision is made with the handle on the dorsal side with a tenotomy of the extensor tendon and an incision through the capsule on the dorsal and medial side of the metatarsophalangeal joint. The toe can now be easily corrected. The skin will be sutured in the corrected position. In another method the proximal phalanx will be completely or partly excised. In that case the toe becomes floppy and it is a good idea to suture the fifth toe partially onto the fourth toe (artificial soft tissue syndactyly).

## Short Toes

Complaint: one or more toes are too short.

Assessment: one or more toes are too short (Fig. 14.10).

Diagnosis: **brachydactyly**

Explanatory note: **brachydactyly.** In the foot the fourth ray is usually shortened. The involved metatarsal bone (or metatarsals) is (are) shorter than the other metatarsals. Generally it is part of an autosomal inborn deformity. Brachydactyly can also be part of a syndrome such as pseudohypoparathyroidism[2] (Albright's hereditary osteodystrophy).

Supplementary assessment: not necessary.

Primary care treatment: none.

When to refer: this is usually a cosmetic problem. Referral can be made in rare cases when there are functional problems.

Secondary care treatment: **brachydactyly.** A lengthening of the involved metatarsal may be considered in exceptional cases. The metatarsal can be lengthened directly 10 mm in one operation or 30 mm can be achieved with gradual distraction.

## Fused Toes

Complaint: 2 or more toes are fused.

Assessment: 2 or more toes are fused.

Differential diagnosis:

**syndactyly**
simple type
complex type

Explanatory note: **syndactyly.** Toe syndactyly occurs mostly between the second and third toe (Fig. 14.11). A syndactyly of the toes is generally not a functional problem.

Simple type. Syndactyly can be simple in which only the soft tissues are involved.

Complex type. In a complex type the bones are also fused. In both situations the syndactyly can be complete in which case the toes are completely fused over the whole length or incomplete in which case the toes are fixed together proximally but not distally. A syndactyly is often isolated but can be seen in combination with polydactyly and brachydactyly and can also be part of a syndrome (Table 14.2).

Supplementary assessment: foot X-ray.

Primary care treatment: none.

[2] See Appendix.

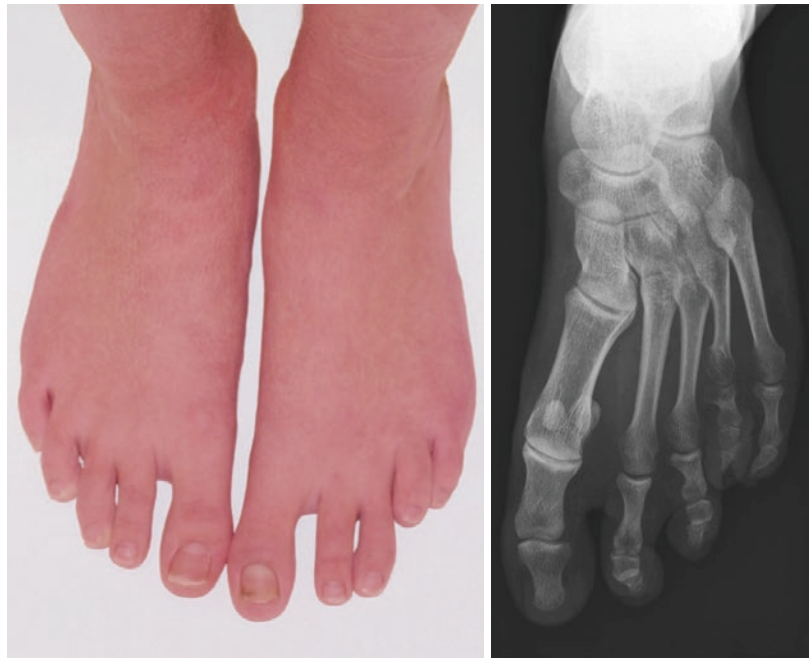


**Fig. 14.10** Brachydactyly involving the fourth ray of the left foot

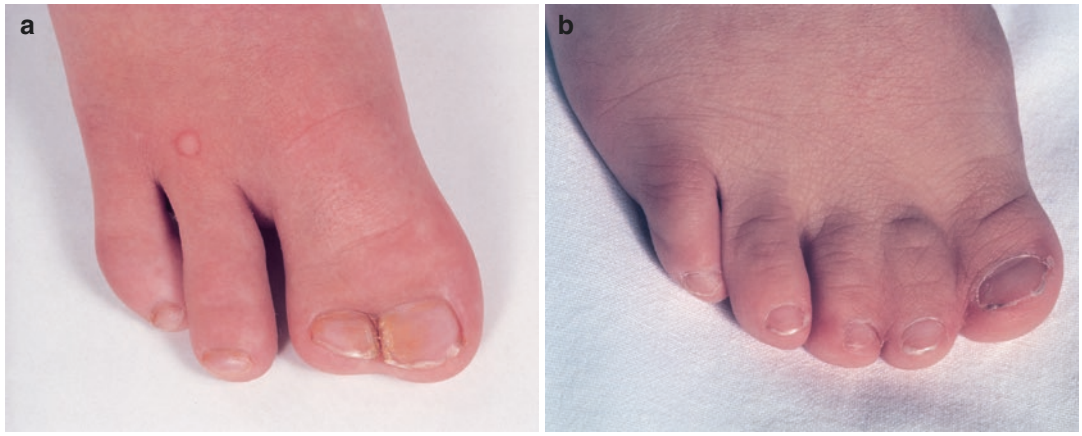



**Fig. 14.11** (**a**) A syndactyly of the big toe and second toe. In this case there is no fifth ray. (**b**) Syndactyly of the large, second and third toes

**Table 14.2** syndromes with toe syndactyly

| |
|---|
| Acrocephalopolysyndactyly |
| Apert syndrome |
| Cornelia de Lange syndrome |
| Goltz syndrome |
| Orofaciodigital syndrome I and II |
| Trisomy13 |
| Trisomy 18 |

See Appendix for details of these syndromes

When to refer: referral is not necessary if there are no associated anomalies such as polydactyly or a specific syndrome that has to be treated for other reasons.

Secondary care treatment: **syndactyly.** Operative correction of a toe syndactyly is not advisable and is not really necessary. It is basically a cosmetic problem.

## The Big Toe Deviates Medially

Complaint: the big toe deviates medially.

Assessment: the big toe is in varus.

Differential diagnosis:

**Metatarsus primus varus**
**hallux varus**
Type I
Type II
Type III

Explanatory note: **metatarsus primus varus.** The first ray is adducted with regard to the rest of the foot (for explanatory note, see p. 261).

**Hallux varus** Three types can be distinguished. In all types there is a short and broad first metatarsal and a bracket shaped longitudinal epiphysis on the medial side. The adducted position of the great toe is caused by a tight fibrous band on the medial side of the first metatarsophalangeal joint. The varus position varies from a few degrees up to 90° (Fig. 14.12).

Type I. In this type there are no other congenital anomalies.

Type II. There are also other congenital foot deformities. Apart from a shortened and broad first metatarsal there may also be a duplication of the big toe (Fig. 14.2).

Type III. This hallux varus is also accompanied by other inborn anomalies in other parts of the body such as in skeletal dysplasia or diastrophic dwarf growth.

Supplementary assessment: foot X-rays.

Primary care treatment: none.

When to refer: in cases of a rigid metatarsus primus varus and in the case of a hallux varus.

Secondary care treatment: **metatarsus primus varus.** A rigid metatarsus primus varus is treated with manual corrections and lower leg plaster cast immobilization. The foot will then be treated at night during 1–2 years with a boot splint after the foot position has been corrected.

**Hallux varus** In a hallux varus the tight medial fibrous band on the medial side of the metatarsophalangeal joint is divided, the capsule on the lateral side of this joint is reefed, the tendon of the abductor hallucis muscle is lengthened, and if necessary extra toe bones or an extra big toe is removed. The central part of the staple shaped longitudinal epiphysis will be removed. The defect will be filled with fat or temporarily with bone cement (Fig. 14.12).

## The Big Toe Deviates Laterally

Complaint: generally involves a girl between 10 and 16 years of age who finds that her foot is ugly because the big toe deviates laterally. This is not usually painful.

Assessment: the big toe is positioned in valgus.

Differential diagnosis:

**Metatarsophalangeal hallux valgus**
**Interphalangeal hallux valgus**

Explanatory note: **metatarsophalangeal hallux valgus.** 90 % of cases are girls with a more than normal valgus position at the level of the first metatarsophalangeal joint (Fig. 14.13). The deformity is usually bilateral. Normally there is a valgus position in the great toe 5–15° with respect to the first metatarsal. There is a hallux valgus if the angle is more than 15°.

The severity of the hallux valgus can be subdivided into a mild type with a valgus angle between 15° and 25° and a moderate to severe type in which the valgus angle is more than 25°.

A hallux valgus is caused by slackening of the soft tissues on the medial side of the first metatarsophalangeal joint and an erosion of the ridge on the plantar side between the medial and lateral sesamoid bones. The proximal phalanx of the big toe is displaced laterally and the first metatarsal

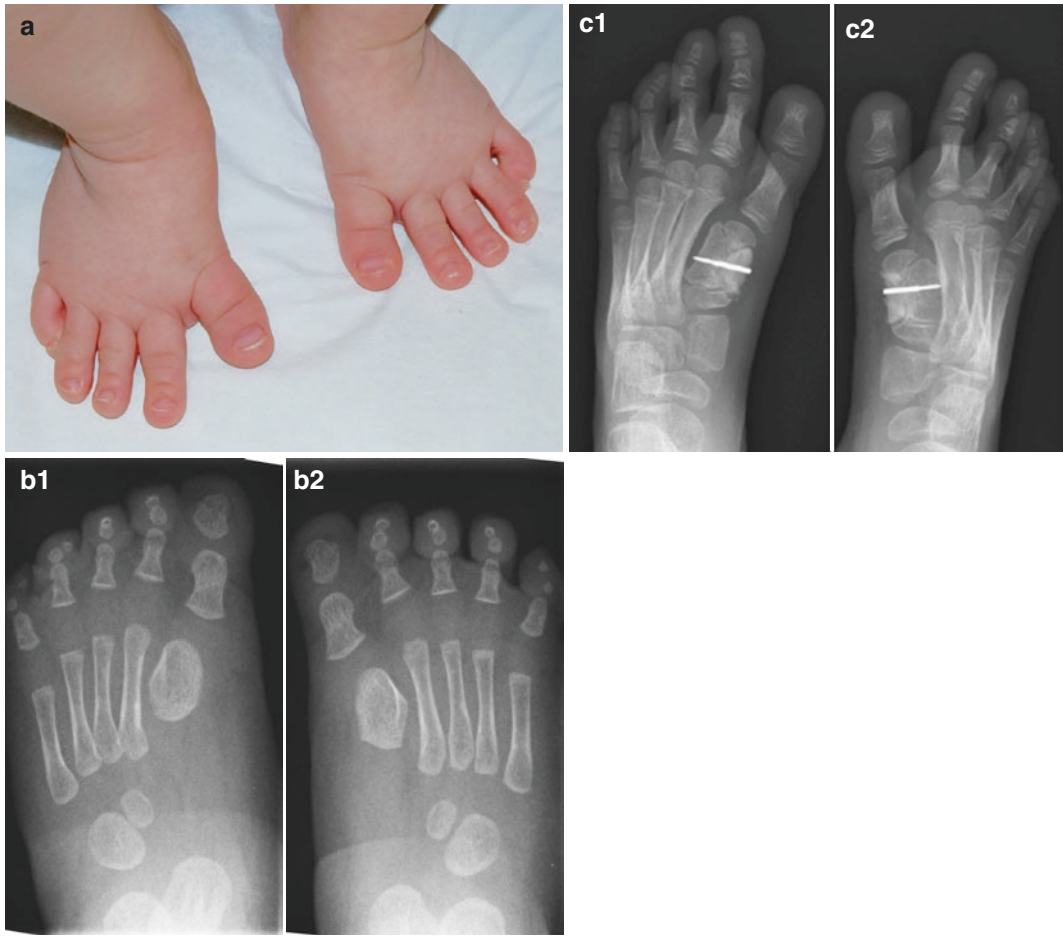



**Fig. 14.12** (**a**) Bilateral hallux varus in a 1.5 year old child. (**b**1, **b**2) Foot X-rays (the same child as in **a**). There is a severely shortened and broad bilateral first metatarsal. (**c**1, **c**2) Bilateral removal of the central area of the staple shaped longitudinal epiphysis, temporarily filled with bone cement and fixed with a Kirschner wire in a 6 year old child

medially. The angle between the first and second metatarsals (intermetatarsal angle) increases (Fig. 14.14). As a result of the increased pressure the bursa on the medial side of the first metatarsal head becomes irritated when wearing insufficiently broad footwear. This is called a painful bunion. Occasionally this is incorrectly described as an exostosis. As a result of the medial displacement of the first metatarsal, the sesamoid bones do not lie symmetrically under the first metatarsal head. The medial sesamoid bone lies under the eroded ridge and the lateral sesamoid bone articulates with the lateral side of the first metatarsal head. The extensor hallucis longus and flexor hallucis longus tendons lie lateral to the first metatarsal head and as a result function as big toe adductors where by the deformity increases. Furthermore, the deformity is made worse due to shortening of the adductor hallucis and the flexor hallucis brevis muscle and the lateral joint capsule of the first metatarsophalangeal joint.

As a result of this a subluxation of the first metatarsophalangeal joint occurs. This is described as an incongruent hallux valgus. This subluxation causes a groove to appear on the medial side of the first metatarsal head with the passage of time. There is also a dorsiflexion and pronation deformity in the big toe in severe cases of hallux valgus of 30–35°. As a result of this the first

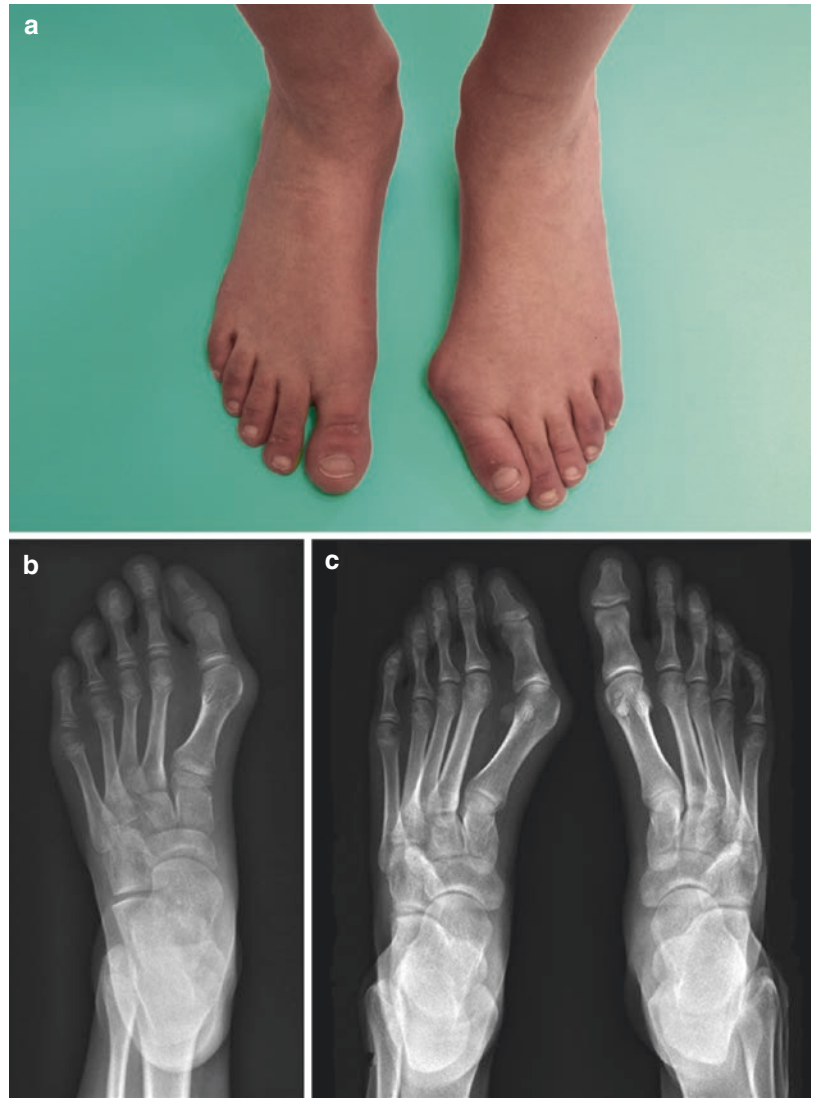



**Fig. 14.13** (**a**) Left metatarsophalangeal hallux valgus in a 10 year old child. (**b**) X-ray of the left foot (the same patient as in **a**). (**c**) X-rays of both feet after the child has stopped growing

metatarsal head bears less weight and this results in overloading of the other heads, particularly the second metatarsal head (transfer metatarsalgia). A claw toe deformity can be seen possibly involving a dislocation in the second metatarsophalangeal joint in serious cases.

As a rule an increased intermetatarsal angle (IMA) is seen in adolescents with a hallux valgus. This angle should not be 10° or more. As a rule a subluxation of the first metatarsophalangeal joint is seen if the intermetatarsal angle is greater than 15° (Fig. 14.14).

In 2 % of cases there is an angulation between the axis of the first metatarsal and the distal joint surface of the first metatarsal (DMAA = Distal Metatarsal Articular Angle). This angle should not be 10° or more (Fig. 14.14). The DMAA is difficult to measure because of large inter observer differences. There is a hypermobile joint between the medial cuneiform bone and the first metatarsal in 2–3 % of cases. This is one of the causes of a hallux valgus. In order to check the hypermobility of the first tarsometatarsal joint the examiner immobilizes the second to fifth metatarsals with the thumb and fingers of one hand using the thumb and index finger of the other hand to grasp the first metatarsal and move it from a plantar lateral to a more dorsomedial

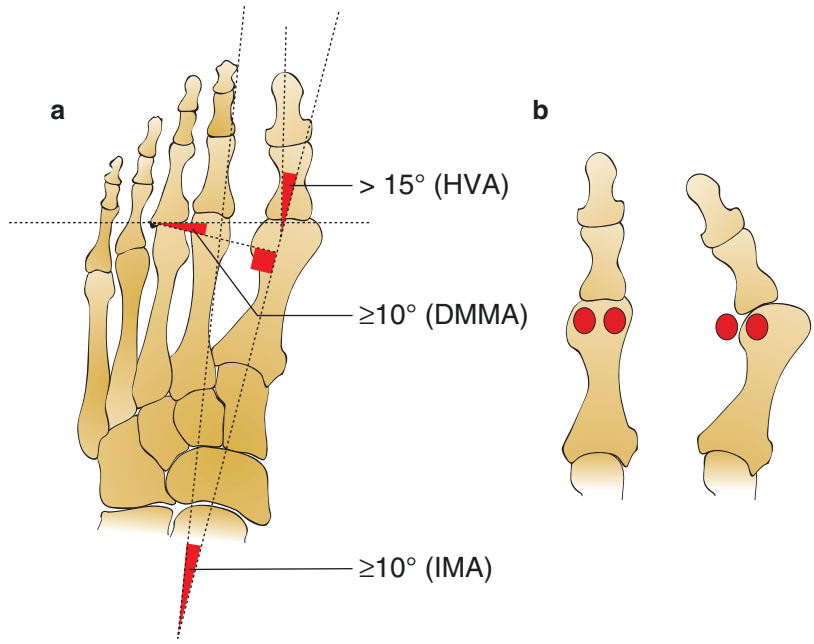



**Fig. 14.14** (**a**) There is a hallux valgus if the angle between the first metatarsal and the big toe is more than 15° (*HVA*). There is usually an increased intermetatarsal angle between the first and second metatarsals in children with a hallux valgus (*IMA* inter metatarsal angle). This angle should not be greater than 10°. There is a metatarsus primus varus if this angle is 10° or more. There is usually no increase in the DMAA (Distal Metatarsal Articular Angle) as an underlying cause in children. The angle between the central axes of the first metatarsal and the distal joint surface of the first metatarsal bone may not be 10° or more. (**b**) The proximal phalanx of the big toe is displaced laterally and the head of the first metatarsal medially. As a result the medial sesamoid bone lies under the eroded edge of the first metatarsophalangeal joint and the lateral sesamoid bone articulates with the lateral side of the first metatarsal head

direction. A movement of more than 9 mm indicates hypermobility in the first tarsometatarsal joint. This examination is not very accurate.

A hallux valgus in adolescents can be distinguished from hallux valgus in adults according to the following points:

- There is a familial occurrence in 70 % (usually on the mother's side of the family)
- Pain is mostly not the primary complaint
- Correction is usually asked for cosmetic reasons
- There is usually a metatarsus primus varus
- The bunion (thickening of the soft tissues on the medial side of the first metatarsal head) is usually not very prominent
- Hallux valgus is often seen in combination with a flatfoot and pronation of the forefoot
- There may also be an interphalangeal hallux valgus apart from the metatarsophalangeale hallux valgus
- There is no degeneration in the first metatarsophalangeal joint
- There is a great chance of recurrence after operative treatment

**Interphalangeal hallux valgus** In this case there is a valgus position at the interphalangeal joint. The upper limit of the interphalangeal hallux valgus is 10º. The deformity varies in severity. In moderate and severe cases pressure areas develop at the level of the interphalangeal joint (Fig. 14.15).

Supplementary assessment: anteroposterior X-rays of the forefoot (Fig. 14.15).

Primary care treatment: advice is given to buy footwear that is sufficiently broad enough otherwise there will be pain as a result of the pressure point (s) or ask the cob-

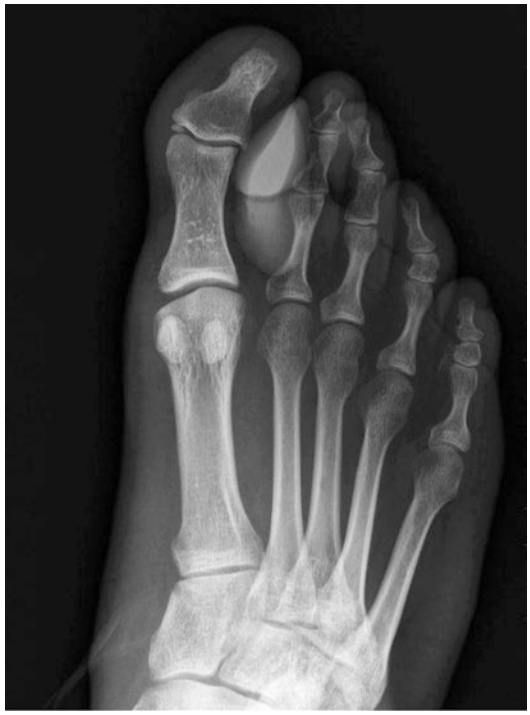


**Fig. 14.15** Interphalangeal hallux valgus of the right big toe

bler to widen the shoe at the level of the pressure point(s).

When to refer: if there is a valgus deformity of the big toe of more than 15° and if the interphalangeal hallux valgus still causes pressure notwithstanding a sufficiently broad shoe.

Secondary care treatment: **metatarsophalangeal hallux valgus.** Operative treatment of a hallux valgus in children is almost always postponed until the child has finished growing because of the high risk of recurrence. There are more than 130 operations described for the treatment of hallux valgus. The patients should be warned that 40% will not be able to wear the shoes that they would prefer. 25–33% of patients will not be satisfied with the result. As a rule a corrective osteotomy of the first metatarsal is adequate in the case of a congruent hallux valgus (no subluxation of the first metatarsophalangeal joint). An additional soft tissue procedure is indicated in the case of an incongruent hallux valgus.

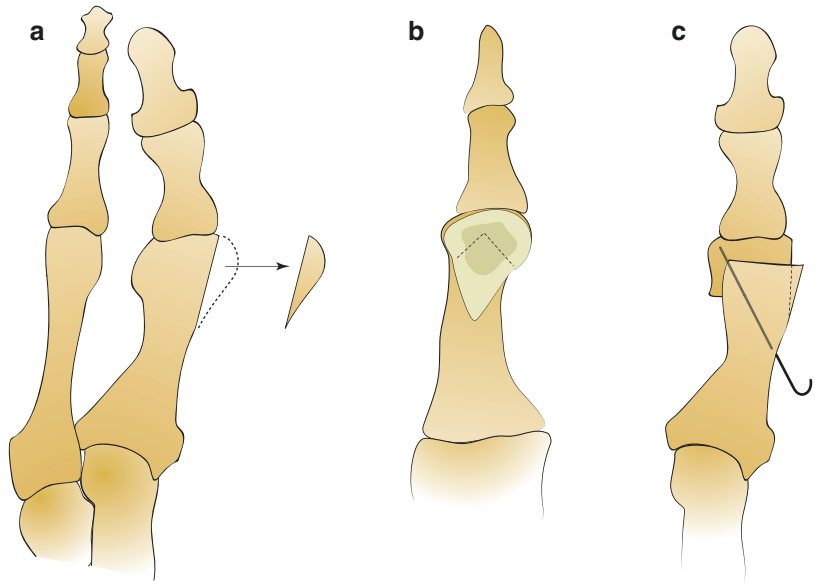



**Fig. 14.16** Chevron osteotomy of the distal part of the first metatarsal. (**a**) Frontview: in line with the medial border of the diaphysis of the first metatarsal, removal of the medial eminence of the head, relieving pressure on the bunion. (**b**) Lateral view: chevron osteotomy (*dotted line*). (**c**) Front view: pushing the distal part laterally and fixation of the osteotomy with a Kirschner wire. Removal of surplus of bone on the medial side. Reefing the medial part of the capsule of the first metatarsal joint

In a soft tissue procedure (DSTP: Distal Soft Tissue Procedure or Mc Bride procedure) a release is carried out of the adductor hallucis muscle and the lateral joint capsule of the first metatarsophalangeal joint. The lateral ligament must be left intact otherwise this can lead to a hallux varus. The medial capsule is reefed after carrying out a VY plasty with the base of the V distal. As a rule it is not necessary to shave the medial side of the first metatarsal head. Too much shaving can lead to damage in that joint causing stiffness in the first metatarsophalangeal joint. Only a soft tissue release is inadequate in the case of an incongruent hallux valgus. Recurrence occurs in 50-70 % of cases. Apart from the soft tissue procedure an osteotomy of the first metatarsal must be carried out.

In the case of a large intermetatarsal angle one can choose between a distal, a diaphyseal or a proximal osteotomy. Standard techniques are: a chevron for a distal (Fig. 14.16), a scarf osteotomy (named after the woodworking equivalent) (Fig. 14.17) in the case of a diaphyseal osteotomy and a crescent shaped osteotomy in the case of a proximal procedure (Fig. 14.18). A distal osteotomy will be carried out in the case of a limited or moderate intermetatarsal angle. There is an increased chance of ischemic necrosis due to damage to the nutrient artery on the lateral side of the neck of the first metatarsal when carrying out a distal osteotomy. This ischemic necrosis is as a rule asymptomatic.

A diaphyseal osteotomy such as the scarf osteotomy will be carried out in the case of a moderate to severe type of hallux valgus. In the same situation a proximal osteotomy of the first metatarsal can be carried out.

In this case one has the choice between an open wedge, a closed wedge or a crescent shaped osteotomy. There is a danger when carrying out a closed wedge osteotomy for shortening of the first metatarsal with as a result overloading of the second metatarsal head (transfer metatarsalgia). In an open wedge osteotomy a lengthening of the first metatarsal occurs with the chance of stiffness in the first metatarsophalangeal joint. A crescentic osteotomy prevents these problems. In the event of a pronounced pronation one can derotate at the level of the osteotomy. Correction of a too great distal metatarsal articular angle (DMMA) can be corrected by adapting the geometry of the chevron osteotomy (biplanar chevron osteotomy). This is also the case with the scarf osteotomy. However, this is not possible when carrying out a proximal osteotomy of the first metatarsal. When carrying

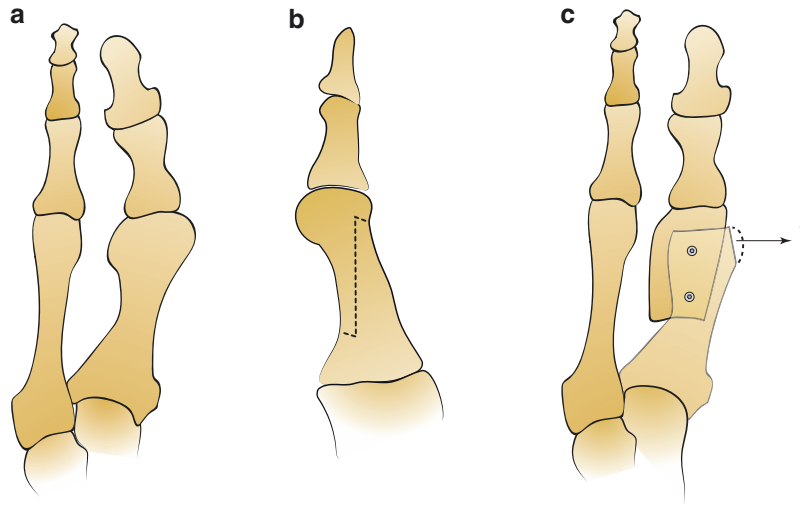



**Fig. 14.17** Scarf (diaphysial) osteotomy. (**a**) Front view of a hallux valgus. (**b**) Lateral view: Z-shaped osteotomy. (**c**) Front view: Lateral transposition of the most distal part. Osteotomy fixation with two small fragment screws. Removal of the excess bone from the medial side of the metatarsal head

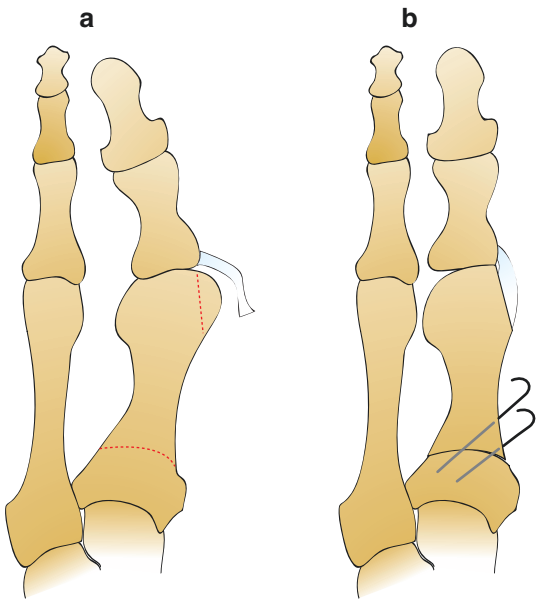



**Fig. 14.18** Crescenteric (proximal) osteotomy. (**a**) Front view of a hallux valgus, removal of the most medial part of the first metatarsal head in line with the medial border of the first metatarsal diaphysis. (**b**) Crescenteric osteotomy of the proximal part of the first metatarsal. Osteotomy fixation with a screw or two Kirschner wires

out a proximal first metatarsal osteotomy a closed distal osteotomy can be combined with a proximal open wedge osteotomy in the case of a large, distal metatarsal articular angle. When carrying out the distal osteotomy the wedge of bone that has been removed can be inserted into the side of the proximal osteotomy (double osteotomy according to Peterson and Newman) (Fig. 14.19).

Hypermobility of the first tarsometatarsal joint can be treated by carrying out an arthrodesis of this joint (Lapidus procedure) together with a distal soft tissue release.

**Interphalangeal hallux valgus** In this case a correction takes place at the level of the proximal phalanx. Preferably in adults in order to avoid damage to the growth plate (Fig. 14.20).

## The Little Toe Is Deviated Medially

Complaint: pain at the level of the head of the fifth metatarsal bone.

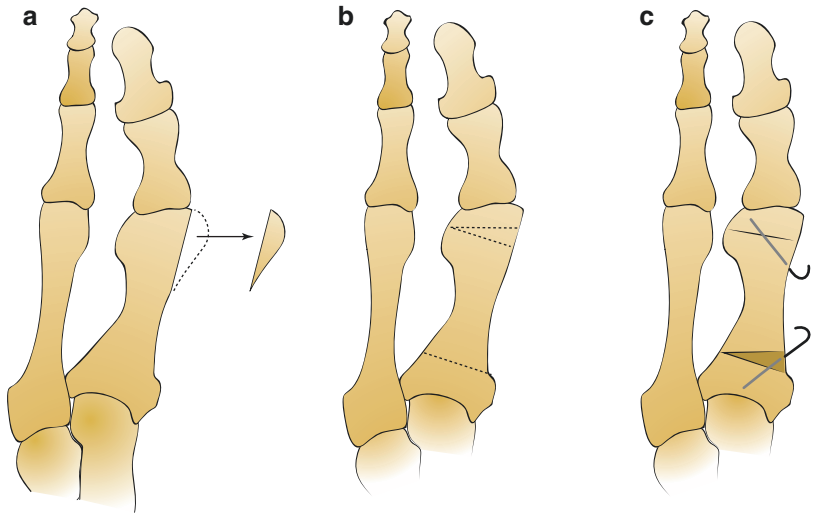



**Fig. 14.19** Double osteotomy according to Peterson and Neuman for a combined too great IMA and DMAA angle. (**a**) Removal of the most medial part of the first metatarsal head in line with the medial border of the diaphysis of the first metatarsal. (**b**) Closed wedge varus osteotomy in the distal part of the first metatasal. (**b**) and (**c**) Proximal first metatarsal open wedge valgus osteotomy with interposition of the bone obtained from the distal osteotomy. Fixation with a screw or two Kirschner wires (Redrawn from: Peterson HA, Newman SR. Adolescent bunion deformity treated with double osteotomy and longitudinal pin fixation of the first ray. *J Pediatr Orthop*. 1993;13:80–4)

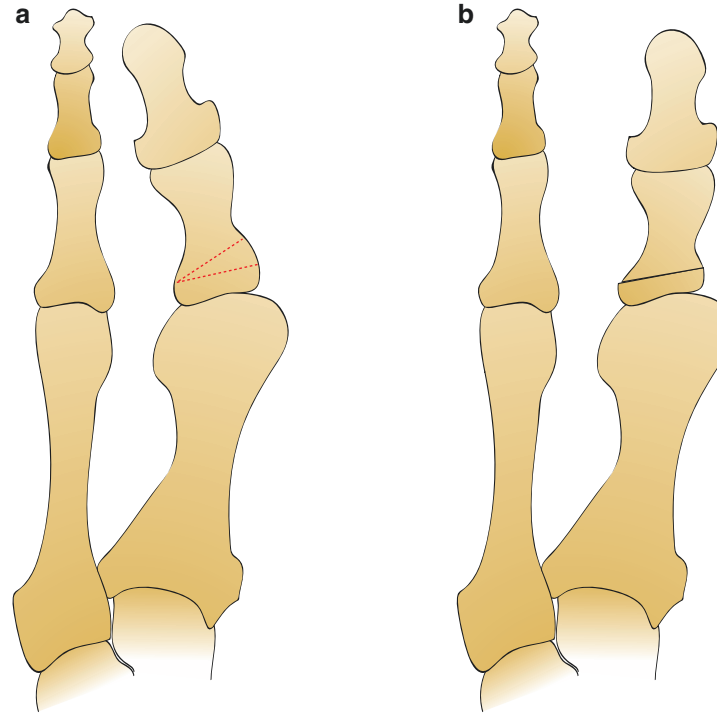



**Fig. 14.20** (**a**) and (**b**) Corrective osteotomy at the level of the proximal phalanx of the big toe because of an interphalangeal hallux valgus (osteotomy according to Akin)

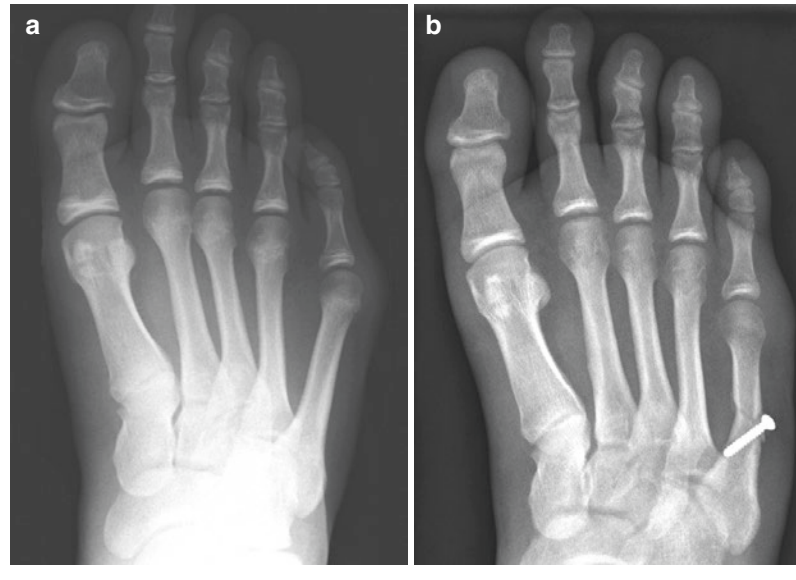



**Fig. 14.21** (**a**) Digitus quintus varus as a result of the metatarsus quintus valgus. (**b**) After a corrective osteotomy of the fifth metatarsal

Usually consultation takes place in young adulthood.

Assessment: broad forefoot. The little toe is deviated in varus.

Diagnosis: **tailor toe (digitus quintus varus, bunionette)**

Explanatory note: **digitus quintus varus.** Normally the metatarsals second up to the fifth are more or less parallel to each other. If the fifth metatarsal is in valgus then the little toe will be in varus (Fig. 14.21). There may be a painful bursa on the lateral side of the head of the fifth metatarsal caused by the footwear.

Supplementary assessment: Anteroposterior X-ray of the foot.

Primary care treatment: advise buying a sufficiently broad shoe. If this does not help sufficiently then the cobbler can be asked to widen the shoe at the level of the head of the fifth metatarsal.

When to refer: if there are still complaints notwithstanding the conservative measures mentioned above.

Secondary care treatment: **digitus quintus varus.** Correction osteotomy of the fifth metatarsal (Fig. 14.21).

## Painful Toes

### Painful Stiff Big Toe

Complaint: this usually involves children in adolescence who complain of pain and stiffness at the level of the joint between the first metatarsal and the big toe.

Assessment: plantar flexion and dorsal extension in the big toe at the level of the first metatarsophalangeal joint is limited and painful.

Diagnosis: **hallux rigidus**

Explanatory note: **hallux rigidus.** A hallux rigidus in children is often caused by an osteochondritis dissecans of the joint surface of the first metatarsal head (Fig. 14.22). In 10 % of cases a free body develops. In general, narrowing of the joints and osteophyte formation in the first metatarsophalangeal joint may be present if there is no osteochondritis dissecans focus. The cause of this early arthritis is not known. Sometimes there is a relatively long first ray.

Supplementary assessment: X-rays of the big toe.

Primary care treatment: advise sufficiently broad shoes to give the big toe more room. Furthermore a supportive insole or a support under the shoe may be prescribed (Fig. 14.23).

When to refer: if there are still complaints in adulthood notwithstanding supportive insoles or shoe modifications.

Secondary care treatment: **hallux rigidus.** Removal of any free body. Curetting the first

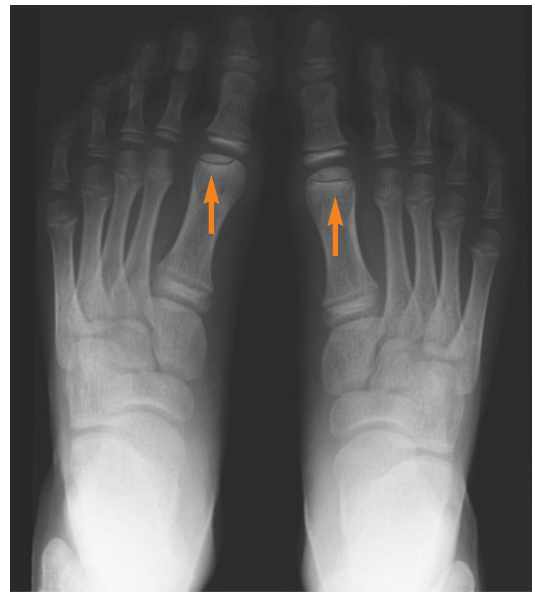


**Fig. 14.22** Osteochondritis dissecans of the heads of the both first metatarsals (*arrows*)

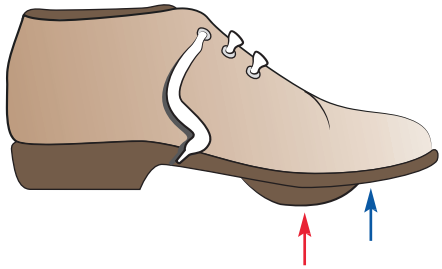


**Fig. 14.23** Supportive bar under the shoe. As a result of this bar (*red arrow*) there is less pressure on the painful metatarsophalangeal joints just in front (*blue arrow*)

metatarsophalangeal joint takes place in arthritis. Possible osteophytes present will also be removed. An arthrodesis of the first metatarsophalangeal joint may be considered in the case of insufficient results.

## Pain at the Level of the Head of the Second Metatarsal (or Third or Fourth or Fifth)

Complaint: this usually involves girls in adolescence older than 13 years of age with pain under the head of the second metatarsal and occasionally under the head of one of the other metatarsals.

Assessment: there is pain on pressure at the level of the head of one of the metatarsals. The mobility of the involved metatarsophalangeal joint is limited.

Diagnosis: **Freiberg disease**

Explanatory note: **Freiberg disease.** This involves an avascular necrosis of one of the heads of the metatarsals. It usually involves the second metatarsal but can also involve the others. It can also be bilateral. It involves girls in 75 % of cases. The avascular bone necrosis according to Freiberg is also known as Köhler II disease.

Supplementary assessment: anteroposterior X-rays of the foot can show ischemic bone necrosis in the head of the involved metatarsal (Fig. 14.24). The head of the metatarsal is flattened and irregular. A child may have complaints for 6 months before anything can be seen on an X-ray.

Primary care treatment: prescribing a supportive insole with a forefoot pressure pad or a shoe with a supportive bar (Fig. 14.23).

When to refer: if the supportive insole or bar gives insufficient support.

Secondary care treatment:

**Freiberg disease** Six weeks of lower leg plaster cast immobilization can be given to unburden the foot if the supportive insole has had no effect. After this a supportive insole with a forefoot pressure pad can be prescribed again. Curetting of the involved head can be considered if the pain continues notwithstanding conservative treatment.

## Toenail Pain

Complaint: toenail pain.

Assessment: palpable swelling beneath the toenail. Pressing on the toenail produces pain.

Diagnosis: **subungual exostosis**

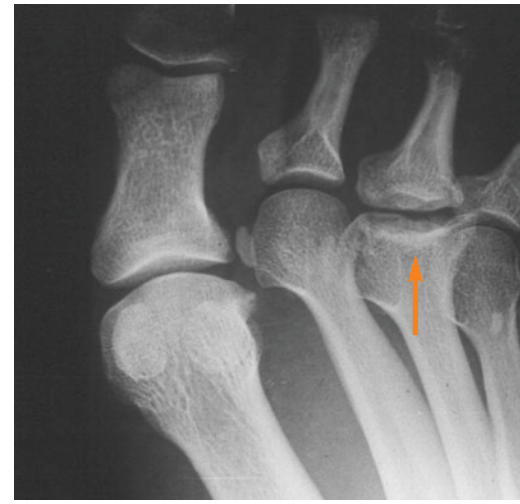


**Fig. 14.24** Freiberg disease (*arrow*). In this case there is an ischemic necrosis of the third metacarpal head (usually involves the second)

Explanatory note: histologically the swelling consists of trabecular bone covered with fibrocartilage. The reason why it occurs on the dorsal aspect of the distal phalanx is because the periosteum is loose in that area. This rare benign tumor occurs in the second decade of life and most commonly affects the big toe.

Supplementary assessment: x-ray of the toe.

Primary care treatment: none.

When to refer: if there is pain.

Secondary care treatment: careful excision of the tumor through a longitudinal incision in the nailbed and suturing the nailbed afterwards.

**Differential Diagnosis Abnormal Toes**

| | |
|---|---|
| **Extra toe** | **Polydactyly** |
| Extra big toe | Tibial polydactyly (preaxial polydactyly, extra big toe) |
| Extra little toe | Fibular polydactyly (postaxial polydactyly, extra little toe) |
| Extra middle toe | Central polydactyly (extra middle toe) |
| Duplication of the fibula and the fibular side of the foot | |
| The tibia and the tibial part of the foot are not present including the big toe and there are 7 or 8 toes | **Fibular dimelia (mirror foot)** |
| **Giant toe growth** | **Macrodactyly** |
| Proportional during growth | Static type |
| Progressive during growth | Progressive type |
| **Bent toe deformity** | |
| The distal phalanx of the toe is deviated medially, flexed and exorotated | **Curly toe** |
| Extension at the level of the metatarsophalangeal joint and flexion in the proximal interphalangeal joint. The distal interphalangeal joint is normal or hyperextended | **Hammer toe** |
| Positional flexion in the distal interphalangeal joint | **Mallet toe** |
| Hyperextension at the level of the metatarsophalangeal joint, flexion in the proximal and distal interphalangeal joint | **Claw toe** |
| **Overlapping toe** | |
| One toe lies on top of another toe | **Digitus superductus (overlapping toe)** |
| **Short toe** | |
| One or more toes are too short | **Brachydactyly** |
| **Fused toes** | |
| Two or more toes are fused together | **Syndactyly** |
| Only soft tissues are involved | Simple type |
| Bones are also fused | Complex type |
| **The big toe deviates medially** | |
| The first metatarsal bone and big toe are in adduction with regard to the rest of the foot | **Metatarsus primus varus** |
| The big toe deviates in varus | **Hallux varus** |
| In this type there are no other congenital anomalies | Type I |
| There are also other congenital foot deformities | Type II |
| Inborn anomalies in other parts of the body | Type III |
| **The big toe deviates laterally** | |
| Valgus position of the big toe at the level of the metatarsophalangeal joint | **Metatarsophalangeal hallux valgus** |
| Valgus position at the level of the interphalangeal joint | **Interphalangeal hallux valgus** |
| **The little toe deviates medially** | |
| Broad forefoot with a varus position in the little toe | **Tailor toe (digitus quintus varus, bunionette)** |

**Differential Diagnosis: Toe Pain**

| | |
|---|---|
| **Painful stiff big toe** | |
| Plantar flexion and dorsiflexion in the first metatarsophalangeal joint are limited and painful | **Hallux rigidus** |
| **Pain at the level of the head of the second metatarsal (or third or fourth or fifth)** | |
| Pain on pressure at the level of the involved metatarsal head | **Freiberg disease** |
| **Toenail pain** | |
| Painful swelling beneath the toenail | **Subungual exostosis** |

# Leg Length Inequality 15

## One Leg is Shorter or Longer than the Other

### One Leg is Shorter than the Other

Complaint: the parents notice that one of the child's legs is shorter than the other or they find that the child has an abnormal gait (Fig. 15.1).

Assessment: on examination leg length difference can be measured with the help of a method using planks.

Differential diagnosis:

congenital
- **idiopathic hemihypotrophy (hemihypoplasia)**
- **Russell-Silver syndrome**
- **congenital deficiency**
- **developmental dysplasia of the hip**
- **talipes equinovarus (pes equinovarus, clubfoot)**

vascular
- **Legg-Calvé-Perthes disease (Perthes disease, coxa plana, Waldenström disease)**
- **ischemic necrosis of the femoral head**

neurological
- **spastic hemiplegia**
- **poliomyelitis**
- **peripheral nerve or plexus injury**
- **plexus injury**

growth plate damage
- **acute osteomyelitis**
- **septic arthritis of the hip**
- **growth plate fracture**
- **slipped capital femoral epiphysis**
- **radiotherapy**

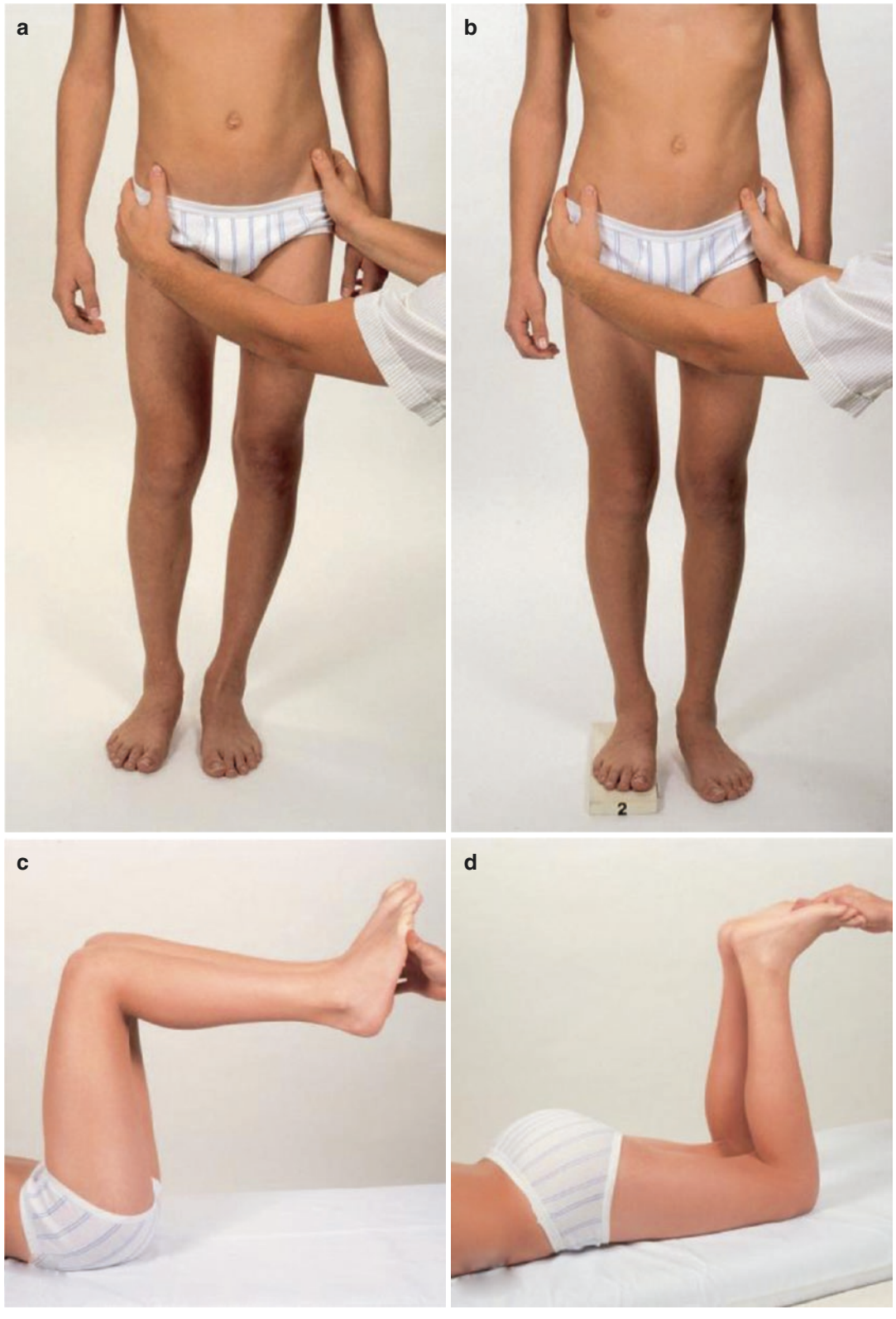
bone tumor
- **unicameral bone cyst (juvenile-, solitary-, simple bone cyst)**
- **osteochondroma (exostosis)**
- **enchondroma**
  - Ollier disease
  - Mafucci syndrome

Explanatory note: congenital

**Idiopathic hemihypotrophy** Hemihypotrophy is usually idiopathic but can also be part of the **Russell-Silver syndrome.**[1] Idiopathic hemihypotrophy can involve not only a complete body half but may involve just one extremity. We are dealing with hemihypotrophy if the short leg is thinner and does not fit in with the rest of the body. Hemihypotrophy occurs in 1 in 100,000 individuals. Apart from the difference in leg length other abnormalities may be present such as palate anomalies, cleft lip and palate, urogenital anomalies and mental retardation. The final difference in leg length is generally not more than 2 cm.

[1] See Appendix.


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_15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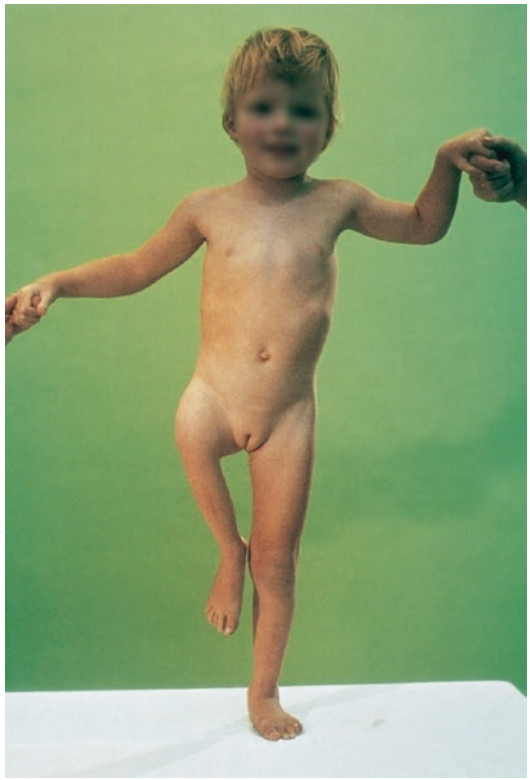


**Fig. 15.2** Congenital deficiency: in this case a proximal focal femur deficiency on the right side

**Congenital deficiency** In this case the whole lower extremity is missing or parts of it and in the latter case there is usually a severe difference in leg length (Fig. 15.2).

**Developmental dysplasia of the hip** An untreated unilateral hip dislocation will lead finally to a leg length difference of 5–6 cm (Fig. 15.3).

**Talipes equinovarus** A talipes equinovarus usually causes a leg length difference of less than 1 cm.

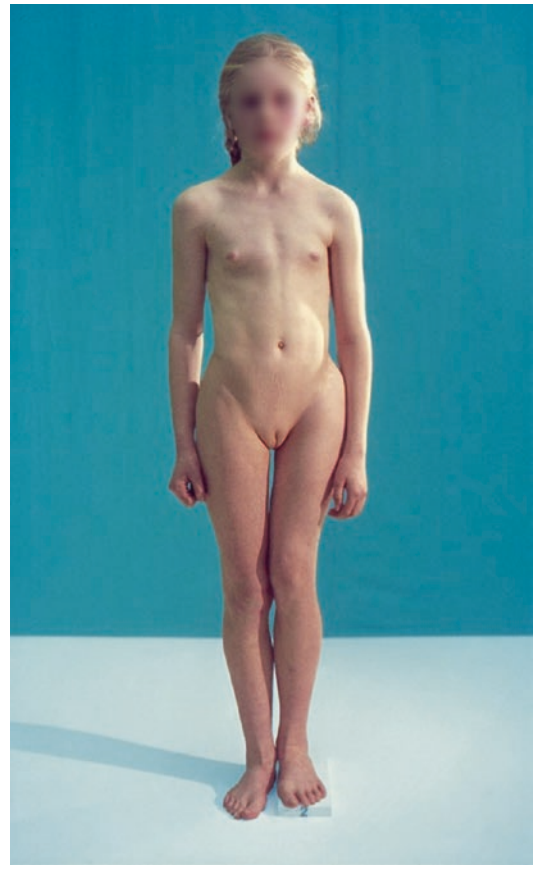


**Fig. 15.3** Leg length difference of 2 cm in the left thigh as a result of a developmental dysplasia with a dislocated left hip. The leg length difference can increase in adulthood up to 5 or 6 cm

## Vascular

**Legg-calvé-perthes disease** Perthes disease generally causes a leg length difference of not more than 1 cm.

**Ischemic necrosis of the femoral head** This can be a complication of a fracture in the proximal

**Fig. 15.1** Measurement of leg length difference using the plank technique. (**a**) The anterior superior iliac spines are palpated with the thumbs. If the thumbs are not at the same height there is a leg length difference. (**b**) If there is a leg length difference, then planks with various thicknesses are placed under the foot on the shorter side, until the anterior superior iliac spines are at the same level. The thickness of the planks is the measurement of the shortening. (**c**) Leg length difference in the thighs can be shown with the child lying on its back with the legs and knees flexed to 90°. If there is a difference in knee height there is a leg length difference in the thighs as in this case (the same patient as in (**a**). (**d**) By allowing the patient to lie supine with the knees flexed to 90° one can see if there is a leg length difference in the lower legs. If the soles of the feet are at the same height as in this case, there is only a difference in the length of the upper legs (the same patient as in (**a**)

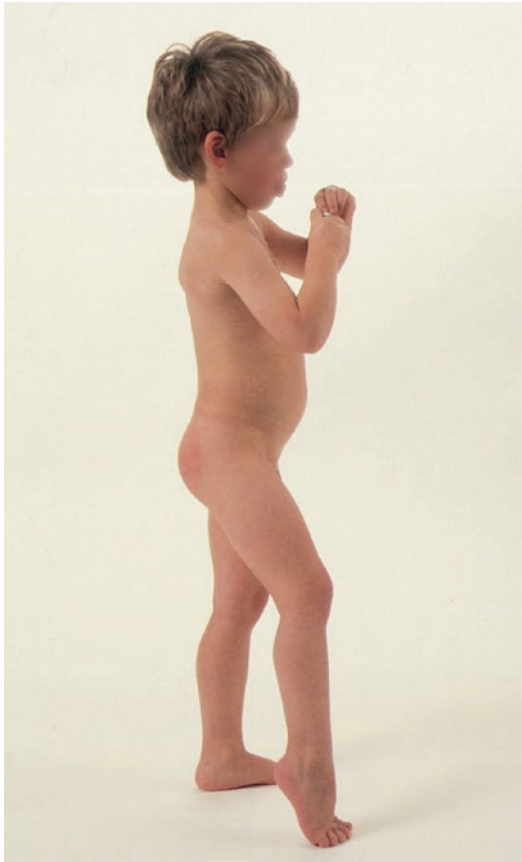


**Fig. 15.4** Right spastic hemiplegia. There will be a maximal leg length difference of 1.5 cm in the involved leg when the child has stopped growing. The leg length difference will be compensated by plantar flexion in the ankle

part of the femur. In children the chance of an ischemic necrosis of the femoral head is 75 % after a transepiphyseal fracture, a transcervical and a cervico trochanteric fracture and 15 % after an intertrochanteric fracture. Ischemic necrosis of the femoral head causes damage to the proximal femoral epiphysis causing a growth disorder. The final leg length difference is dependent on the age at which this complication occurred.

## Neurological

**Spastic hemiplegia** In this case the leg length differences are seldom more than 1.5 cm (Fig. 15.4).

**Poliomyelitis** Leg length differences caused by poliomyelitis are not often seen in better developed countries.

**Peripheral nerve or plexus injury** Hemihypotrophy can occur as a result of a nerve injury or plexus injury.

## Growth Plate Damage

**Acute osteomyelitis** In some cases an acute osteomyelitis can cause a destruction of the growth plate (see Chap. 18). This can lead to substantial growth inhibition. Sometimes the growth plate partially closes and apart from shortening also causes a progressive deformity (Fig. 15.5).

**Septic arthritis of the hip** A septic arthritis of the hip that has been treated too late can lead to destruction and ischemic necrosis of the femoral head. This can lead to a leg length difference of up to 4 or 5 cm, depending on the age of the patient at the time of the infection.

**Growth plate fracture** In 1 % of growth plate fractures damage in the growth plate causes a bony connection between the metaphysis and the epiphysis. A shortening will only occur if this bony connection is located centrally. Apart from shortening a misalignment will occur if the bony connection is asymmetrical. The leg length difference that occurs is dependent on the age of the child and the localization of the fracture. The final leg length difference will be greater the younger the child is. Growth plate closure above or under the knee will lead to a greater leg length difference than growth plate closure in the epiphysis in the proximal part of the femur or in the distal part of the tibia.

**Slipped capital femoral epiphysis** A slipped capital femoral epiphysis seldom leads to a leg length difference of 2 cm or more (see Chap. 9).

**Radiotherapy** Radiotherapy for the treatment of tumors may lead to damage to the growth plate.

## Bone Tumor

**Unicameral bone cyst, osteochondroma and enchondroma** Bone tumors can inhibit growth. Examples of growth inhibition are in solitary

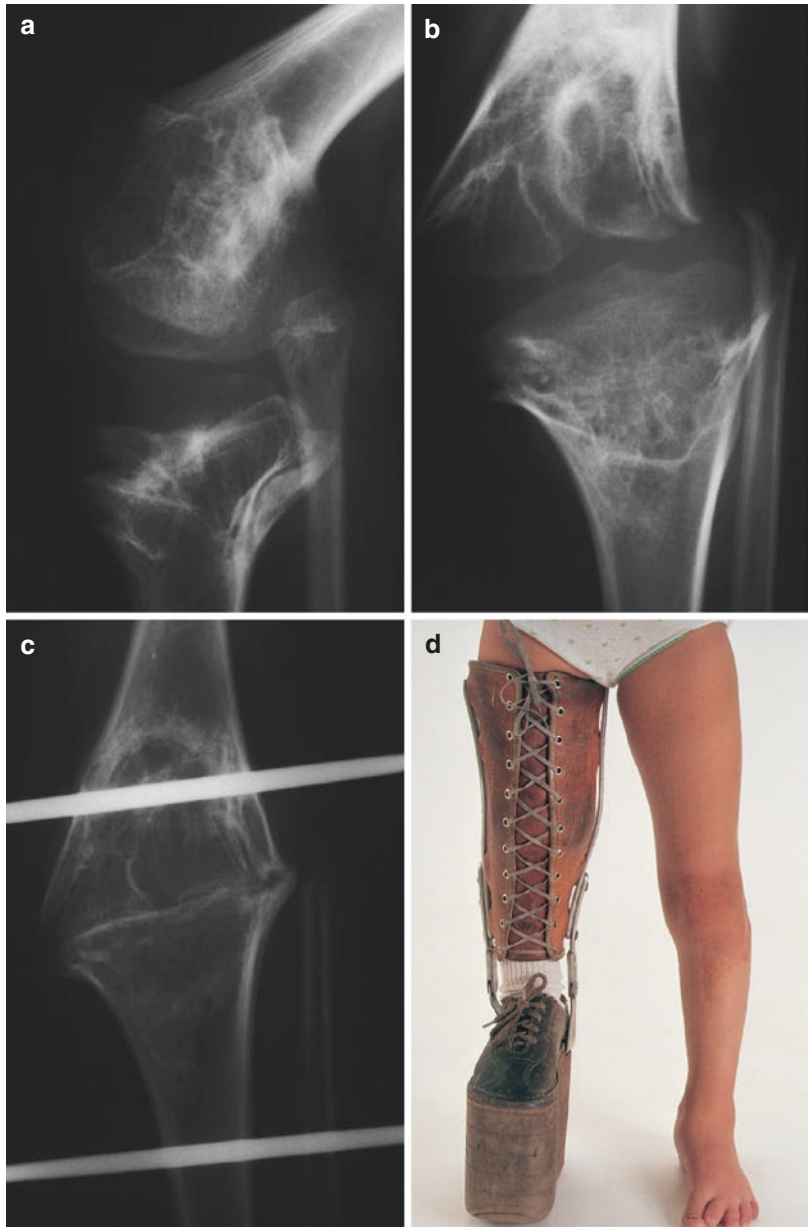



**Fig. 15.5** (**a**–**b**) Situation after a meningococcal sepsis in a newborn child with damage to diverse growth plates in the right knee with serious positional deformities and shortening of the right leg as a result. (**c**) A right knee arthrodesis was carried out because of a serious progressive deformity of the joint. (**d**) After the right knee arthrodesis an extensive leg length difference remains, which in the first instance was dealt with a sole elevation. In order to prevent ankle instability a long leg splint was mounted onto the shoe. Later on a leg lengthening procedure was carried out (see Fig. 15.15). Operation performed in the early eighties

bone cysts, osteochondromata and enchondromas. Enchondromas may be part of Ollier disease[2] (Fig. 15.6) or the Mafucci syndrome[2]. The difference between Ollier disease and the Mafucci syndrome[2] is that in the latter subcutaneous hemangiomas are present.

For supplementary assessment, primary care treatment, when to refer, secondary care treatment, if one leg is longer than the other, see too long leg, pp. 325 and 331 in this chapter.

[2]See Appendix.

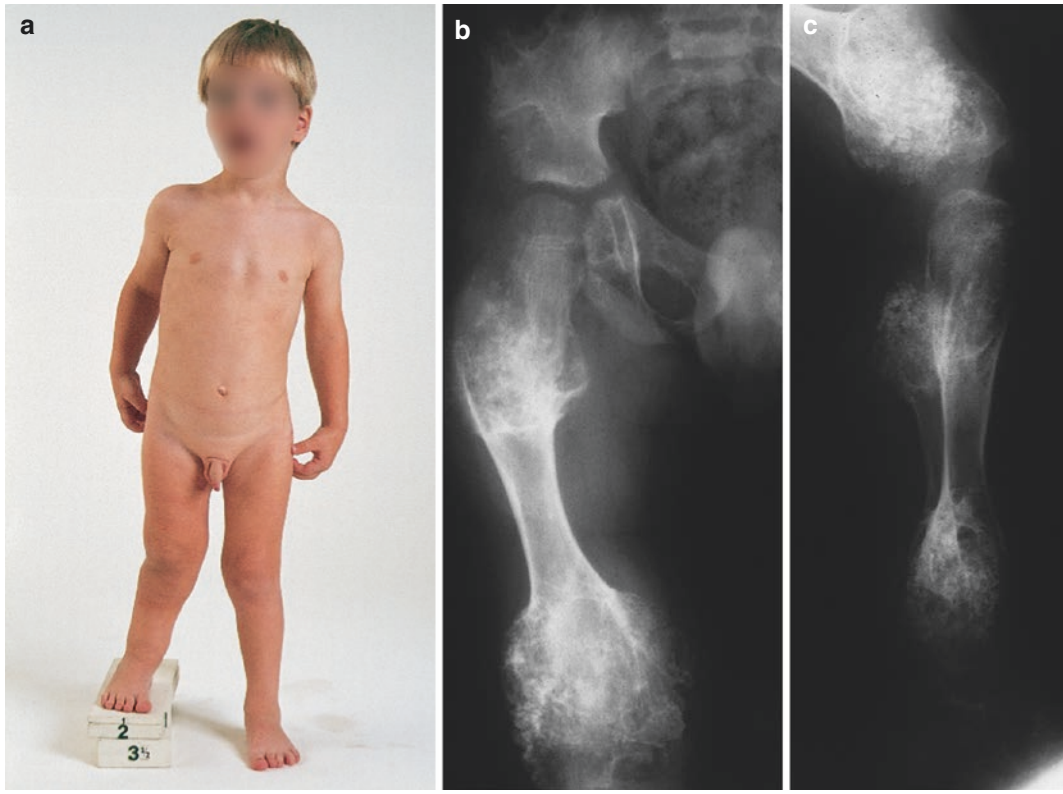



**Fig. 15.6** Ollier disease. (**a**) Positional deformity and shortening of the right leg. (**b**–**c**) Multiple enchondromas in several metaphyses of the right femur and tibia

## One Leg is Longer than the Other

Complaint: the parents feel that the child has a leg length difference or they find that the child walks abnormally.

Assessment: the leg length difference can be measured using the plank method.

Differential diagnosis:

congenital
- **idiopathic hemihypertrophy (hemihyperplasia)**
- **Beckwith-Wiedemann syndrome**
- **Proteus syndrome**

vascular
- **arteriovenous fistulas**
  - Klippel-Trenaunay-Weber syndrome

neurological
- **sympathectomy**

growth plate stimulation
- **acute osteomyelitis**
- **femoral or tibial shaft fractures**

bone tumor
- **fibrous dysplasia**
- **osteoid osteoma**
- **neurofibromatosis**
  - Von Recklinghausen disease[3]

Explanatory note: congenital

**Idiopathic hemihypertrophy** The hemihypertrophy can be idiopathic or is a part of a syndrome such as syndrome such as Beckwith-Wiedemann[3] and Proteus[3]. An idiopathic hemihypertrophy is seen in 1 in 50,000 individuals. Hemihypertrophy is clearly seen several years after birth. The overgrowth can involve a complete half of the body, including the ear, the tongue, the pupil, the nipple, the thorax, and the abdomen. One arm and/

[3] See Appendix.

**Fig. 15.7** Idiopathic hemihypertrophy of the right leg

or leg may be longer and thicker. The difference in size ends precisely in the midline. A crossed hemihypertrophy can occur exceptionally with overgrowth of one leg and the contralateral arm. The leg length difference is seldom more than 5 cm and is usually evenly distributed in the tibia and femur. There is an increased chance of tumor development, in particular a Wilms tumor (a very malignant adrenal gland tumor in children younger than 10 years of age), hepatoblastomas and leiomyosarcomas. The chance of this happening is about 6 % (Fig. 15.7).

## Vascular

**Arteriovenous fistulas** As a result of arteriovenous fistulas in the Klippel-Trenaunay-Weber syndrome[4] there is an increase in vascularization and as a result an increase in leg length growth in the involved leg. The leg length difference is seldom more than 2 cm (Fig. 15.8).

## Neurological

**Sympathectomy** In the past a sympathectomy used to be carried out on the side of the shorter leg. The result was unpredictable and poor and is not carried out anymore.

## Growth Plate Stimulation

**Acute osteomyelitis** Growth stimulation as a result of chronic infection in a metaphysis or diaphysis as a result of increased vascularization in the bone seldom leads to such a great leg length difference that has to be operatively corrected.

**Femur and tibial shaft fractures** In children younger than 12 years of age growth stimulation occurs after a fracture or an operation of the femur or tibial shaft. The younger the child is and the greater the shortening, the greater the growth stimulation will be. Growth stimulation in the femur is maximally 1.5 cm and stops after 18 months. Growth stimulation in de tibia is maximally 1 cm and stops after 15 months. The leg is never lengthened more than 0.5–1 cm as a result of a tibial or femoral shaft fracture.

## Bone Tumor

**Fibrous dysplasia, osteoid osteoma and neurofibroma** Tumors can cause growth stimulation. Examples are seen in fibrous dysplasia, osteoid osteoma and neurofibroma in neurofibromatosis (Von Recklinghausen disease[4]).

Supplementary assessment: X-rays in the first instance are taken to try and elicit the cause of the leg length difference. An X-ray of the pelvis will be carried out if there is a possible congenital hip deformity.

The simplest method to determine leg length difference on X-ray is with the help of a teleroentgenography. In that case a complete X-ray of both legs is taken, including the ankle. An alternative is an orthoroentgenography in which the X-rays of the hip, knees and ankles are put onto one sheet. Using these

[4]See Appendix.

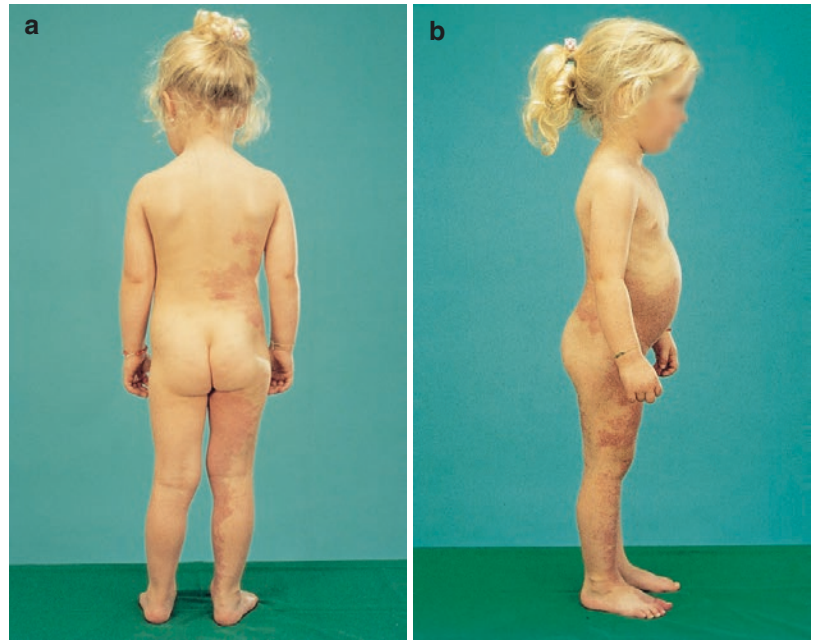



**Fig. 15.8** (**a**–**b**) Right Klippel-Trenaunay-Weber syndrome (See Appendix)

methods the possible fault in measurement is about 3 mm. A disadvantage is that a possible leg length difference caused by the foot, such as in a clubfoot, cannot be measured.

Primary care treatment: if required a leg length difference of 1–1.5 cm can be corrected with an inlay sole with the same thickness. Greater leg length differences must be corrected with a heel sole block lift.

When to refer: referal to an orthopaedic surgeon should be made if the leg length difference is 2 cm or more. Children with hemihypertrophy must also be referred to a pediatrician to check for tumors.

Secondary care treatment: there are a number of methods available to determine the final leg length difference.

## Timing of Correction

**The simple method** This is a method in which a rough estimate can be made assuming that growth inhibition is constant. At the age of 2 the child has about half of the final length as in adulthood. As a result of disproportional growth between the head and torso and the lower extremities in girls aged about 3 and boys aged about 4, half of the normal adult leg length is achieved (see Table 15.1). As an example in a boy aged 4 with a congenital deficiency with a leg length difference of about 3 cm, the leg length difference in adulthood will be about 6 cm.

**The arithmetical method** This is based on the average chronological speed of growth with age. The distal femoral growth plate provides 10 mm in the total length during the last 4 years of growth. For the proximal tibial growth plate in the proximal part of the tibia this is 6 mm, the proximal femoral growth plate 4 mm and in the distal tibial growth plate 4 mm (Fig. 15.9). Growth plates around the knee in girls close with an average age of 14 and boys with an average age of 16.5. This method can be used for long term planning.

**Growth remaining charts for distal femur and proximal tibia for girls and boys** This is determined on the basis of rest growth graphics from Anderson and Green (1963) for boys and for girls.

**Table 15.1** Multiplication factor for the lower extremity

| Multiplication factor | | | Multiplication formula | | |
|---|---|---|---|---|---|
| Age | | | Age | | |
| Year and months | Boys | Girls | Year and months | Boys | Girls |
| **Birth** | **5.080** | **4.630** | | | |
| 0+3 | 4.550 | 4.155 | 7+6 | 1.520 | 1.370 |
| 0+6 | 4.050 | 3.725 | 8+0 | 1.470 | 1.330 |
| 0+9 | 3.600 | 3.300 | 8+6 | 1.420 | 1.290 |
| 1+0 | 3.240 | 2.970 | 9+0 | 1.380 | 1.260 |
| 1+3 | 2.975 | 2.750 | 9+6 | 1.340 | 1.220 |
| 1+6 | 2.825 | 2.600 | 10+0 | 1.310 | 1.190 |
| 1+9 | 2.700 | 2.490 | 10+6 | 1.280 | 1.160 |
| 2+0 | 2.590 | 2.390 | 11+0 | 1.240 | 1.130 |
| 2+3 | 2.480 | 2.295 | 11+6 | 1.220 | 1.100 |
| 2+6 | 2.385 | 2.200 | 12+0 | 1.180 | 1.070 |
| 2+9 | 2.300 | 2.125 | 12+6 | 1.160 | 1.050 |
| 3+0 | 2.230 | 2.050 | 13+0 | 1.130 | 1.030 |
| 3+6 | 2.110 | 1.925 | 13+6 | 1.100 | 1.010 |
| 4+0 | 2.000 | 1.830 | 14+0 | 1.080 | 1.000 |
| 4+6 | 1.890 | 1.740 | 14+6 | 1.060 | |
| 5+0 | 1.820 | 1.660 | 15+0 | 1.040 | |
| 5+6 | 1.740 | 1.580 | 15+6 | 1.020 | |
| 6+0 | 1.670 | 1.510 | 16+0 | 1.010 | |
| 6+6 | 1.620 | 1.460 | 16+6 | 1.010 | |
| 7+0 | 1.570 | 1.430 | 17+0 | 1.000 | |

The multiplication factor is valid for idiopathic hemihypoand hemihypertrophy of the femur, tibia as well as the complete leg. Determination of the final leg length difference: the multiplication factor x leg length difference
From: Paley D, Bhave A, Herzenberg J, Bogen JR. Multiplier method for predicting limb – length discrepancy. *J Bone Joint Surg Am*. 2000;82A:1432–46

**The Rotterdam straight line graph** This is derived from the "Moseley straight line graph". Hereby the skeletal age is determined using the Greulich and Pyle atlas (Fig. 3.19). The final leg length difference can be determined after carrying out a number of measurements during a period of time, for example, three times with periods of 6 months in between.

**Paley multiplication formula** This method is applied in idiopathic hemihypo- and hemihypertrophy, in which the leg length difference proportionately increases during growth. The final leg length difference is determined by multiplying the leg length difference by the age dependant factor (Table 15.1). Example: a girl of 6 years of age has a leg length difference of 2 cm in the thigh. In adulthood the leg length difference will be 1.51 × 2 cm = 3.02 cm. This is also dependant on the contribution of the distal femoral growth plate after 5 years of age which is 10 mm each year (Fig. 15.9). In this case the leg length difference has to be corrected with an epiphysiodesis of the growth plate of the distal part of the contralateral femur at the age of 11.

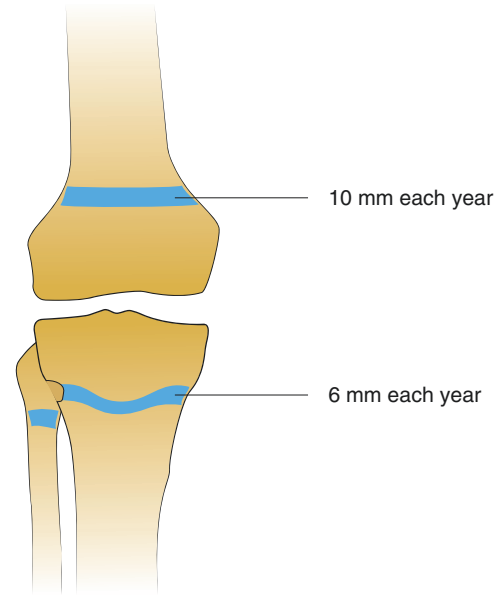



**Fig. 15.9** The growth plate contribution to length in the distal part of the femur is 10 mm during the last 4 years of growth. In the proximal part of the growth plate in the tibia this is 6 mm each year

In order to improve the accuracy of this method one has to also look at the parents. If the parents are tall instead of 10 mm per year growth in the distal femur this may be 11 mm. If the parents are small then instead of 10 mm one can accept 9 mm per year. This is also the case for the tibia, in tall parents instead of 6 mm it can be 7 mm per year. In small parents instead of 6 mm one can expect 5 mm growth per year. Shortly before an epiphysiodesis is to be carried out it is wise to check that the skeletal age is not severely out of step with regard to the calendar age using the atlas of Greulich and Pyle (Fig. 3.19). Hereby

one must realize that the information in this atlas has been collected between 1931 and 1942 and is based on a thousand white children of North European origin from above average backgrounds. However, recent studies have shown that this atlas is still a reliable instrument. The growth potential is increased in an equal skeletal maturity rate. The question remains if this system is also valid for groups of non north european origin. Determining the skeletal age according to Greulich and Pyle atlas is a quick but rather crude method. The consecutive stages generally lie 1 year apart from each other. There must be an enormous difference in skeletal age before this is obvious when using the atlas.

**A too short and too long leg** Generally a leg length difference of 1–2 cm will not be corrected. If one does wish correction then this can be done with an insole of 1–1.5 cm.

The foot will not fit into the shoe if the insole is thicker. A leg length difference of more than 1.5 cm can be compensated by applying a heel sole elevation. In the case of a leg length difference between 2 and 5 cm growth inhibition of the relatively long leg will be proposed if this problem has been identified early enough. The epiphysiodesis will be carried out in the distal femoral growth plate and/or proximal tibial growth plate, depending on the area of difference. If the expected growth from the proximal tibial growth plate is more than 1 cm then one must do an epiphysiodesis in the proximal fibular growth plate, otherwise there will be an overgrowth in the fibula head. A few decades ago the open Phemister technique was used in which a rectangular piece of bone was removed medial and lateral at the level of the growth plate, two thirds metaphysial and one third epiphyseal. After removal of the rest of the lateral part of the growth plate this block of bone was rotated and placed into the defect. Since the beginning of the nineties the medial and lateral areas in the epiphysis were percutaneously drilled out and destroyed with a sharp spoon or curette through two small medial and lateral incisions under X-ray screening (Fig. 15.10). Possible complications as a result of insufficient destruction of the growth plate are that the leg continues to grow or grows crooked because one or both sides have been insufficiently destroyed. On the same day or the day after the children can be mobilized whereby they can completely bear weight on the operated leg.

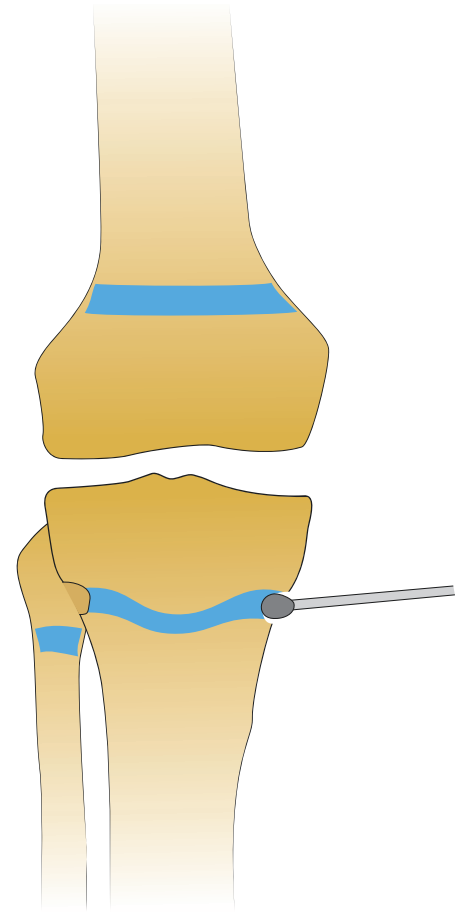


**Fig. 15.10** Epiphysiodesis of the proximal part of the tibia. The epiphysis is destroyed on the medial and lateral side

A shortening osteotomy of the femur or tibia and fibula can be proposed in children who have stopped growing. The femur can be shortened by 2–6 cm and the tibia 2–3 cm. In the femur one has the choice of inter- or subtrochanteric shortening after which the osteotomy is fixed with a hook plate (Fig. 15.11). One can also

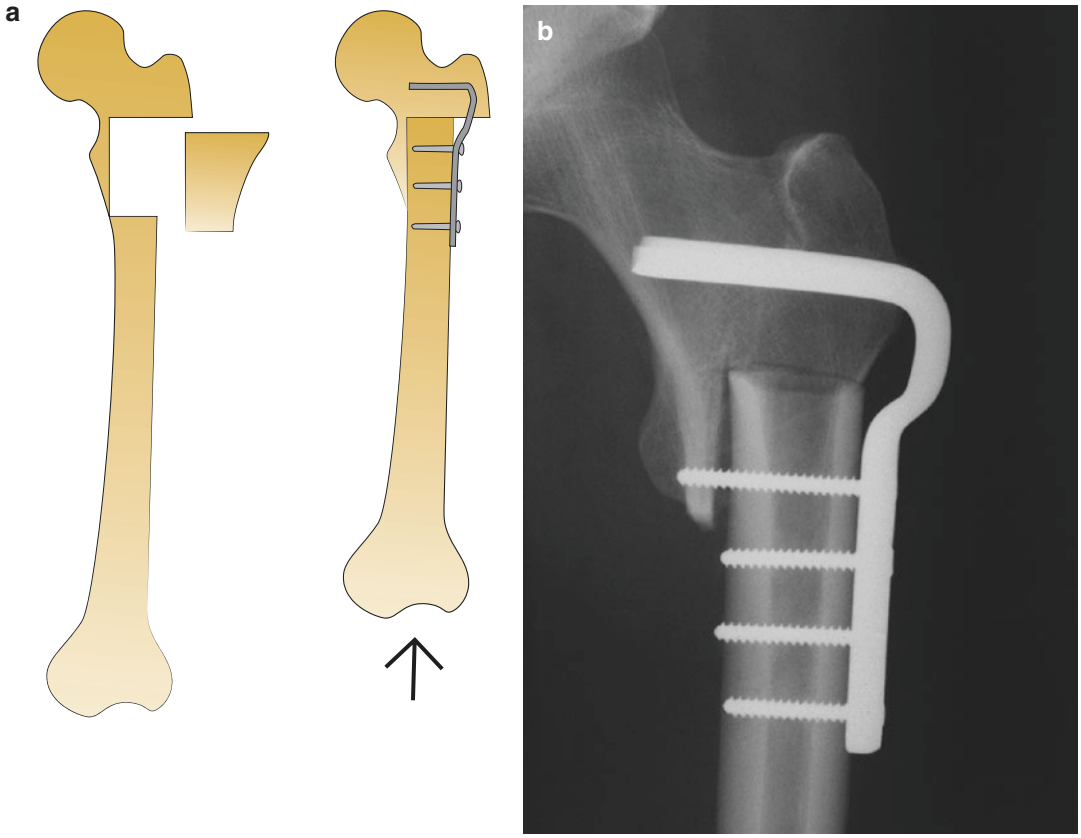



**Fig. 15.11** (**a**) Shortening of the upper leg can be carried out by removing a piece of bone just under the hip joints with fixation of the bone using a hook plate and screws. (**b**) Postoperative X-ray

choose closed intramedullary shortening as in the Winquist method (Fig. 15.12). Hereby a fragment of bone of the required length is removed from the femoral shaft with an intramedullary saw. After the osteotomy the ends have been approximated and the femur is fixed with an intramedullary nail. It is also possible to do an open procedure in which the femoral shaft is shortened and fixed with an intramedullary nail. An operation to lengthen the leg will be advised with a leg length difference of more than 5 cm (Fig. 15.13). A leg length difference of less than 5 cm can be an indication for lengthening in children who are expected to remain small.

Combinations of the above mentioned techniques are also possible.

In cases in which the leg length difference is more than 15 cm one may consider a Syme amputation and a prosthesis (see Chap. 17).

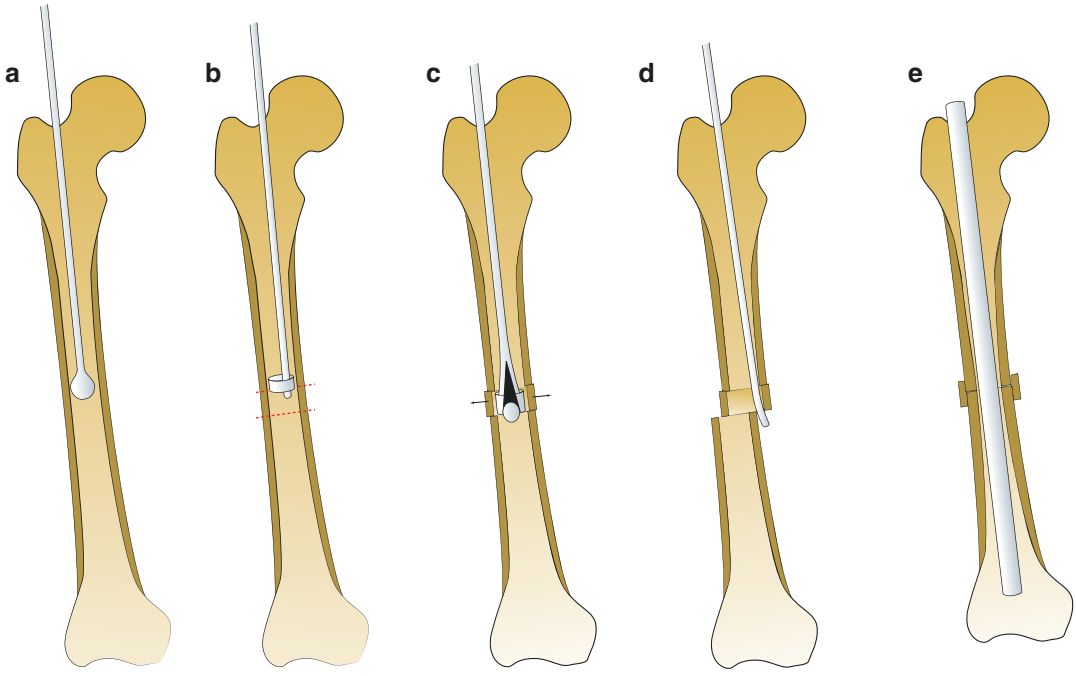



**Fig. 15.12** Closed intramedullary shortening of the femur using Winquist method. (**a**) Removal of bone marrow. (**b**): 2 saw cuts. The distance between the saw cuts is dependant on the degree of shortening required. (**c**) Fragmentation of the bone area between the saw cuts. (**d**) Displacement of the fragments. (**e**) Intramedullary fixation (Winquist RA. Closed intramedullary osteotomies of the femur. *Clin Orthop Relat Res*. 1986;212:155–64)

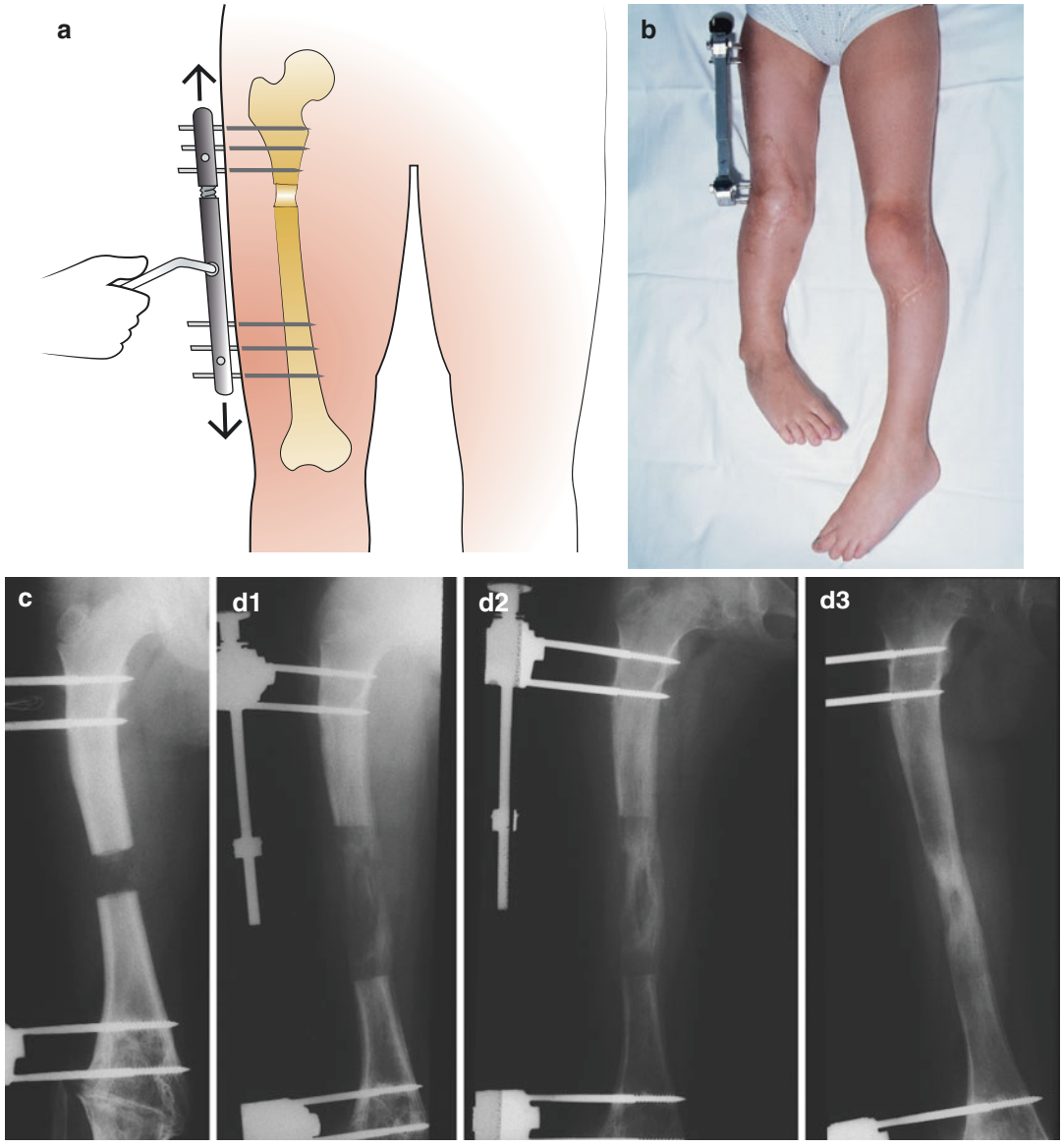



**Fig. 15.13** (**a**) Thigh lengthening, in this case with a unilateral external fixator on the lateral side of the upper leg. A femoral osteotomy is carried out before applying the external fixator. The fixator can be opened up by the patient with a frequency of 1 mm each day divided into 4 steps. New bone tissue is formed at the level of the osteotomy. The callus is pulled apart (callus distraction) until the required length is achieved. The fixator remains in position until the callus is strong enough to allow removal. Deciding the time for removal is difficult but on average one should accept 1 month for each cm of lengthening. (**b**) Lengthening of the thigh with a unilateral external fixator in the same patient as in Fig. 15.6. (**c**) 7–10 days after the osteotomy the fixator will be opened up by the patient with a frequency of 1 mm each day divided into 4 steps. (**d1**) Callus distraction until the required length is achieved. (**d**2–**d**3) The fixator remains in place until the lengthened piece of bone is strong enough to bear weight. Same patient as in Fig. 15.5. Operation performed in the eighties

**Differential Diagnosis: Leg Length Inequiality**

| Cause | One leg is shorter than the other | One leg is longer than the other |
|---|---|---|
| Congenital | **Idiopathic[a] hemihypotrophy (hemihypoplasia)** | **Idiopathic hemihypertrophy (hemihyperplasia)** |
| | **Russell-Silver syndrome** | **Beckwith-Wiedemann syndrome** |
| | **Congenital deficiency** | **Proteus syndrome** |
| | **Developmental dysplasia of the hip** | |
| | **Talipes equinovarus* (clubfoot)** | |
| Vascular | **Legg-Calvé-Perthes disease[a] (Perthes disease, coxa plana Waldenström disease)** | **Klippel-Trenaunay-Weber syndrome[a]** |
| | **Ischemic necrosis of the femoral head[a]** | |
| Neurological | **Spastic hemiplegia[a]** | **Sympathectomy[a]** |
| | **Poliomyelitis** | |
| | **Perifpheral nerve lesion[b]** | |
| | **Plexus injury[b]** | |
| Growth plate damage | **Acute osteomyelitis[b]** | |
| | **Septic arthritis of the hip Growth plate fractures[b]** | |
| | **Slipped capital femoral epiphysis[a]** | |
| | **Radiotherapy[b]** | |
| Growth plate stimulation | | **Acute osteomyelitis[a]** |
| | | **Femoral shaft fractures[a]** |
| | | **Tibial shaft fractures[a]** |
| Bone tumors | **Unicameral bone cyst (juvenile-, solitary-, simple bone cyst)** | |
| | **Osteochondroma[a] (exostosis)** | **Fibrous dysplasia** |
| | **Enchondroma[a]** | **Osteoid osteoma[a]** |
| | Ollier disease | **Neurofibromatosis** |
| | Mafucci syndrome | **(Von Recklinghausen disease)** |

[a]The diagnosis marked with an asterix seldom lead to a leg length difference of 2 cm or more
[b]Leg length difference is dependent on the age at which the damage is sustained

# Abnormal Gait

# 16

## Introduction

A child that has just started to walk has a broad aligned gait whereby the upper arms are kept in abduction in order to keep balance. The knees are not flexed and the toes reach the ground first and not the heel. At 2 years of age there is a normal gait. During the gait cycle knee flexion begins and the lateral part of the heel reaches the ground first.

In the first year of life there is an increased external rotation in the hip joints. After the age of about 2–4 years there is as a rule a symmetrical distribution between internal- and external rotation (Fig. 16.1). In the lower legs it is just the opposite. In newborns there is on average 5° of internal-up to 5° of external tibial torsion. At the age of two there is an external torsion in the lower legs of 10–20° (Fig. 16.2). The medial side of the foot should be straight (Fig. 16.2). In a normal gait the heel of the shoe wears out at most on the lateral side (Fig. 16.3).

If the child walks with the toes turned inwards because of strong internal rotation of the hips or internal tibial torsion of more than 5°, then the heel wears out at most on the medial side (Fig. 16.4).

Neurological problems causing an abnormal gait have been left out of this discussion

## Gait Disorders

### Limping

Complaint: the parents complain that the child is limping.

Assessment: there is a leg length difference of 2 cm or more.

Diagnosis: **one leg is shorter or longer than the other** (for explanatory note, supplementary assessment, primary care treatment, when to refer, secondary care treatment, see Chap. 15).

### Waddling Gait

Complaint: the child walks like a duck (sags as it were through the hips).

Assessment: the air space between the legs just under the perineum is widened (thigh gap). The Trendelenburg test is positive.

Differential diagnosis:

**coxa vara**
congenital coxa vara
acquired coxa vara
**missed bilateral hip dislocation**

Explanatory note: in the Trendelenburg test the strength of the hip abductors are tested. One asks the patient for instance to stand on the right leg and lift the left leg. The pelvis


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_16

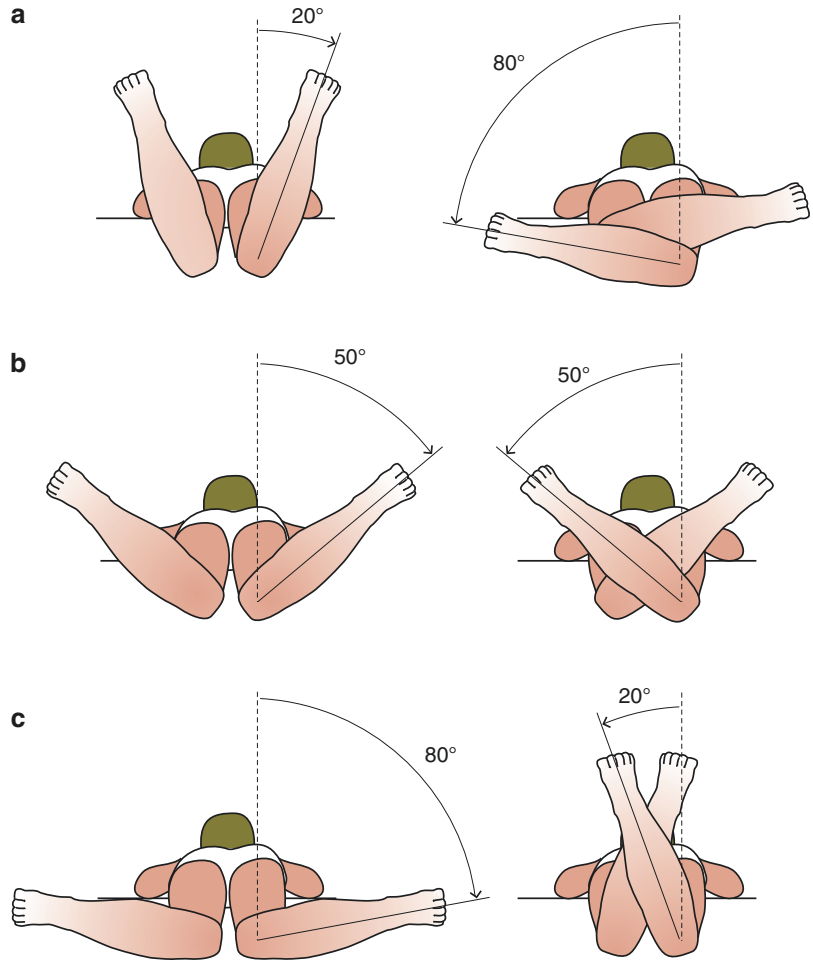



**Fig. 16.1** Rotations of the hips are measured prone with extended hips and 90° of knee flexion. Turning the lower legs outwards is internal rotation, moving the lower legs in the opposite direction is external rotation. (**a**) In newborns there is a strong external rotation in the hips of 80–90° and a diminished internal rotation of 10–20°. (**b**) From the age of 2–4 years there is as a rule a symmetrical distribution between internal- and external rotation. (**c**) In some children between the ages of 4 and 6 years there is a marked internal rotation and less external rotation. If the internal rotaton is 70° or more then the child walks with the toes turned inwards. The sum of the rotations in extended hips is usually 100°. Thus, with an internal rotation of 80° degrees there is an external rotation of 20°

on the left side will be lifted up if the hip abductors have normal strength. In that case the Trendelenburg test is negative (Fig. 16.5b). If the distance between the origin and insertion of the hip abductors is shortened as in coxa vara and a hip dislocation, for instance on the left side, then the hip abductors are relatively too long and therefore less powerful. The Trendelenburg test is positive if the pelvis sags down on the right side while the child stands on the left leg and tries to lift the pelvis on that side (Fig. 16.5c).

In a congenital or acquired coxa vara the angle between the femoral head/neck and the femoral shaft is less, in which case the greater trochanter lies higher (more proximal) (Fig. 16.6). If the greater trochanter is at the same level as the upper edge of the femoral head as in coxa vara there is in 50 % of cases a positive Trendelenburg test. If

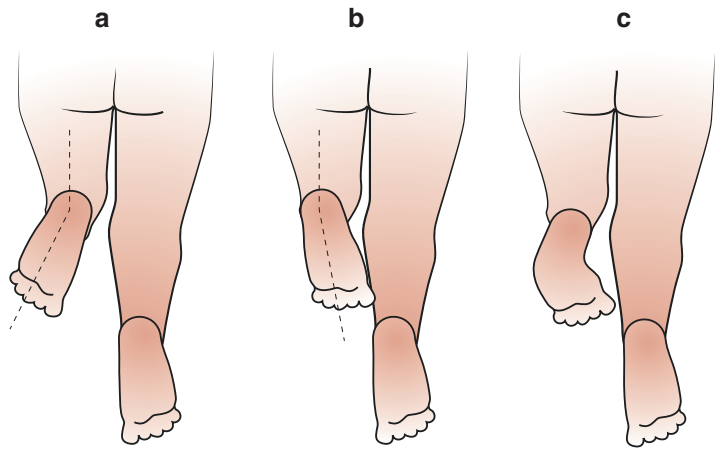



**Fig. 16.2** Assessment of torsion in the lower leg with a child lying prone with the left knee in 90° of flexion. (**a**) The foot is turned outwards 10–20° with regard to the upper leg (external tibial torsion). (**b**) In internal tibial torsion the foot is turned inwards with regard to the upper leg. (**c**) Inspection of the medial foot edge with the child lying prone and the left knee in 90° of flexion. The medial foot edge should be straight. In a metatarsus adductus and a metatarsus varus the mid- and fore-foot is angled inwards with regard to the hindfoot

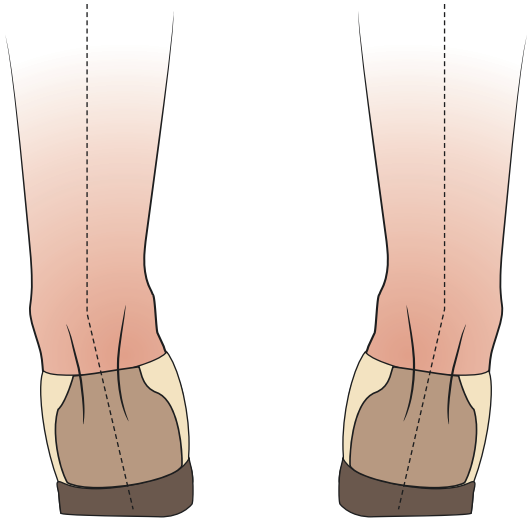


**Fig. 16.3** With a normal gait the heel of the shoe wears out on the lateral side

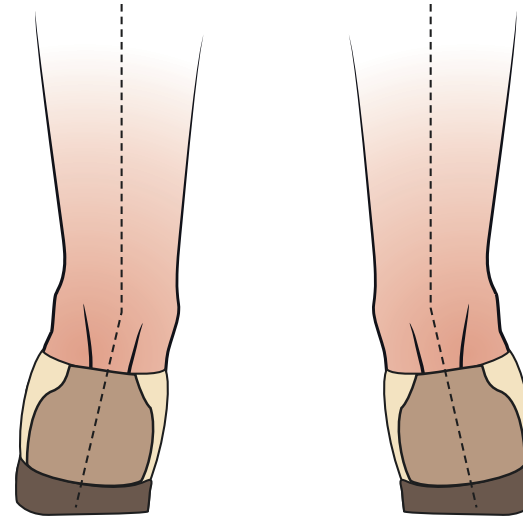
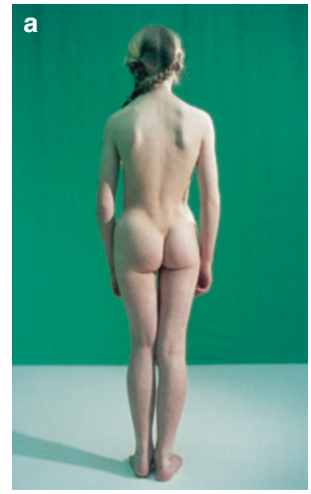


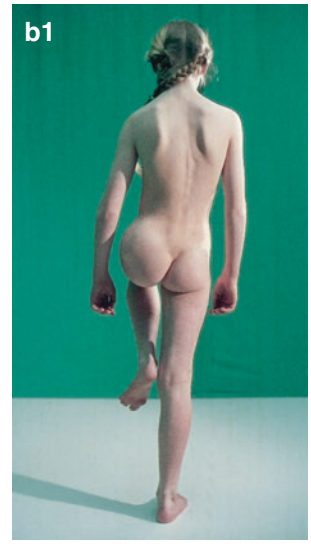
**Fig. 16.4** If the child walks with intoeing then the heel will wear out on the medial side

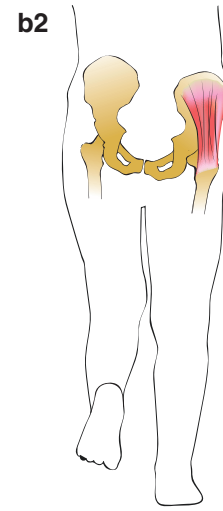
the proximal part of the greater trochanter lies more proximal than the upper edge of the femoral head, then there is 50–100 % chance of a positive Trendelenburg test.

*Congenital coxa vara* This type is rare and is present at birth. It is often associated with other inborn errors such as congenital short femur (femur hypoplasia), proximal focal femoral deficiency and a dysostosis cleidocranialis. This type is often recognized earlier due to the other deformities.

*Acquired coxa vara* This type of hip deformity is as a rule finally diagnosed between the ages of 3 and 5. As a child gets older and heavier, the





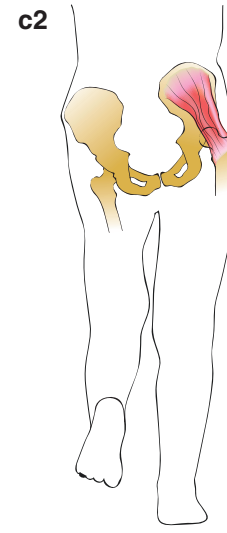



**Fig. 16.5** Trendelenburg test. (**a**) Patient with a left hip dislocation stands with both feet on the ground. (**b**1–**b**2) One asks the patient to lift the left leg. The left pelvis half is lifted up if there is normal strength in the abductors of the right hip. The Trendelenburg test is negative. (**c**1–**c**2) In a dislocation of the left hip in which the distance between the origin and insertion of the abductors is shortened there is loss of strength in these left hip muscles and the right side of the pelvis cannot be lifted up. The Trendelenburg test is positive

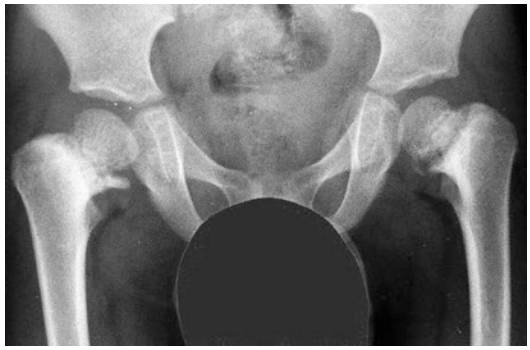


**Fig. 16.6** Anteroposterior X-ray of the pelvis: bilateral coxa vara. In a coxa vara the angle between the femoral head/neck and the femoral shaft is lessened, whereby the greater trochanter is situated more proximal than normal

epiphysis glides slowly downwards and the angle between the femoral shaft and femoral neck, the so-called caput collum diaphyseal angle (CCD-angle), decreases whereby the greater trochanter finally lies proximal to the femoral head.

**Missed bilateral hip dislocation** Apart from a widened air space between the legs just below the perineum (thigh gap), the Trendelenburg test for both hips is always positive. As a result of flexion contractures in the hips there is a compensatory hollow back (hyperlordosis), the pelvis is turned over anteriorly (Fig. 16.7) and there will be a waddling gait (Fig. 16.8). For further explanation, see Chap. 9.

Supplementary assessment: anteroposterior X-rays of the pelvis.

Primary care treatment: none.

When to refer: if the X-ray is abnormal.

Secondary care treatment: see Chap. 9.

## Toe Walking

Complaint: the child walks on the toes.

Assessment: apart from a bilateral limited dorsiflexion in the ankle joints no other orthopaedic or neurological anomalies will be found. As a rule the ankle can just be brought into a neutral position.

**Fig. 16.7** Hyperlordosis of the pelvis which is tilted anteriorly in a bilateral hip dislocation

Diagnosis: **habitual toe walker (idiopathic toe walking, congenital short achilles tendons**).

For explanatory note, suplementary assessment, primary care treatment, when to refer and secondary care treatment, see pp. 276–279.

## Out-Toeing

Complaint: from the beginning the child walks like Charlie Chaplin, with the toes turned outwards.

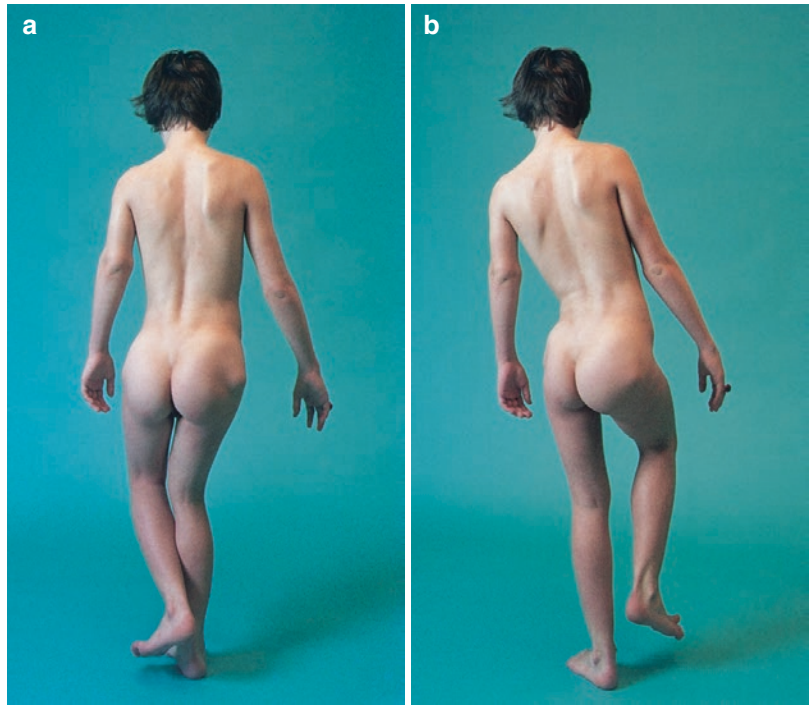



**Fig. 16.8** (**a**, **b**) There is a waddling gait as a result of less powerfull abductors in a bilateral hip dislocation

Assessment: the child walks with an out-toeing gait. The rotations in the hips are measured lying prone with extended hips and 90° of knee flexion. Turning the legs outwards is external rotation and inwards internal rotation. Examination of the torsion in the lower legs is carried out in the same position.

Differential diagnosis:

**increased external rotation of the hips**
**increased external tibial torsion**

Explanatory note: **increased external rotation of the hips**. The hips lie in external rotation in the uterus. The feet turn outwards (out-toeing) when the child stands. This is caused by an increased external rotation of 80–90° and a lessened internal rotation of 10–20° in the hips. As a rule this out-toeing normalizes in the second year of life. Sometimes this process takes more time (Fig. 16.9).

**Increased external tibial torsion** At the age of 2 years there is external tibial torsion of 10–20°. A greater increase in external tibial torsion seldom occurs. This is seen for example in congenital patella dislocation (see pp. 192 and 193) but the cause is usually unknown.

Supplementary assessment: none.

Primary care treatment: none.

When to refer: an increased external rotation in the hips is as a rule normalized in the second year of life. In cases of increased external tibial torsion referral is necessary after this age.

Secondary care treatment: **increased external tibial torsion**. In a few cases supramalleolar internal rotational osteotomies will be considered.

## Intoeing

Complaint: the child walks with the toes turned inwards and often stumbles.

Assessment: the cause of this condition can be located at three levels. There may be an adduction of the forefoot with regard to the

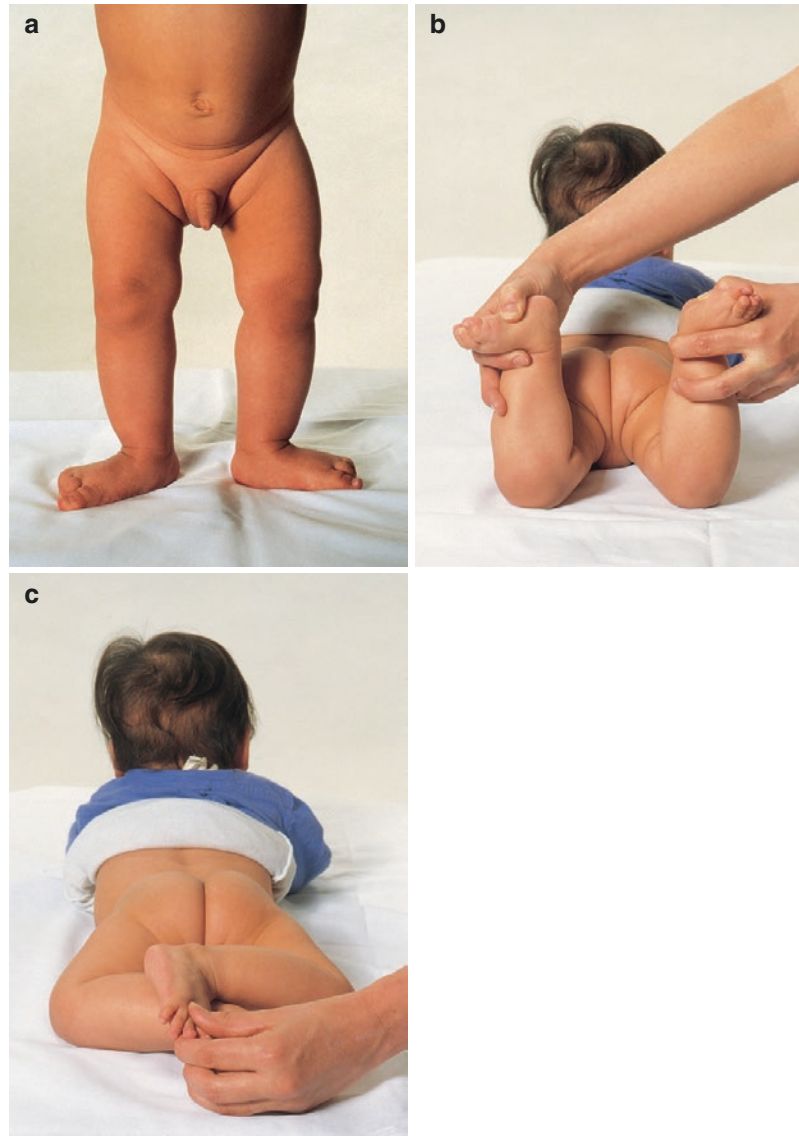



**Fig. 16.9** (**a**) In the first year of life children's feet turn outwards (out-toeing). (**b**) The same patient as in (**a**). There is a reduced internal rotation of 10° bilaterally. (**c**) The same patient as in (**a**). There is an increased bilateral external rotation of 90°

hindfoot, an internal tibial torsion and/or an increased internal rotation in the hip joint. The child lies prone with the knees in 90° of flexion. In this position the position of the forefoot with regard to the hindfoot can be observed and the internal tibial torsion can be assessed by moving the lower leg inwards or outwards.

Differential diagnosis:

**metatarsus adductus and metatarsus varus**
**internal tibial torsion >5°**
**increased internal rotation of the hips >70°**

Explanatory note: **metatarsus adductus and metatarsus varus**. The forefoot is turned inwards with respect to the hindfoot. This is present from birth but reaches a peak when the children stand because the forefoot deformity becomes more obvious when weight bearing. On examination when standing the toes turn inwards, the patellae face anteriorly. The child is examined lying prone with the knees in 90° of flexion, in which situation the mid- and forefoot turn inwards with respect to the hindfoot. The medial foot border should be in a straight

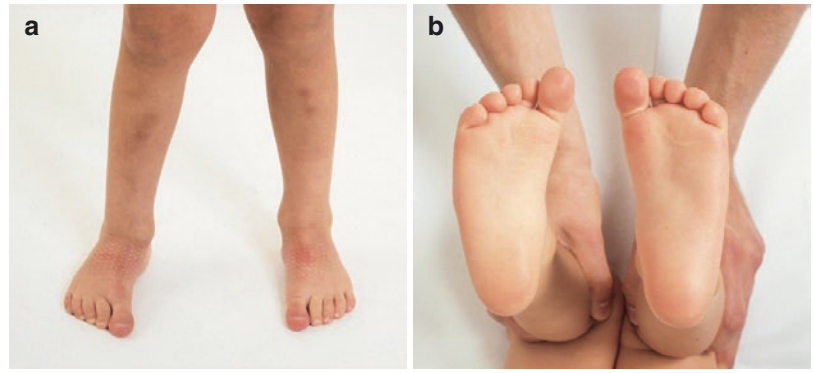



**Fig. 16.10** (**a**) Right metatarsus adductus. The right forefoot is in adduction. (**b**) The same child as in (**a**). The child lies prone with the knees flexed to 90°. The medial foot border of the left foot has a straight line. The right mid- and forefoot is turned inwards with regard to the hindfoot

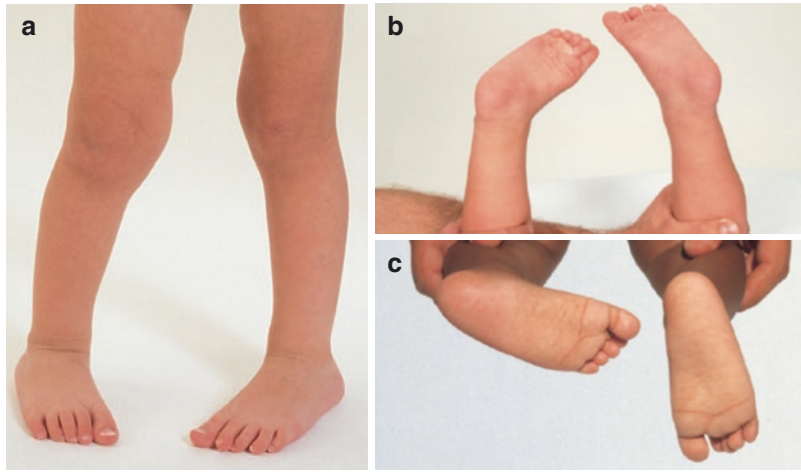



**Fig. 16.11** (**a**) Internal tibial torsion. The patellae are facing forwards and the toes are turned inwards. (**b**) The same patient as in. (**a**). The child lies prone with the knees flexed to 90°. Both lower legs have a severe internal tibial torsion. The child will walk with the toes turned inwards if there is 5° or more internal tibial torsion. (**c**) The left lower leg has an internal tibial torsion of 70° and the right leg has an external tibial torsion of 10°

line (Fig. 16.10). There is furthermore a supination component in a metatarsus varus (Fig. 13.3) (for explanatory note, see pp. 261 and 262).

**Internal tibial torsion** The legs turn inwards. The patellae face anteriorly and the toes are inwardly directed. An internal tibial torsion is the most important cause of intoeing and is usually seen at the age of 2–4 years. The child is examined prone with the knees flexed to 90°. In newborns the foot faces inwards or slightly outwards with respect to the thigh. As the children grow older the foot lies 10–20° outwards with respect to the thigh. If the foot has more than 5° of internal tibial torsion the child will walk with an intoeing gait (Fig. 16.11). The examiner should not passively turn the foot inwardly or outwardly. This can lead to a wrong interpretation. One may only stabilize the foot with one finger. In most cases the internal tibial torsion will mostly spontaneously be corrected before the fourth year of age. An internal tibial torsion can be asymmetrical with a preference for the left side in contrast to an increased internal rotation in the hips which is almost always symmetrical (Fig. 16.11).

**Increased internal rotation in the hips** Not only do the toes turn inwards but also the patellae (Fig. 16.12). It often looks as if the child has knock knees. They disappear after the child has been asked to rotate the toes straight forward. In

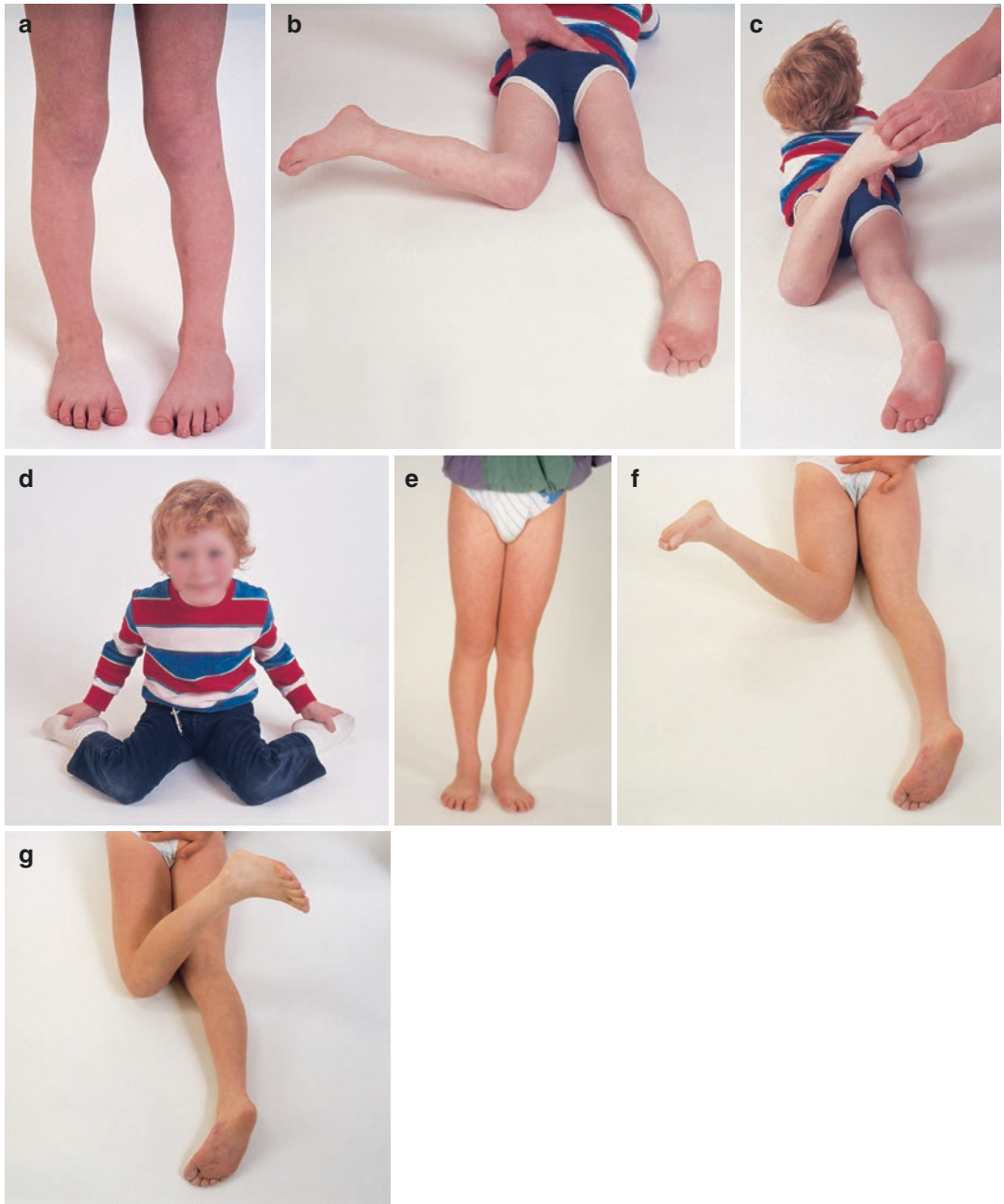



**Fig. 16.12** (**a**) A 5 year old boy. Inwardly directed patellae and feet (intoeing). (**b**) The same patient as in (**a**). There is an increased internal rotation of 80° with extended hips and 90° flexion in the knee. The examiner stabilizes the pelvis with one hand when rotations in one leg are being measured. (**c**) The same patient as in A with an external rotation of 20° in an extended hip with a 90° of flexion in the knee. (**d**) W-position in the same patient as in (**a**). (**e**) The same patient as in (**a**), now 12 years of age. The patellae and also the feet are directed straight forward. (**f**) The same patient as in (**a**) now 12 years of age. The internal rotation has spontaneously been corrected from 80 to 40° in the course of 7 years. (**g**) The same patient as in (**a**), now 12 years of age. The external rotation has spontaneously been corrected from 20 to 40° in 7 years

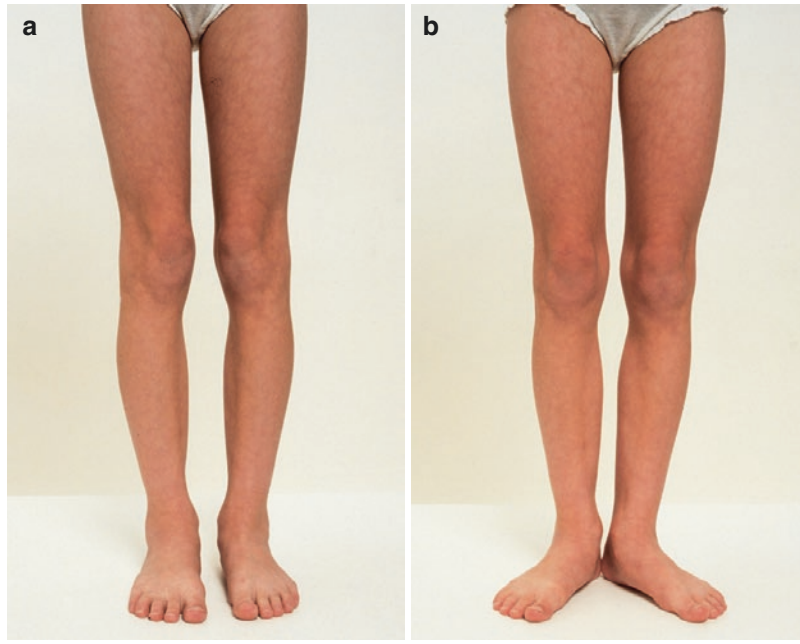



**Fig. 16.13** (**a**) Increased internal rotation of the hips which leads to infacing patellae, compensated by an increased external tibial torsion. (**b**) The same patient as in (**a**). If the patellae face directly forwards then the lower legs are in increased external torsion

an examination with a child lying down, the child lies prone with the knees in 90° of flexion. Turning the lower leg outwards gives a degree of internal rotation and rotating inwardly a degree of external rotation. The examiner fixes the pelvis with one hand. As a rule there is a symmetrical distribution between internal and external rotation in children 4 years or older. An increased internal rotation and a lessened external rotation occurs in some children between 4 and 6 years of age (Fig. 16.12). The sum of the rotations in extended hips is usually 100° between 4 and 6 years of age. An exception to this rule is in children with hypermobility. The sum of the rotations in these cases will be more than 100°. The child will walk with the toes turned inwards if the internal rotation is 70° or more. These children prefer to sit in their favorite position which is the so-called W-position and sleep preferably on their abdomens. The increased internal rotation and diminished external rotation will spontaneously correct in 80 % before they reach the age of 12 (Fig. 16.12). Hip rotations will decrease in the course of time. At the age of 12 years maximal internal and external rotation of 40° is usually normal. In adults the internal rotation as well as external rotation is normally 30°.

In cases in which the increased internal rotation does not correct spontaneously there is usually an increased external tibial torsion. Hereby one sees inwardly turned patellae and an increased external torsion (more than 20°) in the lower legs whereby the toes normally face forward. If the patellae face directly forwards, then the lower legs are in increased external torsion (Fig. 16.13). In some cases the increased internal rotation of the hip is compensated by a subluxation of the talonavicular joint. The patellae again face inwardly. Compensation occurs by abduction in the mid- and forefoot and a flatfoot develops as a result (Fig. 16.14).

Supplementary assessment: in principal not necessary.

Primary care treatment: the metatarsus adductus can usually be passively corrected by the examiner. In 90 % of cases the mid- and forefoot can generally be brought into abduction with respect to the hindfoot. If this is the case, the metatarsus adductus will spontaneously correct itself and the patient does not need to be referred.

When to refer: metatarsus adductus. Referral is indicated when the forefoot with respect to

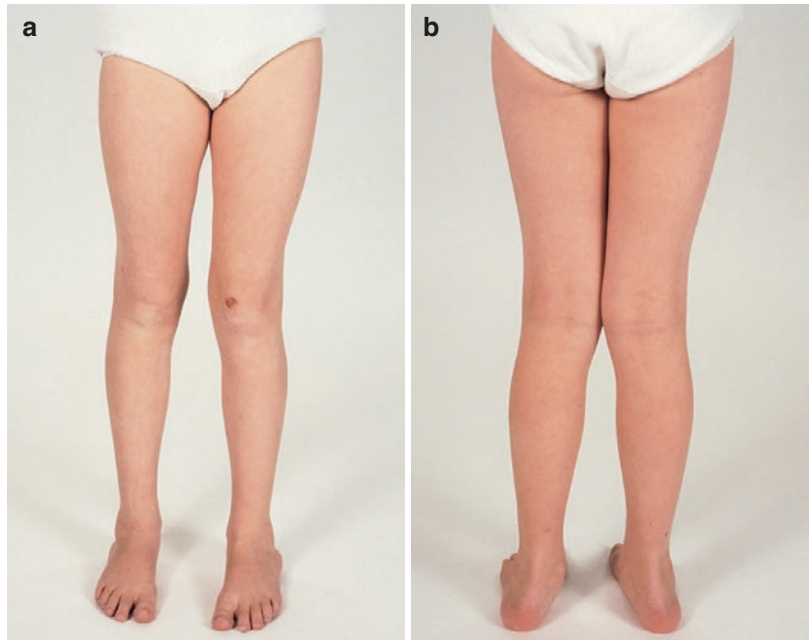



**Fig. 16.14** Increased internal rotation in the hips which causes infacing of the patellae compensated by flatfeet. (**b**) The same patient as in (**a**), looking from behind, where- by the valgus positon of the hindfeet can be clearly seen

the hindfoot can just or just not be brought into neutral. In these cases spontaneous correction will not happen (rigid metatarsus adductus). Metatarsus varus. A metatarsus varus will never be spontaneously corrected and must be referred.

Internal torsion of the lower legs. An internal torsion of the lower legs will in almost all cases be spontaneously corrected before the 4th year of life. A referral will be made only in extreme cases in children older than 4 years of age with an internal tibial torsion.

Increased internal rotation in the hips. In 80 % of cases a spontaneous correction occurs. Only in those cases where there is an internal rotation contracture are eligible for operative treatment after 12 years of age.

Secondary care treatment: **metatarsus adductus and metatarsus varus**. As a rule conservative treatment is carried out with serial manual corrections and plaster immobilization and after treatment for 1 or 2 years with an ankle-foot orthosis during the night. In rare cases operative correction (see also p. 267).

**Fig. 16.15** Contrarotational system. The shoes can be adjusted with respect to the frame. In an internal rotation of the lower legs the shoes will be turned outwards 20° in a few steps

**Internal tibial torsion** In extreme cases and in children older than 4 years of age, who frequently stumble, treatment with a contrarotational system during 1 year at night can be prescribed (Fig. 16.15). This treatment has not been proved scientifically but parents have in any event a feeling that something has been done.

**Increased internal rotation of the hips** Frequently the children will be stimulated to sit crosslegged and to sleep on their backs instead of their abdomens. It has not been shown however, that these

measures actually correct the anomalous rotation. The child sits or sleeps like that because the increased internal rotation in the hips is present and not the reverse. It has also never been shown that the natural course will be influenced by wearing external rotational systems.

Parents often have cosmetic objections and persist in their wish for referral to an orthopedic surgeon. A possible treatment to correct the position of the legs is an external rotational osteotomy of the proximal part of the femurs after which the patellae face forwards. In the case of a secundary increased external torsion of the lower legs the child will have out-toeing after such an operation. An internal rotational osteotomy of both lower legs must take place in these cases at the same time. An extensive operative treatment is not justified in a child with a slightly poor gait and cosmetic objections from the parents. Above all, spontaneous correction will occur in 80 % of cases. Mother: "Can you make my child's knees point straight forward?" Doctor: "That is of course possible but not easy. We have to saw through the thigh bones just under the hips and then turn these outwards. Next, the ends of the bones have to be fixed to each other with a plate and screws. This frequently gives ugly scars at the level of the hips. After the operation the children can not walk for 6 weeks because the bone has to grow together. When they start walking again the knees point straight forwards but the feet point outwards just like Charlie Chaplin (Fig. 16.13) and you can understand that this looks very ugly.

(Mother nods in agreement.) But that is not a problem, after this we can saw through the lower leg bones and turn them inwards. This requires two operations and there will be ugly scars on the lower legs." Having told this story, I have never known parents who would agree to this treatment. Operative treatment is only justified in rare cases in which there is an internal rotation contracture after 12 years of age.

## Unburdening Hip Gait

Complaint: the child complains of pain in the groin, thigh and/or the knee and walks abnormally.

Assessment: when walking the upper part of the body bears weight on the painful hip in the standing phase and the internal rotation of the hip is limited and/or painful.

Differential diagnosis:

**acute transient synovitis of the hip** (**coxitis fugax**, **irritable hip syndrome**, **observation hip**, **toxic synovitis**, **acute transient epiphysitis**)
**Legg-Calvé-Perthes disease** (**Perthes disease**, **Waldenström disease**, **coxa plana**)
**chronic slipped capital femoral epiphysis**

Explanatory note: the child will not bear weight on the involved leg in the case of a very painful hip anomaly as in a fracture or a solitary bone cyst, an acute epiphysiolysis capitis femoris or an acute phase in chronic slipped capital femoral epiphysis, an acute osteomyelitis or a septic arthritis. In less painful hip anomalies such as in acute transient synovitis of the hip, Perthes disease and chronic epiphysiolysis capitis femoris the child walks with an antalgic gait (an is a prefix meaning no or without and algos is Greek for pain). By shifting the centre of gravity to the painful hip during the standing phase the power that the abductors must exert will be lessened in order to keep the pelvis in balance and the pressure on the hip joint during the standing phase will be reduced from four times the body weight to one.[1] This is only valid for the hip joint, not for example the knee or ankle joint whereby the pressure on these joints during the standing phase is about one time the body weight.

[1] See addendum

Supplementary assessment: anteroposterior and frog-leg lateral X-ray's of the pelvis. On conventional X-rays no anomalies will be seen in the case of acute transient synovitis of the hip. In other cases the anomaly will be clearly seen, particularly on the frog-leg lateral X-ray's of the pelvis.

Primary care treatment: in a coxitis fugax the child may not bear weight for 3 or 4 days.

When to refer: in Perthes disease and in a chronic slipped capital femoral epiphysis.

Secondary care treatment: see Chap. 9.

## Gaits with a Shortened Standing Phase

Complaint: the child has pain and bears weight on the painful leg for a shorter period than the other leg.

Assessment: there is a shortened standing phase on the painful leg when walking.

Differential diagnosis:

(**stress**) **fracture**
**ligament rupture**
**sprain**

Explanatory note: a gait with a shortened standing phase is also an antalgic gait. As a rule this involves traumatic injuries such as (stress) fractures, ligamental injuries or distortions.

Supplementary assessment: take X-rays of the painful area.

Primary care treatment: in distortions of the knee, ankle and foot weight-bearing mobilization depending on the complaints.

When to refer: (stress) fractures and ligament injuries.

Secondary care treatment: depending on the nature of the injury.

**Differential Diagnosis: Abnormal Gait**

| | |
|---|---|
| **Limping** | |
| Leg length difference > 2 cm. | **One leg is shorter or longer than the other** |
| **Waddling gait** | **Coxa vara** |
| Widened thigh gap and normal lumbar lordosis . | Congenital coxa vara |
| | Acquired coxa vara |
| Widened thigh gap and lumbar hyperlordosis. | **Bilateral hip dislocation** |
| **Toe walking** | |
| Limited or loss of dorsal extension in the subtalar joint. | **Habitual toewalker**, (**idiopathic toe walking**, **congenital short achilles tendons**) |
| **Out-toeing** | |
| 1–2 years of age. | **Increased external rotation of the hips** |
| External torsion of the lower legs > 20°. | **Increased external tibial torsion** |
| **Intoeing** | |
| Newborn: 1–2 years of age. Forefoot adduction. | **Metatarsus adductus** |
| Newborn: 1–2 years of age. Forefoot adduction and supination. | **Metatarsus varus** |
| 2–4 years of age. | **Internal tibial torsion > 5°** |
| 4–6 years of age. | **Increased internal rotation in the hips > 70°** |
| **Unburding hip gait** | |
| The body is brought above the painful hip during the weight bearing standing phase. | **Acute transient synovitis of the hip** (**coxitis fugax**, **irritable hip syndrome**, **observation hip**, **toxic synovitis**, **acute transient epiphysitis**) |
| | **Legg-Calvé-Perthes disease** (**Perthes disease**, **Waldenström disease**, **coxa plana**) |
| | **Chronic slipped capital femoral epiphysis** |
| **Gait with a shortened standing phase** | |
| Shortened weight bearing standing phase | (**Stress**) **fracture**<br>**Ligament rupture**<br>**Sprain** |

# Limb and Back Reduction Deficiency Present at Birth

# 17

## Limb General

The term congenital deficiency is used for an absent extremity or part thereof.

## Classification

A classification is meant to define and distinguish different anomalies from each other as much as possible. Different classifications have been published throughout the years. The most usable classification is based on missing parts of the skeleton. In this classification a distinction is made between transverse and paraxial deficiencies. In transverse deficiencies a complete extremity is absent or part thereof. In paraxial deficiencies a part is absent in the length axes parallel and distal to the elbow or knee. Paraxial deficiencies can be divided into preaxial (radial and tibial), postaxial (ulnar and fibular) and central (cleft hand and foot) deficiencies.

The term terminal is used if all parts of the extremity distal to a particular part are missing and the term intercalary is used if a part of the extremity between the trunk and hand or foot is missing. Four groups can be distinguished (Fig. 17.1):

**Transverse terminal deficiency** In a transverse terminal deficiency the extremity is normally developed down to the level of the absent part. The transverse terminal deficiency can vary from missing distal phalanges or a complete extremity. The transverse terminal deficiencies are named after the absent part. I.e. if the foot is absent the term apodia is employed (podos is Greek for foot, apodia is absence of one or both feet). In amelia the whole extremity is absent (melos is Greek for limb). The term congenital amputation is often used for terminal transverse deficiencies. This is incorrect because it involves a constructional deformity and that which is not present cannot be amputated (amputare is Latin for circumferentially cutting off). Congenital amputations are caused by an amnion band constriction in principle in a normal extremity.

**Transverse intercalary deficiency** Hereby a whole segment is absent between the trunk on one side and the hand and foot on the other. For instance the foot is fixed to the pelvis and the rest of the leg is absent. This deficiency is further divided into a complete deficiency in which the hand and foot are fixed to the trunk, a proximal deficiency in which the upper arm or thigh is completely missing and the distal deficiency whereby the forearm or the lower leg is completely missing. The old term phocomelia is frequently used (phoke is Greek for seal and melos for limb; the direct translation is a seal limb). The anomaly whereby the proximal part of the thigh including or not including the hip is an anomaly which is referred to as a proximal focal femur deficiency, that can also be referred to as an incomplete proximal type of phocomelia.


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_17

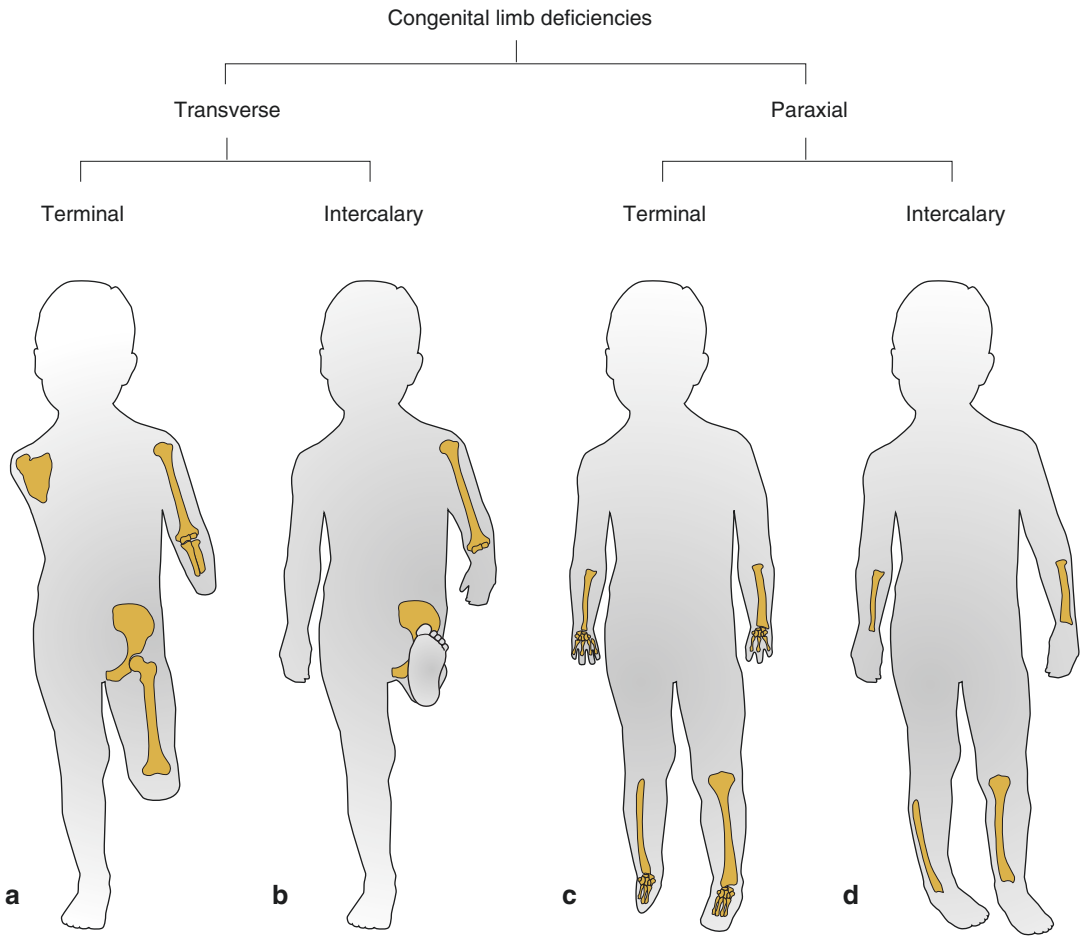



**Fig. 17.1** Frantz-O'Rahilly classification of congenital deficiencies (Redrawn from: Frantz CH, O'Rahilly R. Congenital skeletal limb deficiencies. J Bone Joint Surg Am. 1961;43A:1202–4) modified from Hall CB, Brooks MB, Dennis JF. Congenital skeletal deficiencies of extremities. Classification and fundamentals of treatment. J Am Med Assoc. 1962;181:590–9. (**a**) Transverse terminal deficiencies: amelia of the right arm, incomplete hemimelia of the left arm and hemimelia of the left leg. (**b**) Transverse intercalary deficiencies: distal phocomelia of the left arm, complete phocomelia of the left leg. (**c**) Paraxial terminal deficiencies: radius aplasia of the right arm, ulnar aplasia of the left arm, tibial aplasia of the right leg and fibular aplasia of the left leg. Absent thumb and fingers. (**d**) Paraxial intercalary deficiencies: radius aplasia of the right arm, ulnar aplasia of the left arm, tibial aplasia of the right leg and fibular aplasia of the left leg. The hand and foot are complete

**Paraxial terminal deficiency** This involves a congenital complete or partial absence of one bone or piece of bone in the forearm (radius or ulna), or lower leg (fibula or tibia) with the adjacent rays or parts of the rays of the hand and foot.

A completely missing piece of bone is called aplasia, i.e. radius aplasia (a means not present and plassein is Greek for forms). If a piece of bone is partly absent then this is called hypoplasia as in the case of radius hypoplasia (hypo is Greek for too little or too short).

**Paraxial intercalary deficiency** This involves congenitally complete or partial absence of one of the pieces of bone in the forearm (radius or ulna) or lower leg (tibia of fibula) in which the hand or foot are completely present.

**Central deficiency** This deficiency may be limited to a cleavage of hand or foot but may be so extensive missing the central three rays.

## Frequency

In 1 in 2000 births children miss an extremity or a part thereof. It involves the upper limb twice as often as the lower limb. If there are more absent parts in other limbs then one should consider the possibility of a syndrome.

*Upper extremity* In 100 children born with a congenital deficiency about 70 will miss an arm or a part thereof. In most cases it involves a terminal transverse deficiency. In 98 % of cases this is unilateral. In 2 or 3 of these 100 children the radius or ulna or part thereof is absent.

*Lower extremity* In 30 of 100 children it involves the lower extremity whereby in about 20 of the 30 cases the fibula or a part thereof is absent. In 4 of these 30 children there is a deficient proximal part of the femur whereby the femoral head may or may not be absent.

# Upper Limb

## Absence of the Whole or Part of the Arm

Complaint: the arm or a part of the arm and/or the hand is absent.

Assessment: the arm or the end of the arm is absent. The remaining part is normally developed.

Differential diagnosis (Fig. 17.2):

Transverse terminal deficiency

**Amelia**
Absence of the whole arm.

**Incomplete amelia**
Absence of the hand, forearm and a part of the upper arm.

**Hemimelia**
Absence of the hand and forearm

**Incomplete hemimelia**
Absence of the hand and part of the forearm

**Acheiria**
Absence of the hand and wrist.
(a is Greek for none, cheir is hand)

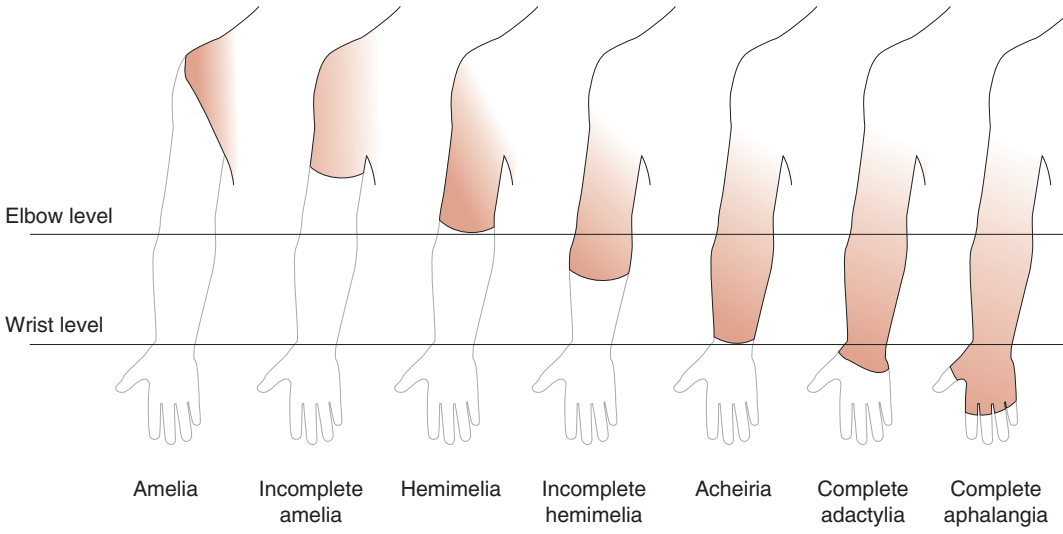



**Fig. 17.2** Classification terminal transverse deficiencies of the upper extremity

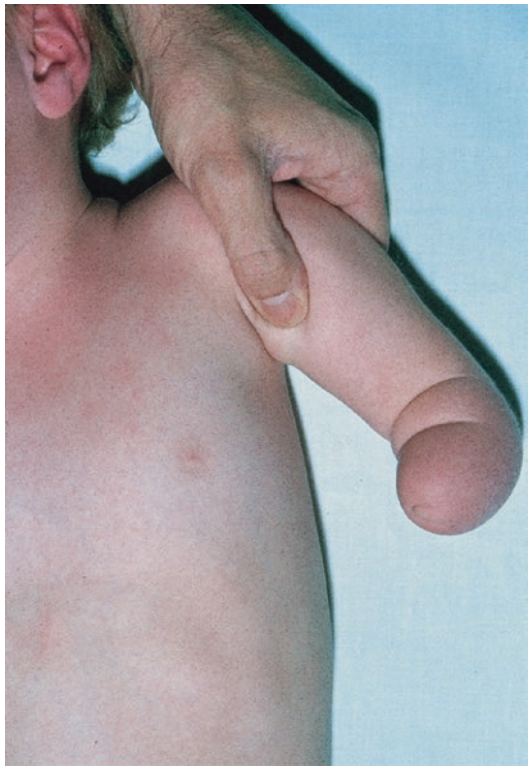


**Fig. 17.3** Incomplete hemimelia of the left arm

**Complete adactylia**
Absence of the thumb and all the fingers and metacarpals.
(dactylos is Greek for finger)

**Complete aphalangia**
Absence of the distal phalanx of the thumb and all the distal finger phalanges.

Explanatory note: **transverse terminal deficiency.** The most frequent terminal deficiency in the upper limb is whereby one third of the forearm is present (incomplete hemimelia). The length of the forearm in a newborn with an incomplete hemimelia is as a rule not more than 7 cm (Fig. 17.3) and in adults not more than 10 cm. Afunctional rudimentary remaining fingers may be present. Pro- and supination are as a rule possible.

Apart from cases where the mother has used Thalidomide (Softenon) during pregnancy (in the 1950s and 1960s), no cause can generally be found for unilateral terminal transverse deficiency. Only if there is a bilateral or multifocal terminal transverse deficiency may one consider autosomal recessive inheritance or a syndrome.

**Fig. 17.4** Forearm prosthesis with an artificial hand without grip function on the right side

Supplementary assessment: X-rays of the involved arm if necessary.

Primary care treatment: the parents should be informed that there are prostheses available but that their child can usually function well without a prosthesis.

When to refer: before the age of 5 and 6 months.

Secondary care treatment: treatment is only necessary if the child is functionally limited because of the deficiency. Future prospects for an improvement in function is a prerequisite for treatment. Improvement in the appearance of the arm or hand must not be at the expense of function. In any event one waits until the child has sitting balance before prescribing an upper extremity prosthesis. As a rule this is at the age of 5 or 6 months. In the first instance a simple prosthesis with an artificial hand without grip function is prescribed (Fig. 17.4). One waits until 2 years of age before prescribing a myoelectric prosthesis (Fig. 17.5). Wearing an upper extremity prosthesis is dependent on the child. The attitude of the parents and others in the direct surroundings play a big

**Fig. 17.5** Right forearm myoelectric prosthesis

role. Older children choose themselves whether to use or not use a prosthesis. One child may wear the prosthesis the whole day and another only for certain activities. Children with a long forearm stump use the prosthesis much less often than children with a short forearm stump. If the child is fitted with a prosthesis before the third year of age, then the chances of successful use are greater than in older children. Extensive operative corrections will preferably be carried out before 3 years of age, because after that a functional improvement becomes more difficult to achieve.

**Amelia** These children will usually not accept a prosthesis. A prosthesis is experienced as unhandy and heavy (Fig. 17.6). Children with a bilateral amelia often develop formidable compensatory possibilities using their feet and toes. As a result they can often use their feet to eat, brush their teeth, write and operate a computer.

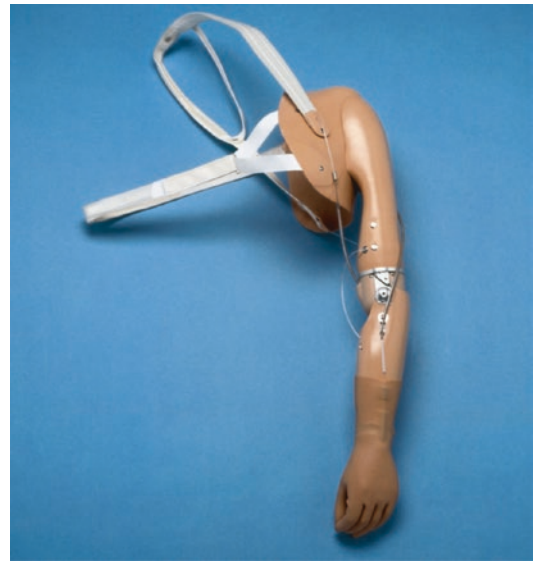


**Fig. 17.6** Body powered shoulder exarticulation prosthesis for an amelia. In unilateral cases the energy required to move the prosthesis is produced by movements of the contralateral shoulder

Patients with a bilateral amelia can possibly benefit from an electrical shoulder exarticulation prosthesis.

**Incomplete amelia or hemimelia** An upper arm prosthesis is given for cosmetic reasons and in order to be able to grip objects. Because there is no elbow, a special elbow hinge is necessary to allow the prosthesis to be fixed at a particular angle. This may be necessary in order to be able to hold particular objects against the body. When walking the elbow hinge must be mobile so that a natural swinging movement can be made. The upper arm prosthesis is heavier and has much less ease of operation compared to a forearm prosthesis. The functional possibilities of the prosthesis are limited and the prosthesis will be worn particularly for cosmetic reasons (Fig. 17.7).

**Incomplete hemimelia** The elbow is stable and can as a rule be overextended to about 30° and has excellent flexion. One can prescribe a prosthesis with an artificial hand that has no grip function when the child has a good sitting balance (Fig. 17.4).

**Fig. 17.7** Upper arm prosthesis

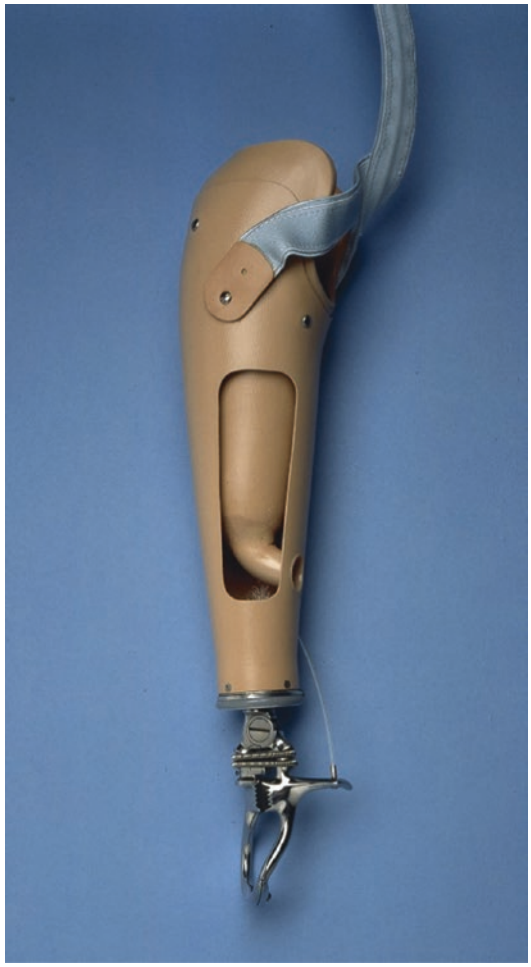


**Fig. 17.8** Lower arm prosthesis with a hook

*Body powered prostheses* After the age of 2 years one can choose for a prosthesis with a hook, which the child can open and close (Fig. 17.8). The prosthesis is ideally suitable for grabbing objects and holding them tight. One can choose a prosthetic hand instead of a hook. A prosthetic hand has more grip function and looks more natural than a hook. The objects to be gripped are less easily seen and more energy is required in order to open the hand than is the case with a hook prothesis (Fig. 17.9).

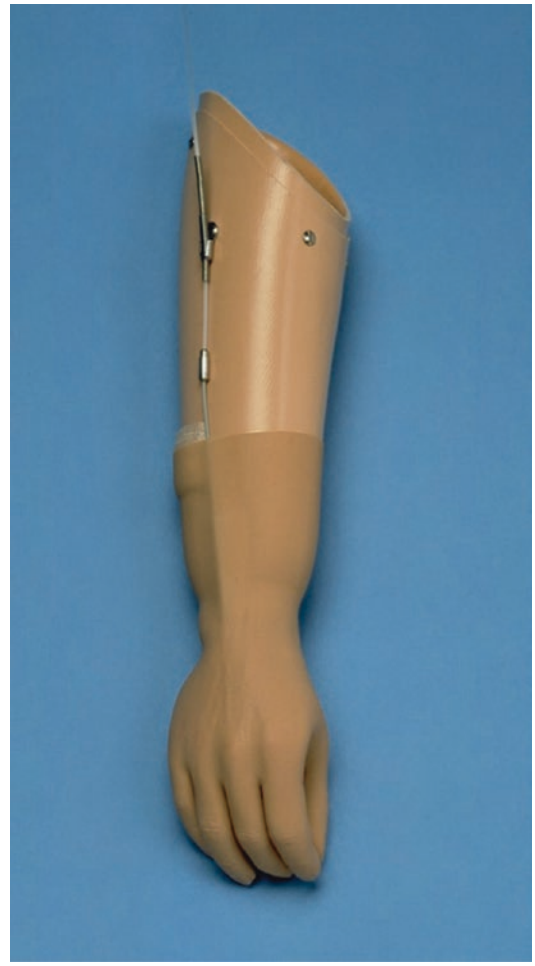


**Fig. 17.9** Body powered lower arm prosthesis with an artificial hand

The energy required to open the prosthetic hook is provided by movement in the elbow or the contralateral shoulder. These two previously mentioned prostheses are called body powered prostheses because the patient has to provide the energy required to open and close the hook or the hand.

*Myoelectrically powered prostheses* There are electrically powered prostheses (Fig. 17.5). The energy required to open and close the artificial hand is provided by a battery. Some electrical prostheses are operated with a pull switch. Other prostheses are operated through a myoelectrical signal from the skin. There is an alternating electrical signal on contracture of the flexors and extensors in the forearm. The myoelectrical hand can be hereby opened and closed. More concentration is required for gripping with a myoelectric prosthesis than with a bodily powered prosthesis.

**Acheiria** As a rule a prosthesis is not useful in patients who have a forearm but no wrist or hand (Fig. 17.10). A prosthesis with a grip function would be longer than a normal arm because of technical reasons. Sometimes a prosthesis may be provided in an older child for cosmetic reasons. In bilateral cases and in blind children a so-called "Krukenberg procedure" can be carried out. Hereby the radius and ulna are operatively separated creating "chopsticks" (Fig. 17.11). The other side can be possibly fitted with a prosthesis with a grip function. One needs good eyesight in order to grip an object with such a prosthesis, because this does not allow "feeling". A blind child cannot be fitted with such a prosthesis with a grip function for this reason.

**Complete adactylia** Hereby the thumb, all of the fingers and the metacarpals are absent. Just as in acheiria a prosthesis is as a rule not meaningful. Grip function can possibly be provided by a so-called opposition orthosis (Fig. 17.12).

**Complete aphalangia** If there is a transverse reduction defect of the distal phalanges of the fingers, the thumb and the fingers are shorter than

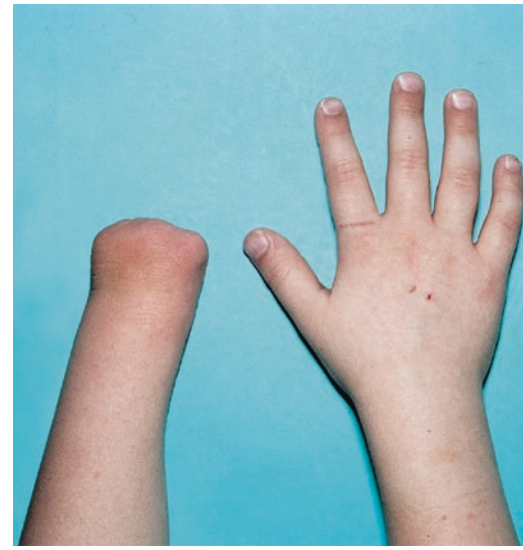


**Fig. 17.10** Acheiria. A prosthesis is as a rule not useful in a patient with an absent wrist and hand

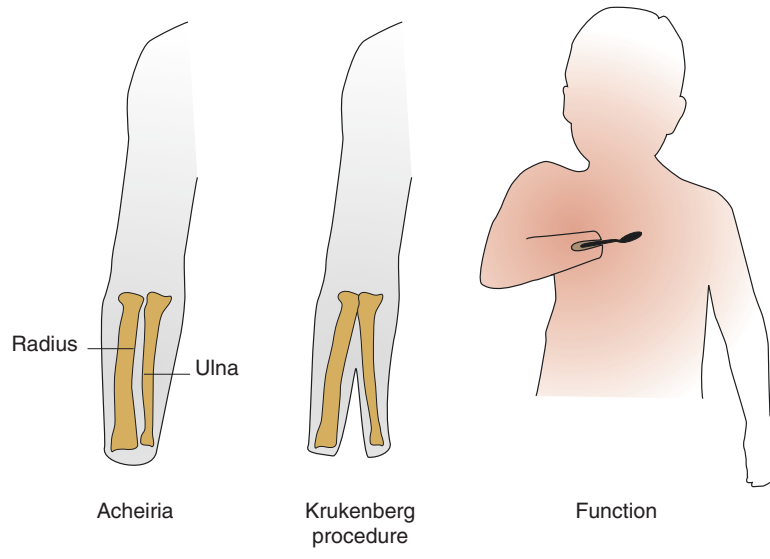



**Fig. 17.11** A Krukenberg procedure can be carried out with an absent wrist and hand. Hereby the radius and ulna are operatively separated creating sort of "chopsticks"

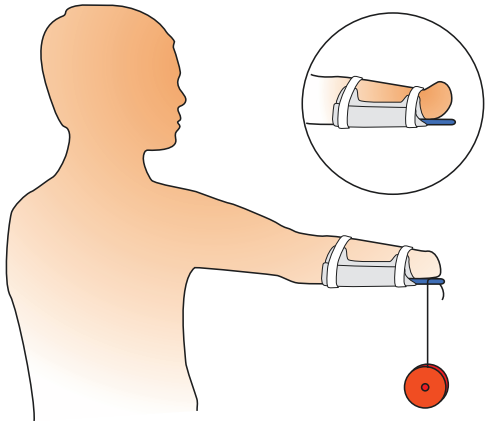


**Fig. 17.12** Opposition orthesis in a patient in which the wrist is present but the rest of the hand is absent (adactylia)

normal, they can often move more or less normally. Often only small objects can be picked up. In order to allow the child to pick up and hold larger objects the soft tissues in the webspace between the thumb and index finger can be operatively widened.

## Absence of the Whole Arm or Upper Arm or Forearm Between the Trunk and the Hand in Which Case the Elbow Joint Is Always Absent

Complaint: between the trunk and the hand the whole arm or the upper arm or forearm is absent. The hand is fixed directly or with the forearm onto the trunk or onto the upper arm.

Assessment: a complete segment is absent from the arm between the trunk and the hand. The joint ligaments are flaccid and as a result (passively) hypermobile. The elbow is always absent. The fingers may also be absent. The strength in the extremity is lessened (Fig. 17.13).

Differential diagnosis (Fig. 17.14):

Transverse intercalary deficiency (phocomelia)

**Complete phocomelia**

The upper arm and forearm are absent. The hand is fixed to the shoulder girdle.

**Proximal phocomelia**

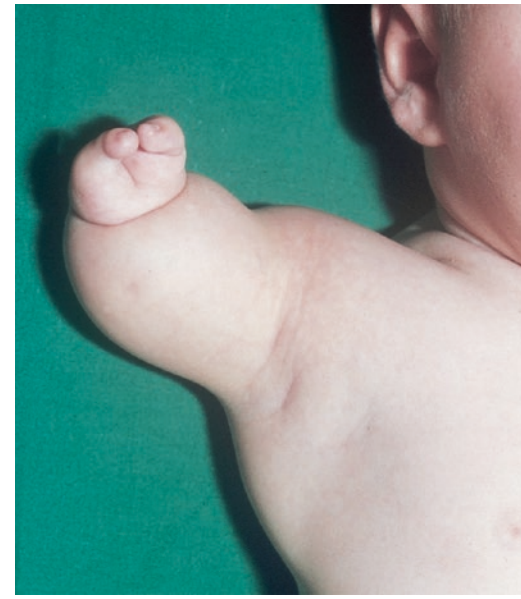


**Fig. 17.13** Distal phocomelia of the left arm. The forearm is absent, the hand is fixed to the upper arm and the fingers are absent

The upper arm is absent. The forearm is fixed to the shoulder girdle.

**Distal phocomelia**

The forearm is absent. The hand is fixed to the upper arm.

Explanatory note: **transverse intercalary deficiency (phocomelia).** Nowadays the deformity is rarely seen except in patients born in the 1950s and 1960s, when pregnant women often received Thalidomide (Softenon).

Supplementary assessment: as a rule not necessary.

Primary care treatment: none.

When to refer: during the first 6 months of life.

Secondary care treatment: **complete phocomelia.** A shoulder exarticulation prosthesis can be prescribed for a complete phocomelia of the upper extremity.

**Proximal and distal phocomelia** In these cases a prosthesis is usually not prescribed. The hand function is as a rule good in the proximal type.

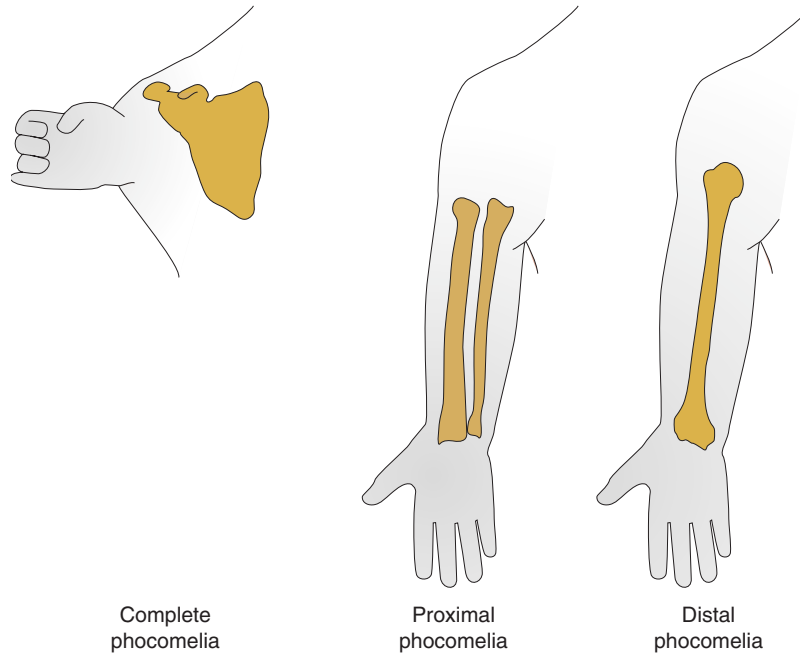



**Fig. 17.14** Classification of intercalary transverse deficiencies (phocomelia) in the upper extremity (Redrawn from: Tachdjian MO. Tachdjian's paediatric orthopaedics. 2nd ed. Philadelphia: Saunders Company; 1990)

The results of an operative treatment to lengthen the arm in the proximal type or to improve shoulder stability are disappointing.

## Abnormal Forearm and/or Hand

Complaint: the hand has an abnormal position and the thumb and/or some of the fingers are possibly absent.

Assessment: the hand may be completely normal in the case of a radial deviation. The thumb may be flaccid and/or small or completely absent. The styloid process of the ulna is prominent. The forearm is short and is possibly curved with the convex side on the ulnar side.

If there is an ulnar deviation the hand can be complete but fingers and/or the thumb may be absent, most often the ring and the little finger. The forearm is short and may be curved radially. In many cases the upper arm is also shortened. Also, if the forearm is curved convexly on the radial side with ulnar deviation of the hand, this does not usually lead to functional problems.

The radial head can be dislocated or fixed to the humerus or there may be an unstable elbow.

There may also be deficiencies in which only the thumb or one or several fingers are absent without forearm deformities.

Differential diagnosis:

Preaxial terminal and intercalary deficiency
**radial hypoplasia or aplasia (radial clubhand)**
type I
type II
type III
type IV
**thumb aplasia**
Postaxial terminal and intercalary deficiency
**ulnar hypoplasia and aplasia (ulnar clubhand)**
type I
type II

type III
type IV
**finger aplasia**
Central deficiency
**cleft hand (ectrodactyly, splithand, lobster-claw hand)**
typical type
atypical type

Explanatory note: **paraxial terminal, paraxial intercalary and central deficiency.** In the classification of hypoplasia and aplasia of the radius and ulna no distinction is made between the presence or absence of a complete hand. Although in both cases the lower arm is involved, the deformities are very different (Table 17.1). The term cleft hand is used for a central deficiency.

**Radial hypoplasia and aplasia** This occurs in 1 in every 100,000 births. In 50 % of cases it is bilateral. It occurs just as frequently in boys and girls. The deformity is not inheritable. If the radius is completely or partly replaced with a connective tissue strand that does not grow with the ulna, then the ulna will be curved with a convexity on the ulnar side. The hand is positioned in radial deviation and the deformity is also known as a radial club hand. The thumb can be normal but can also be absent or flaccid. The deformity can be classified into four types (Fig. 17.15). This is a radius hypoplasia (type I, II and III), if the radius is partly present and if the radius is completely absent then one speaks of radial aplasia (type IV). Usually there are functional problems in the wrist but not in the elbow (Fig. 17.16). There is generally a reasonable function even in an extensive radial deviation.

**Table 17.1** Radial and ulnar hypoplasia and aplasia have very different aspects

| | Radial hypoplasia or aplasia | Ulnar hypoplasia or aplasia |
|---|---|---|
| Incidence | 1 in 100,000 | 1 in 500,000 |
| Part of a syndrome | 25 % | Seldom |
| Bilateral | 50 % | Seldom |
| Inheritable | Not | Not |
| Aplasia | Often | Seldom |
| Elbow function | Good | Type III unstable |
| | | Type IV synostosis |
| Wrist function | Reasonable | Good |
| Hand | Thumb regularly absent | Variable absence of thumb and fingers |

*Type I* Slight shortening of the distal part of the radius. The ulna is not curved. The epiphysis in the proximal part of the radius is normal. The epiphysis in the distal part of the radius is seen later and has less growth potential than normal. There is only a slight radial deviation in the hand

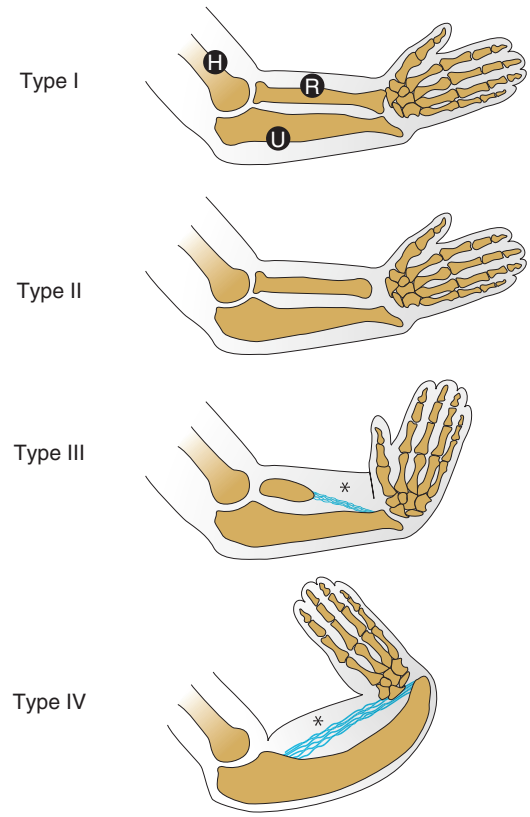



**Fig. 17.15** Heikel's classification for radial hypoplasia and aplasia. Hypoplasia and aplasia of the radius can be divided into four types. If the radius is partially absent then there is a radius hypoplasia (*type I, II and III*), if the radius is completely absent then this is radius aplasia (*type IV*). *H* humerus, *R* radius, *U* ulna, * connective tissue strand (Redrawn from: Heikel HV. Aplasia and hypoplasia of the radius: studies on 64 cases and on epiphyseal transplantation in rabbits with the imitated defect. Acta Orthop Scand. 1959;39:1–115)

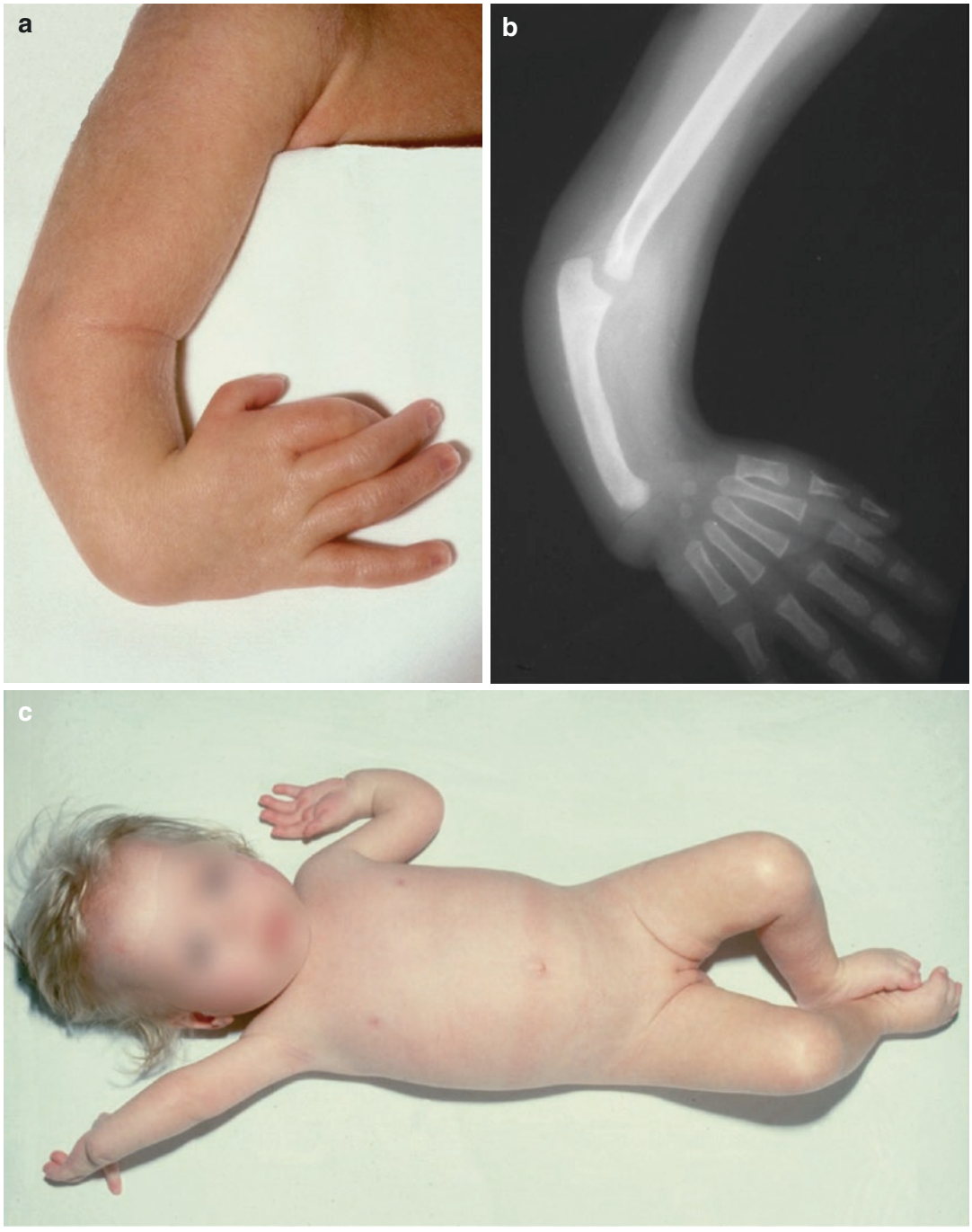



**Fig. 17.16** (**a**) and (**b**): radial aplasia of the right forearm (Heikel type IV). There is an underdeveloped thumb. (**c**) Bilateral radial aplasia

with respect to the wrist. The mobility of the elbow and wrist is normal.

*Type II* The radius is shortened more than in type I. The ulna is thickened and curved convexly to the ulnar side. The proximal and distal epiphysis of the radius are visualized later and the growth potential is less than normal. The hand has a slight radial deviation. The mobility of the elbow and the wrist is normal.

*Type III* The radius is partly absent at the level of the proximal, central or distal part, which is the most frequent. The ulna is thickened, shortened and curved convexly to the ulnar side. The elbow function is mostly normal. The stability of the wrist joint is less because of the absent distal part of the radius. There is a severe radial deviation.

*Type IV* Completely absent radius (radius aplasia). This is the most frequent type (Fig. 17.16). The ulna is severely curved convexly on the ulnar side. There is a radial deviation of the hand which can be as much as 90°. There is a false joint between the hand and the radial side of the distal part of the ulna. The humerus and the clavicle are shorter and the scapula is usually smaller than normal. By the time the child has grown up the total length of the upper extremity is about $^{2}/_{3}$ the length of the normal extremity. The thumb and first metacarpal are absent in more than 80% of cases. The scaphoid and trapezium are absent in more than half of the patients and the lunatum, trapezoid and pisiform in 10%.

The long head of the biceps muscle and the wrist extensors are absent or severely hypoplastic if the radius is completely absent. The finger extensors are present. If the thumb is absent then all the muscles associated with thumb mobility are not present. The radial nerve stops at the elbow and sensation is provided by an abnormal branch of the median nerve. The musculocutaneous nerve may be absent. The ulnar and median nerves are complete. The radial artery is usually rudimentary or absent.

The condition can be an isolated malformation but can also occur in combination with other deformities. It is part of a syndrome in 25% of cases (Table 17.2).

**Thumb aplasia** Absence of the thumb is mostly seen in a hypoplasia or aplasia of the radius or ulna but may also be part of a syndrome (Table 17.3).

**Ulnar hypoplasia and aplasia** This is an anomaly that occurs in 1 in 500,000 births. It is usually unilateral but there are often anomalies in the contralateral hand, such as absent fingers. The anomaly occurs with the same frequency in boys and girls but is not inheritable. The radius will be curved with a convexity towards the radial side if the ulna has been completely or partly replaced with connective tissue which does not grow at the same rate as the radius. The anomaly can be classified into four types (Fig. 17.17). Some fingers may be absent, mostly the ring finger and little finger (Fig. 17.18), but sometimes the thumb and the index finger are absent. If the ulna is partly absent then one calls this ulna hypoplasia (type I and II) and if the ulna is completely absent an ulnar aplasia (type III and IV).

**Table 17.2** Syndromes with radial hypoplasia or aplasia

| |
|---|
| Fanconi anemia |
| Holt-Oram syndrome |
| Juberg-Hayward syndrome |
| Rothmund-Thomson syndrome |
| TAR syndrome |
| Trisomy 13 |
| Trisomy 18 |
| VATERor VACTERL-association |

See Appendix for syndrome characteristics

**Table 17.3** Syndromes with an absent thumb

| |
|---|
| Holt-Oram syndrome |
| Rothmund-Thomson syndrome |
| Trisomy 18 |

See Appendix for characteristics of syndromes

*Type I* Shortened ulna. The ulna is present but shortened. The radius is hardly curved and there is no or limited ulnar deviation of the hand (Fig. 17.19).

*Type II* The distal part of the ulna is absent. The proximal part has a normal joint with the humerus. The radial head may have a normal articulation with the humerus but lateral and posterior dislocations also occur. The elbow is stable and mobile but however, particularly pro- and supination are limited. The radius is curved convexly to the radial side. The hand is deviated towards the ulnar side (Fig. 17.20).

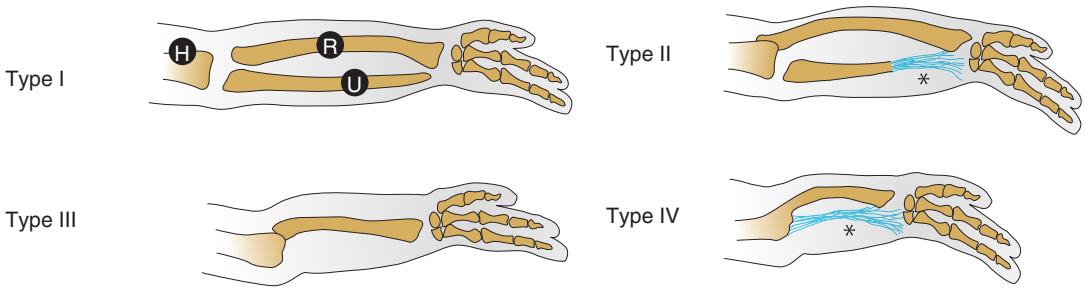



**Fig. 17.17** Bayne classification for ulnar hypoplasia and aplasia. If the ulna is partly present one refers to this as ulnar hypoplasia (*type I and II*) and if completely absent as ulnar aplasia (*type III and IV*). *H* humerus, *R* radius, *U* ulna, * connective tissue strand (Bayne LG, Klug MS. Long-term review of the surgical treatment of radial deficiencies. J Hand Surg Am. 1987;12:169–79)

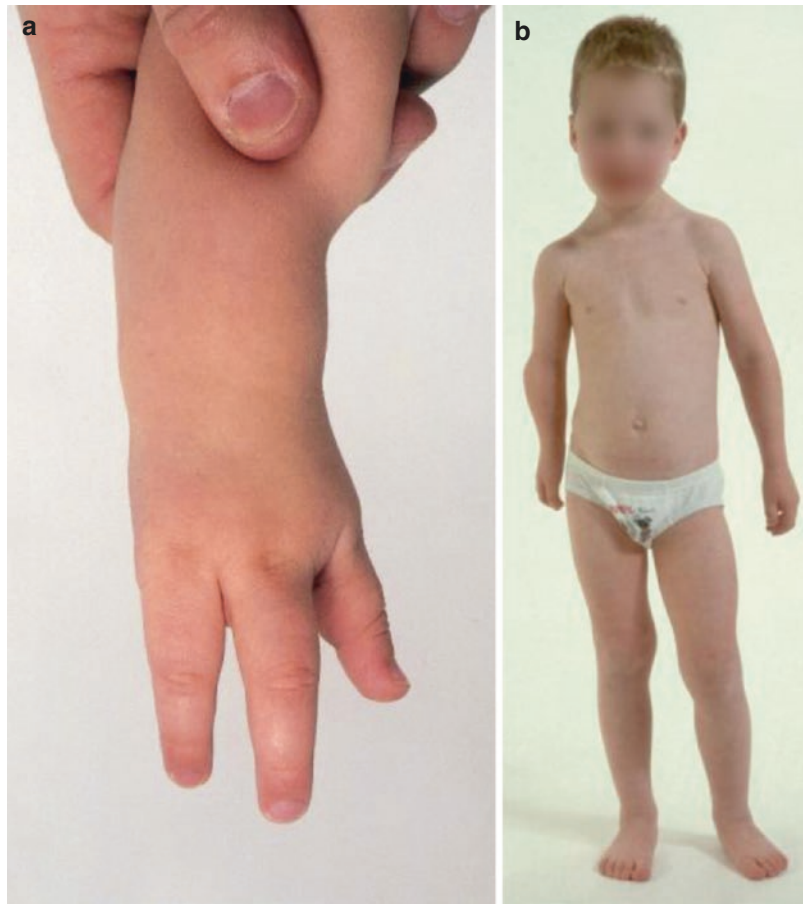



**Fig. 17.18** Ulnar hypoplasia of the right arm with (**a**): absent ring finger and little finger. (**b**): the forearm and upper arm are short

*Type III* This is the rarest type. The ulna is completely absent.

The child has an unstable elbow. The radius is straight because there is no rudimentary connective tissue strand in place of the ulna. The hand is not deviated towards the ulnar side.

*Type IV* The ulna is absent and the radius is fixed to the humerus (so-called humeroradial

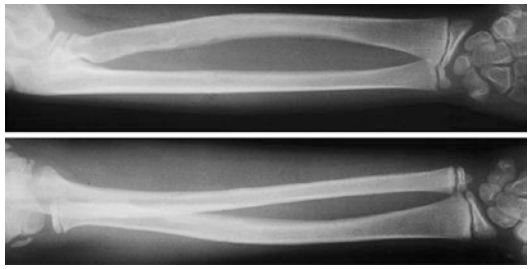


**Fig. 17.19** Ulnar hypoplasia type I in a 13 year old patient. The radius is slightly curved with the convexity towards the radial side

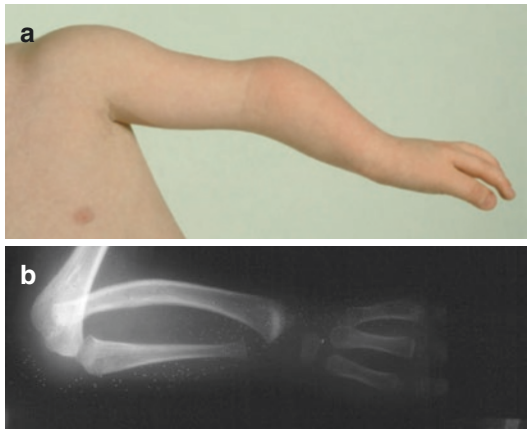



**Fig. 17.20** (**a**) Ulnar hypoplasia type II. The radius is curved convexly to the ulnar side and there is ulnar deviation of the hand. (**b**) The distal part of the ulna is absent. The radial head is displaced laterally and posteriorly

synostosis) (Fig. 17.21). Usually there is a rudimentary ulna and that limits length growth in the radius. The radius increasingly bends with a radial convexity and, above all, wedging of the distal radial epiphysis occurs causing an increase in the ulnar deviation of the hand. The length of the forearm remains limited to 2/3 of normal. In type IV in which the distal humeral epiphyseal growth is lacking, the final length of the upper arm will be four fifths of normal (the epiphyseal growth plate in the distal part of the humerus contributes 20 % to the humeral length growth).

Other inborn errors that occur in combination with ulnar hypoplasia or aplasia are as a rule limited to the locomotor apparatus in contrast to the radius.

Associated deformity's are absent fingers of the contralateral hand. However, a clubfoot, fibular hypoplasia and aplasia, spina bifida and an absent patella may also be part of the problem.

**Finger aplasia** Absence of a finger (Fig. 17.22) is as a rule not accompanied by functional problems.

**Cleft hand** There are two types: a typical and an atypical type.

*Typical type* This is often familial, often bilateral and is associated in 50 % of cases with cleft feet. The simplest type is a cleavage between the third and fourth rays in which there is no tissue missing. In severe types there is a V-shaped defect in the central part of the hand (Fig. 17.23). One or more fingers are absent: middle finger; middle and index finger; middle and ring finger or middle, index and ring finger, all of which can be combined with absent metacarpals. The thumb and the little finger and their metacarpals are always present. Syndactyly of the thumb and index finger and of the ring- and little finger is frequently seen.

*Atypical type* There is a U-shaped defect and the central rays are absent and the thumb and little finger are underdeveloped (Fig. 17.24). Rudimentary fingers between the thumb and the little finger may be present. The opposition function of the thumb and the little finger is reduced. This type is usually unilateral, non familial and is not associated with foot anomalies.

Supplementary assessment: X-rays of the whole upper extremity.

Primary care treatment: the parents should be told that the function of the arm and hand will be reasonable even in the case of a extensive type of hypoplasia or aplasia of the radius and the ulna. The function of a typical cleft hand is reasonable or good but very limited in the case of an atypical cleft hand. Absence of a finger does not lead to functional invalidity. There are operative possi-

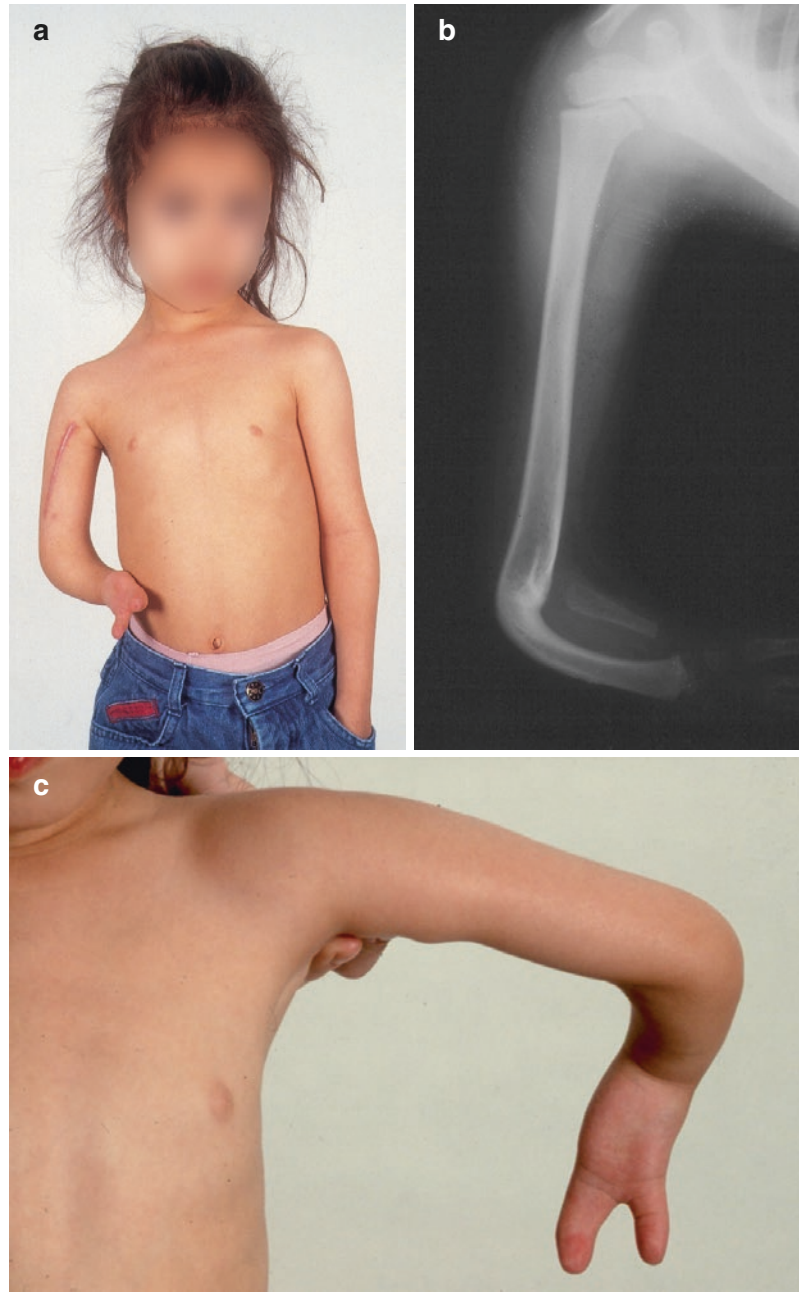



**Fig. 17.21** (**a**) Ulnar aplasia type IV. (**b**) The ulna is absent. there is a humeroradial synostosis. (**c**) There are only two fingers

bilities for an absent thumb. In hypoplasia or aplasia of the radius other anomalies have to be excluded.

When to refer: in a hypoplasia or aplasia of the radius the child should be referred to a pediatrician in order to check for other possible associated anomalies or syndromes. At the same time an appointment must be made with a pediatric orthopedic surgeon or pediatric plastic surgeon. Referral should be made to a pediatric orthopedic surgeon in the case of ulnar hypoplasia and aplasia.

The child should be referred to a pediatric plastic surgeon before 9 months of age if the thumb is

absent. The same applies if there is a serious divergence between the ulnar and radial component of a cleft hand.

Secondary care treatment: **radial hypoplasia and aplasia.** There has been a lot of discussion about the treatment of this condition. Even if the hypoplasia or aplasia of the radius is not treated, the final function of the hand will be reasonable.

**Type I** In this case treatment is as a rule not necessary. A pollicisation procedure may be

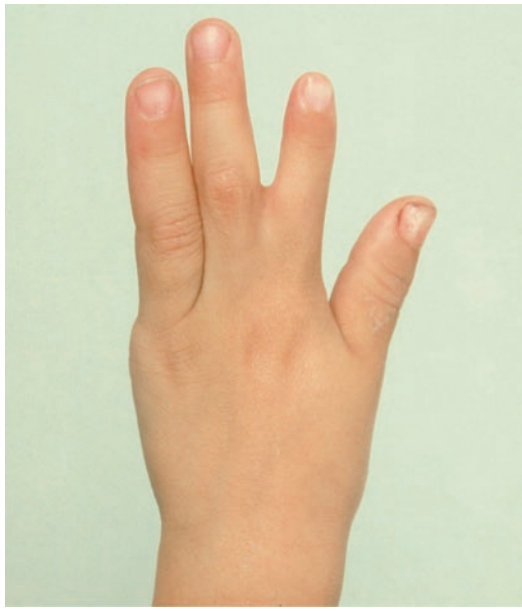


**Fig. 17.22** Finger aplasia (absent little finger). There is also an incomplete syndactyly between index and middle finger

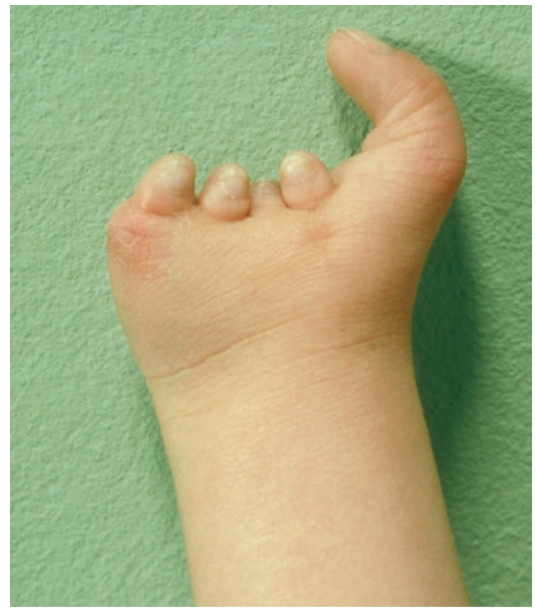


**Fig. 17.24** Atypical cleft hand

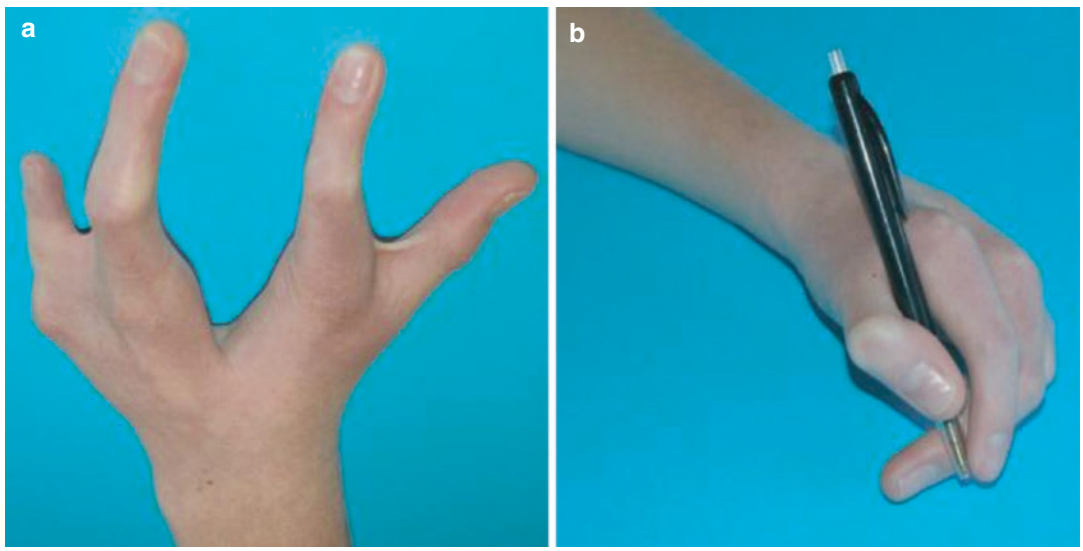



**Fig. 17.23** (**a**) Typical type of cleft hand. Absent middle finger. Incomplete syndactyly of the thumb and index finger and of the ring finger and little finger. (**b**) The function of this hand is good

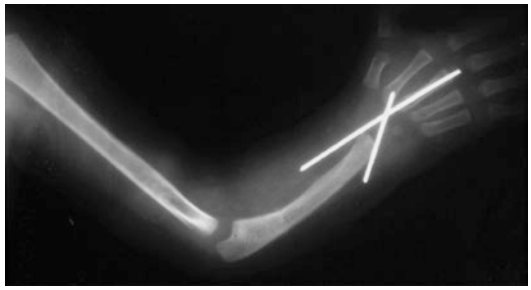


**Fig. 17.25** Centralization of the hand in a type IV radial club-hand

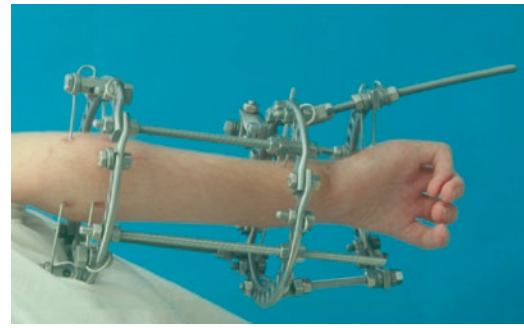


**Fig. 17.26** Centralization of the hand in radial clubhand at an older age, preferably using an Ilizarov frame

considered if the thumb is absent. The index finger will be made into a thumb to allow opposition. This procedure can best be carried out between 9 and 12 months of age, because this gives the best final result.

**Type II** The radial deviation of the hand will be treated with manual corrections and plaster cast immobilization. This should be started as quickly as possible after birth. A splint will be prescribed 3 months after birth. This will be worn in the first instance day and night and must only be taken off when the child is washed. There is no point in applying plaster corrections in children older than 2 years of age because the shortened radial structures have to be loosened operatively.

**Type III and IV** Centralization of the hand will mostly be chosen for cosmetic reasons (Fig. 17.25). Operative treatment will preferably be carried out between the ages of 3 and 6 months if there are no contraindications such as in Fanconi anemia with heart anomalies. The hand will be centralized distally on the ulna. In a unilateral anomaly the hand is fixed in 30° pronation. In the case of a bilateral deformity the dominant hand is centralized with 30° of pronation and the non dominant hand in 30° of supination because the dominant hand will be used later for eating, drinking and writing and the non dominant hand for cleaning the bottom after using the toilet. An osteotomy of the ulna can be carried out during the same operation if the lower arm is severely bent. After this the child must wear a lower arm splint day and night up to the age of 6 years. After 6 years of age the brace will only be worn at night until the child has finished growing. Occasionally an anterior transposition of the triceps muscle must be carried out for elbow flexion in case of absence of the biceps tendon or the musculocutaneous nerve. Centralization of the hand at a later age is preferably treated with an Ilizarov frame (Fig. 17.26).

**Thumb aplasia** A pollicisation of the index finger may possibly be considered in order to create opposition function.

**Ulnar hypoplasia and aplasia** The treatment in the first instance consists of frequent manual corrections of the ulnar deviation with plaster cast immobilization. If a good position is achieved the treatment will be continued with a removable splint which keeps the hand in a good position.

**Type I** The lengthening procedure for the ulna after 10 years of age. This procedure may, if neccesary, be repeated between the ages of 12 and 16 years.

**Type II** The connective tissue strand has to be divided between the ages of 6 and 12 months in order to prevent increase in the curvature of the radius. A correction osteotomy may be necessary in case of a severely bent radius.

**Type III** An upperarm splint with an elbow hinge may be considered in the case with an unstable elbow.

**Type IV** Division of the rudimentary ulna connective tissue strand at 6–12 months of age. A corrective osteotomy may be considered in the case of an extensively curved radius.

**Finger aplasia** This does not lead to functional problems and does not need to be treated.

**Cleft hand** The divergence between the ulnar and radial component can be a problem with regard to good function in the typical type. The atypical type has no possible opposition function.

**Typical type** If the ulnar and radial component are strongly divergent then they can be operatively approximated between the ages of 6 months and 2 years of age.

**Atypical type** In order to achieve opposition function a Z-plasty or Z-plasties and osteotomies of one or more metacarpals can be carried out before 3 years of age. Rudimentary fingers between the thumb and little finger will be removed. In a one fingered hand one can consider a toe to thumb transplant or a toe to little finger transplant.

**Differential Diagnosis: Complete or Partial Absence of an Arm**

| **Absence of the whole or part of the arm** | **Transverse terminal deficiency** |
|---|---|
| Absence of the whole arm. | **Amelia** |
| Absence of the hand, lower arm and part of the upper arm. | **Incomplete amelia** |
| Absence of the hand and lower arm. | **Hemimelia** |
| Absence of the hand and part of the lower arm. | **Incomplete hemimelia** |
| Absence of the hand and wrist. | **Acheiria** |
| Absence of the thumb, all the fingers and metacarpals. | **Complete adactylia** |
| Absence of phalanges in the whole width of the hand. | **Complete aphalangia** |
| **Absence of the whole arm or upper arm or forearm between the trunk and the hand in which the elbow joint is always absent** | **Transverse intercalary deficiency (phocomelia)** |
| Absence of the upper and forearm. The hand is fixed to the shoulder girdle. | **Complete phocomelia** |
| Absence of the upper arm. The forearm is fixed to the shoulder girdle. | **Proximal phocomelia** |
| Absence of the forearm. The hand is fixed to the upper arm. | **Distal phocomelia** |
| **Abnormal forearm and/or hand** | **Paraxial terminal and intercalary deficiency** |
| Absence or partial absence of the radius | **Radial hypoplasia and aplasia (radial clubhand)** |
| Limited radial deviation of the hand. Radius slightly shortened, straight ulna. | Type I |
| Limited radial deviation of the hand. Radius is shortened, the ulna is thickened and bent (convex to the ulnar side). | Type II |
| Severe radial deviation of the hand. Partial absence of the radius, the ulna is shortened, thickened and bent (convex to the ulnar side). | Type III |
| Severe radial deviation of the hand. Total absence of the radius, the ulna is short, thickened and bent (convex to the ulnar side). | |
| The thumb is absent in 80 % of cases. | Type IV |
| Absence of only the thumb. | **Thumb aplasia** |
| Absence or partial absence of the ulna | **Ulnar hypoplasia and aplasia (ulnar clubhand)** |

| | |
|---|---|
| Ulnar deviation of the hand. Ulna shortened, the radius is straight. | Type I |
| Ulnar deviation of the hand. Distal part of the ulna is absent, radius is slightly bent (convex radially), possible radial head dislocation. | Type II |
| No ulnar deviation of the hand. Total absence of the ulna, the radius is straight, the elbow is unstable. | Type III |
| Ulnar deviation of the hand. Totally absent ulna, the radius is short, thickened and severely bent (convex radially) and fixed to the humerus. | Type IV |
| Absence of only a finger. | **Finger aplasia** |
| Cleft hand or absence of central fingers | **Central deficiency** |
| Cleft hand or absence of one or more central rays of the hand. | **Cleft hand (ectrodactyly, splithand, lobster-claw hand)** |
| V-shaped defect. | Typical type |
| U-shaped defect. | Atypical type |

# Lower Limb

## Absence of the Whole or Part of the Leg

Complaint: the leg or part of the leg and/or the foot is absent.

Assessment: the leg or part of the leg or foot is absent. The rest of the leg has developed normally.

Differential diagnosis (Fig. 17.27):
Transverse terminal deficiency

**Amelia**
Absence of the whole leg.

**Incomplete amelia**
Absence of the foot, lower leg and part of the upper leg.

**Hemimelia**
Absence of the foot and lower leg.

**Incomplete hemimelia**
Absence of the foot and part of the lower leg.

**Apodia**
Absence of the foot and the ankle.

**Complete adactylia**
Absence of all toes and metatarsals.

**Complete aphalangia**
Absence of the distal phalanges of all toes.

Explanatory note: **transverse terminal deficiency.** A terminal transverse deficiency of the lower extremity usually gives less functional invalidity than in the upper extremity. The child can stand and walk with a weight bearing telescopic prosthesis which can be lengthened with adaptations to the shoes.

Supplementary assessment: X-rays of the involved leg.

Primary care treatment: inform the parents that their child fitted with a prosthesis or with shoe adaptations will be able to manage reasonably well or even extremely well.

When to refer: before the age of 1 year.

Secondary care treatment: one can begin with a prosthesis for the lower extremity at 1 year of age. In the first instance one will provide a prosthesis without a knee joint in the case of (incomplete) amelia or hemimelia. At the age of 2 the knee joint can be fitted to the prosthesis. Prostheses must be checked every 3 or 4 months because they often become defective because of the intensive use and because the children grow quickly and the prosthesis must be regularly adjusted.

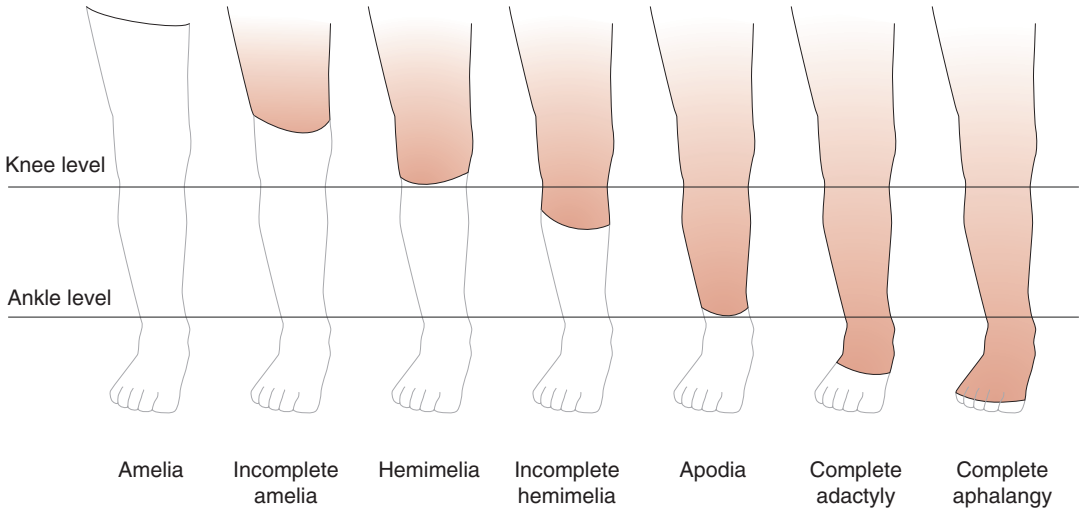



**Fig. 17.27** Classification of terminal transverse deficiencies of the lower extremity

**Amelia** The leg is completely absent. In the first instance these children will be provided with a simple stilt prosthesis (Fig. 17.28). This can later be replaced with a prosthesis with a hip and knee joint, a so-called hip exarticulation prosthesis. In bilateral cases they will be at first be provided with a pelvic basket, which is mounted onto a plate with wheels. Hereby the child can get used to the vertical position of the body. Later on this can be replaced with a bilateral prosthesis with hip and knee hinges whereby in the first instance the hinges will be fixed. The hinges will be made functional once the child can stand well.

**Incomplete amelia and hemimelia** A simple upper leg prosthesis without a knee joint will be provided once the child has just started to stand or walk. After the age of about 2 years one can provide the child with a prosthesis with a knee joint (Fig. 17.29).

**Incomplete hemimelia** Usually half of the lower leg is present. The stump is sometimes slightly in varus. These children get a lower leg prosthesis (Fig. 17.30).

**Apodia** The foot is absent. The lower leg, including the epiphysis of the tibia and fibula are present. These children get a lower leg prosthesis.

**Complete adactylia** The toes and metatarsals are absent, mostly including the cuneiform bones and the cuboid bone. There is also an equinus position in the hind foot. They cannot walk properly without the forefoot. The children get a shoe with a high firm shaft and the anterior space is filled up with felt or foam rubber.

**Complete aphalangia** Shoe modification.

## Absence of the Whole Leg or Thigh or Lower Leg Between the Trunk and the Foot, in Which Case the Knee Joint Is Always Absent

Complaint: the upper and/or lower leg are completely absent between the pelvic girdle and the foot (Fig. 17.31). The foot is fixed directly onto the pelvic girdle or onto the lower leg or thigh.

Assessment: absence of one segment between the trunk on the one side and the foot on the other. The ligaments of the joints are flaccid and apart from that (passively) hyper mobile. The knee joint is always absent. Toes can also be absent. The leg strength is diminished.

Differential diagnosis (Fig. 17.32):

**Fig. 17.28** Prosthesis for amelia of the right leg (and a proximal focal femoral deficiency of the left leg). In the first instance a stilt prosthesis will be given. This can later be replaced with a prosthesis with a hip and knee joint

Transverse intercalary deficiency (phocomelia)

**Complete phocomelia**

Absence of the upper and lower leg. The foot is attached to the pelvic girdle

**Proximal phocomelia**

Absence of the upper leg.

The lower leg is attached to the pelvic girdle.

**Distal phocomelia**

Absence of the lower leg.

The foot is attached to the upper leg (Fig. 17.33).

Explanatory note: **transverse intercalary deficiency (phocomelia).** The deformity is extremely rare. In the 1950s and 1960s it

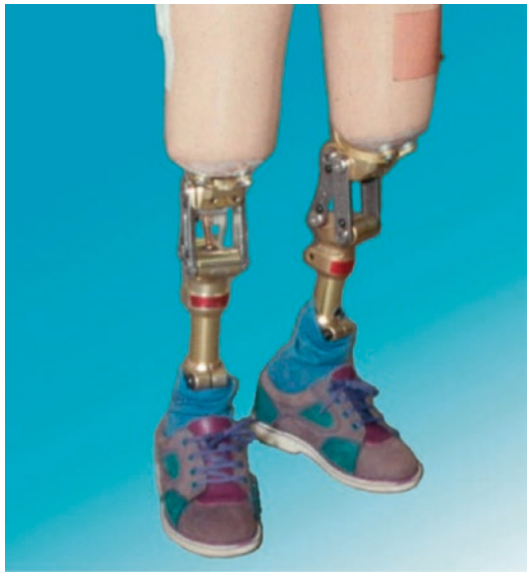


**Fig. 17.29** Prostheses with knee joints

occurred more frequently because of the use of Thalidomide (Softenon) during pregnancy.

Supplementary assessment: X-rays of the whole leg.

Primary care treatment: none.

When to refer: during the first 6 months of life.

Secondary care treatment: **complete phocomelia.** In the complete type an unconventional hip exarticulation prosthesis will be provided in which there is an opening in the prosthesis for the foot. The foot is used to improve balance. A stilt prosthesis will be used in the first instance. Hip and knee hinges are added once the child is older.

**Proximal phocomelia** The ligaments between the pelvis and tibia are flaccid so that a prosthesis with a tuberosity support will be chosen.

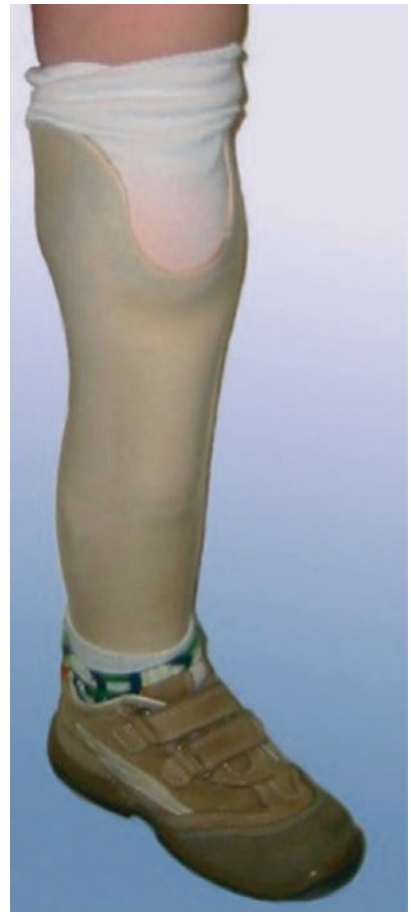


**Fig. 17.30** Lower leg prosthesis

**Distal phocomelia** A lengthening prosthesis will be provided for the distal type.

## Short Upper Leg, the Knee Joint Is Always Present

Complaint: there is an abnormally short thigh.

Assessment: the deformity is usually discovered directly after birth. There is an abnormally short upper leg, sometimes so bad that the foot on the involved side may not reach further down than the normal knee.

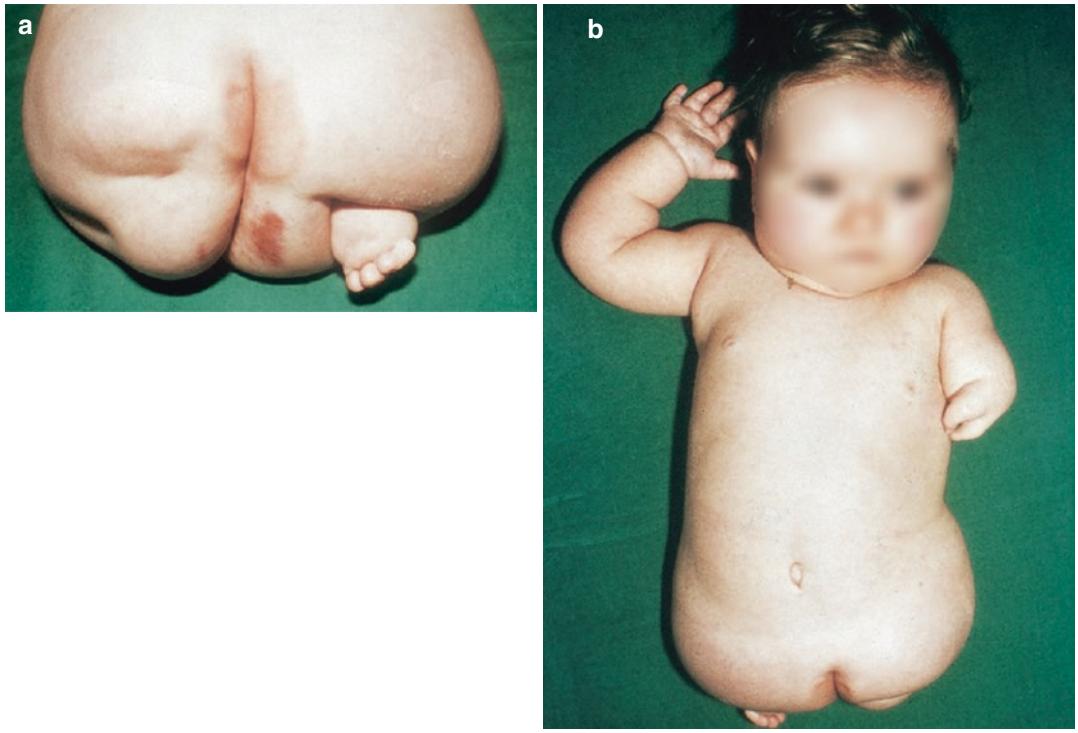



**Fig. 17.31** (**a**) Complete phocomelia of the right leg and amelia of the left leg. (**b**) The left arm shows a distal phocomelia. A number of fingers of the left hand are absent

Differential diagnosis:

**congenital short femur (femoral hypoplasia)**
- type I
- type II
- type III
- type IV

**proximal focal femoral deficiency**
- type I
- type II
- type III
- type IV

Explanatory note: the frequency of this anomaly is 1 in 50,000 life births. The radiological classification in newborns gives problems because a great deal of the skeleton, particularly the proximal femoral epiphysis, is not yet ossified. A more pragmatic classification is a clinical one with two groups:

**Congenital short femur (group 1)** The foot on the short thigh is at the level of the middle part of the lower leg or more distal compared to the normal (contralateral) leg. This is the group with a congenital short femur or femoral hypoplasia (Fig. 17.34 and 17.35). In these children the hip joint is present with a possible varus or valgus positon of the femoral neck with respect to the femoral shaft. In some children there may also be a slight flexion contracture and/or valgus position in the knee, with possible knee instability. The lower leg is often shortened.

*Type I* Hypoplastic femur with a normal hip joint.

*Type II* Hypoplastic femur with a valgus position in the proximal part of the femur, the femoral neck and head are smaller.

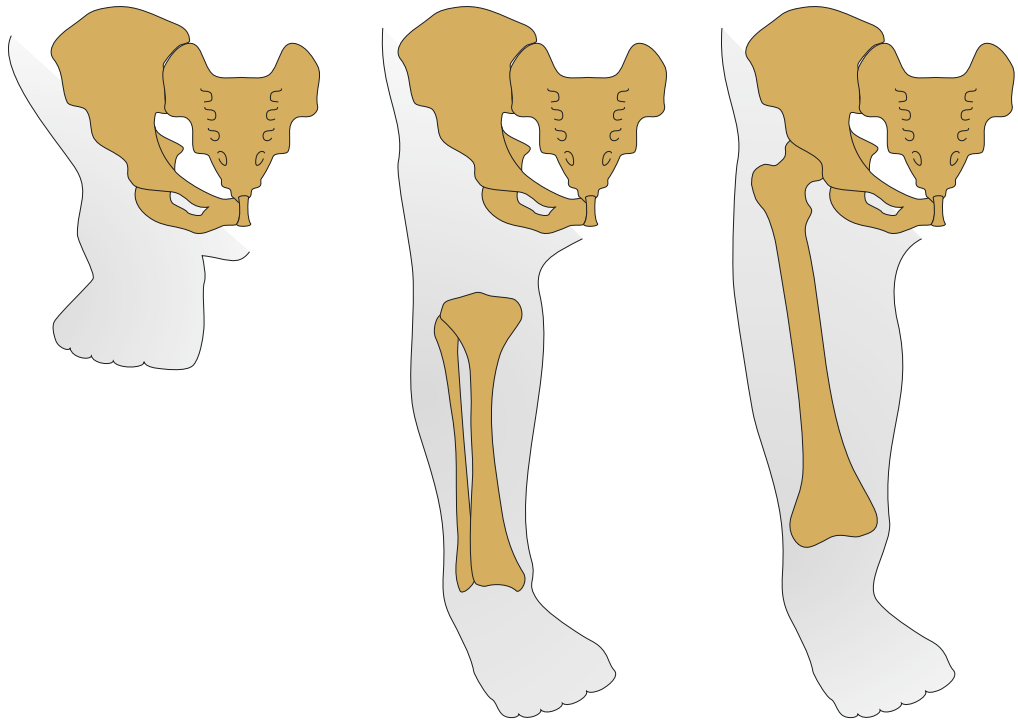



**Fig. 17.32** Classification of intercalary transverse deficiencies (phocomelia) of the lower extremity

*Type III* Hypoplastic femur with a varus position in the proximal part. The cortex in the proximal part of the femoral diaphysis is thickened and the lateral femoral condyle is often deficient.

*Type IV* Hypoplastic femur with an irregular epiphysis in the distal part of the femur, the knee joint is absent and only part of the fibula or tibia is present.

**Proximal focal femoral deficiency (group 2)** In this group the foot of the shortened thigh is at the level of the knee or more proximal compared to the contralateral normal side. The thigh is short and thick, there is a flexion, abduction and external rotation contracture in the hip. The knee joint is always present but has a flexion contracture (Fig. 17.36). The tibia is shortened and in 50 % of cases there is a completely or partially absent fibula (aplasia c.q. hypoplasia of the fibula) (Figs. 17.37, 17.38, 17.39, and 17.40).

*Type I* Normal acetabulum, varus deformity of the proximal part of the femur.

*Type II* Normal acetabulum, pseudarthrosis of the proximal part of the femur.

*Type III* Underdeveloped acetabulum, femoral head absent. Proximal ossification of the remaining part of the femur.

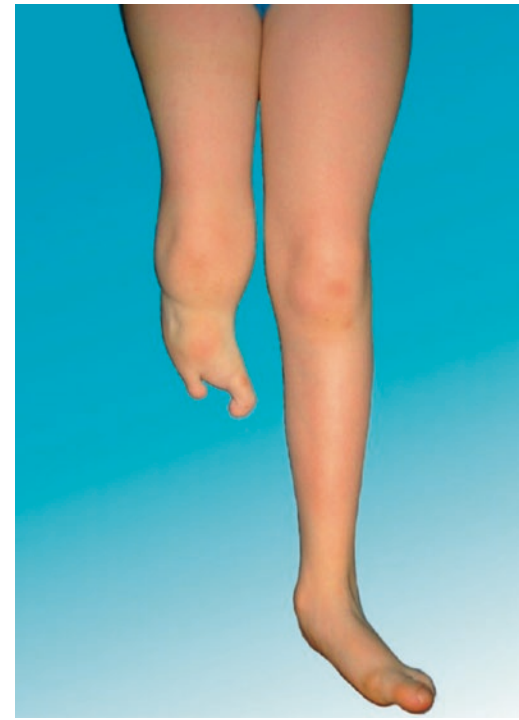


**Fig. 17.33** Distal phocomelia of the right leg. The foot is attached to the upper leg. A number of toes are absent in the right foot

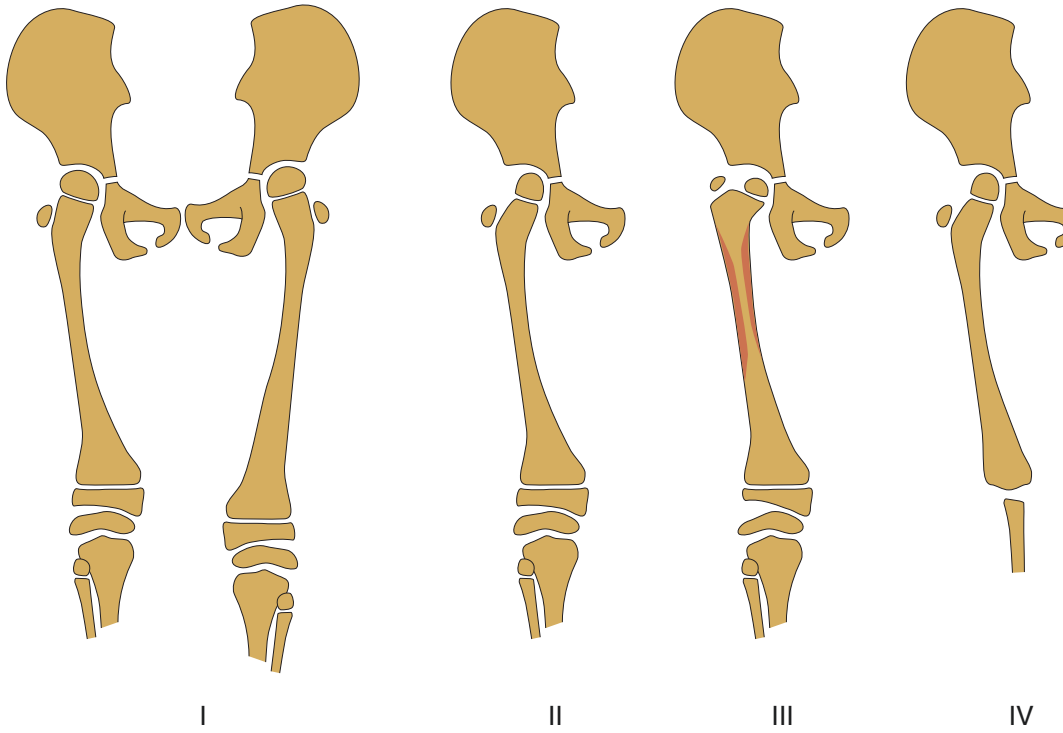



**Fig. 17.34** Radiological classification or a congenital short femur (femoral hypoplasia) according to Pappas, types VI to IX. There is more or less a normal hip joint (Redrawn from: Pappas AM. Congenital abnormalities of the femur and related lower extremity malformations: classification and treatment. J Pediatr Orthop. 1983;3:45–60)

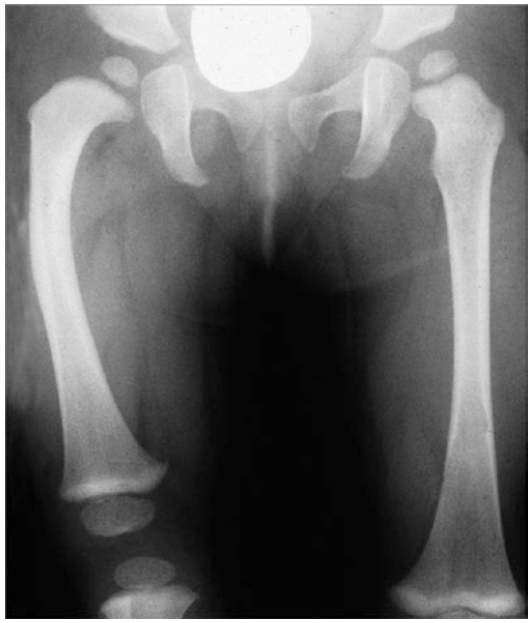


**Fig. 17.35** Congenital short femur, type III on the right side

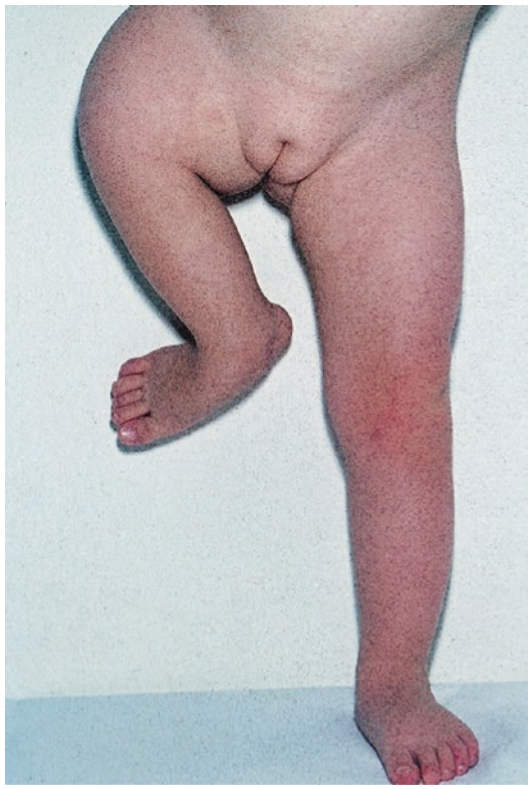


**Fig. 17.36** Right proximal focal femoral deficiency

*Type IV* No acetabulum, only the distal part of the femur is present.

Supplementary assessment: X-rays of both lower extremities.

Primary care treatment: explain to the parents that their child with a prosthesis can begin to walk at the same age as other children.

When to refer: referral to a pediatric rehabilitation center in the first year where the parents get an overview of the treatment possibilities and can be brought into contact with other children who are being treated for the same problem.

Secondary care treatment: **congenital short femur (group 1).** If the child is ready to stand and walk it will be fitted with a rigid leg prosthesis in which the leg length difference is corrected. The foot is positioned in equinus for cosmetic reasons (Fig. 17.41). The foot must be moved daily so that it remains flexible. In case of knee instability the lower leg prosthesis can be extended with a thigh brace and a knee hinge.

A thigh lengthening can be carried out if there is a stable knee if the leg length difference in the thigh is less than 30 %. The van Nes rotation plasty can be considered if the shortening of the thigh is 50 % or more. In this procedure a knee arthrodesis is carried out and at the same time an osteotomy of the lower leg at the level of the knee, whereby the foot can be internally rotated so that it is posteriorly directed. The ankle must be at the level of the contralateral knee joint for a good prosthetic application. The growth plate will be left intact. At this time a possible epiphysiodesis can be carried out if the posteriorly directed ankle lies distal to the contralateral knee joint (Fig. 17.42). There should be a normally mobile ankle joint if a van Nes procedure is to be carried out. The deformity must not be bilateral. The leg should also have sufficient length so that the posteriorly directed ankle is at the same height as the contralateral knee joint. Preferably there should be a stable hip. This technique gives good functional results but the psychological

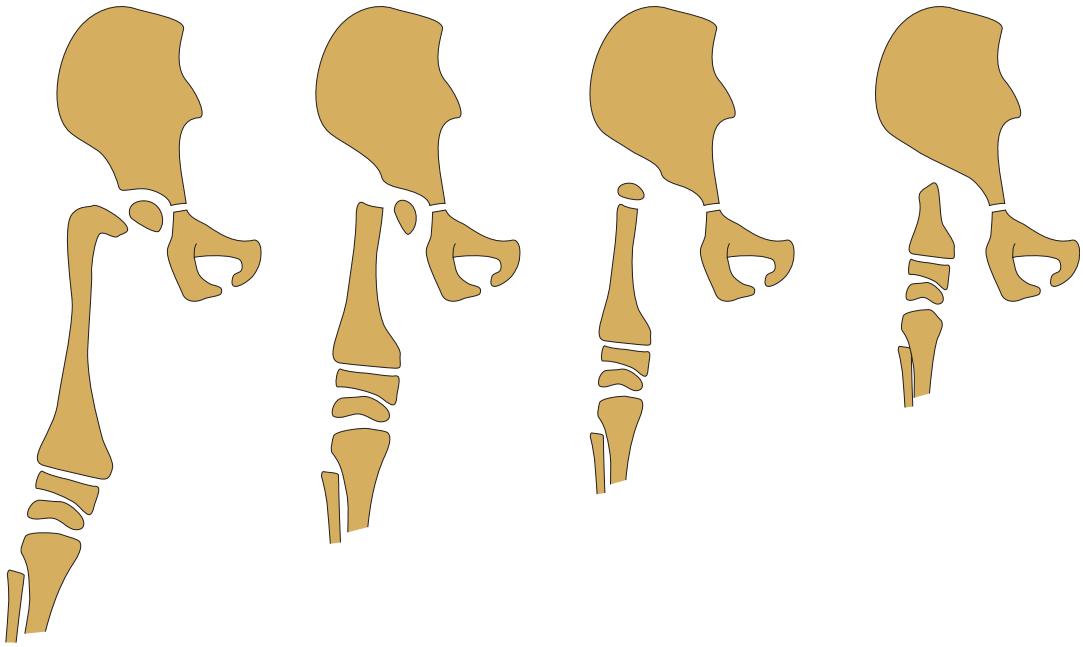



**Fig. 17.37** Radiological classification of proximal focal femoral deficiency according to Aitken. The hip joint is absent in half of the cases. In cases in which the hip joint is present there is a varus deformity or a pseudarthrosis of the proximal part of the femur (Redrawn from: Aitken G. Proximal femoral focal deficiency: a congenital anomaly. In: Aitken GT, editor. A symposium on proximal femoral focal deficiency: a congenital anomaly. Washington, DC: National Academy of Sciences; 1969. p. 1)

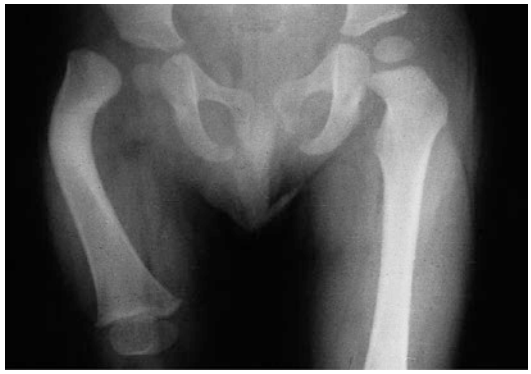


**Fig. 17.38** Right sided proximal focal femoral deficiency type I

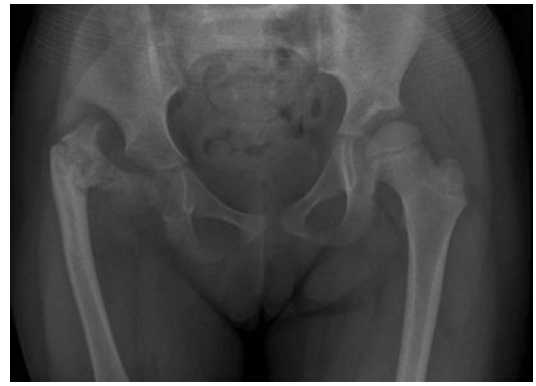


**Fig. 17.39** Right sided proximal focal femoral deficiency type II

aspects should not be underestimated. The operation can be carried out from 2 years of age onwards, however under the age of 12 a spontaneous derotation in the leg can often occur so that a new rotation osteotomy has to be performed sometimes on three occasions.

**Proximal focal femoral deficiency (group 2)** Operations are only necessary in order to provide as good base of a leg lengthening prosthesis (Fig. 17.43). In some cases the prosthesis must be fitted with a tibial tuberosity support to unburden a femoral part that is not supported by the pelvis.

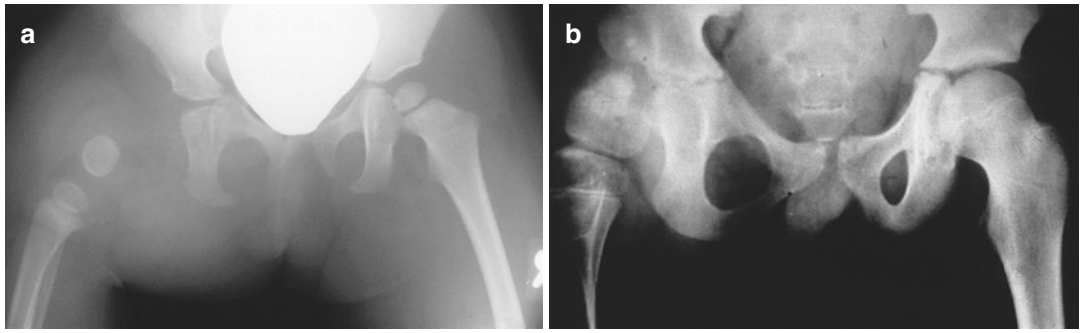



**Fig. 17.40** Proximal focal femoral deficiency type IV. (**a**) Right unilateral, at the age of 2 years. (**b**) At the age of 16 years

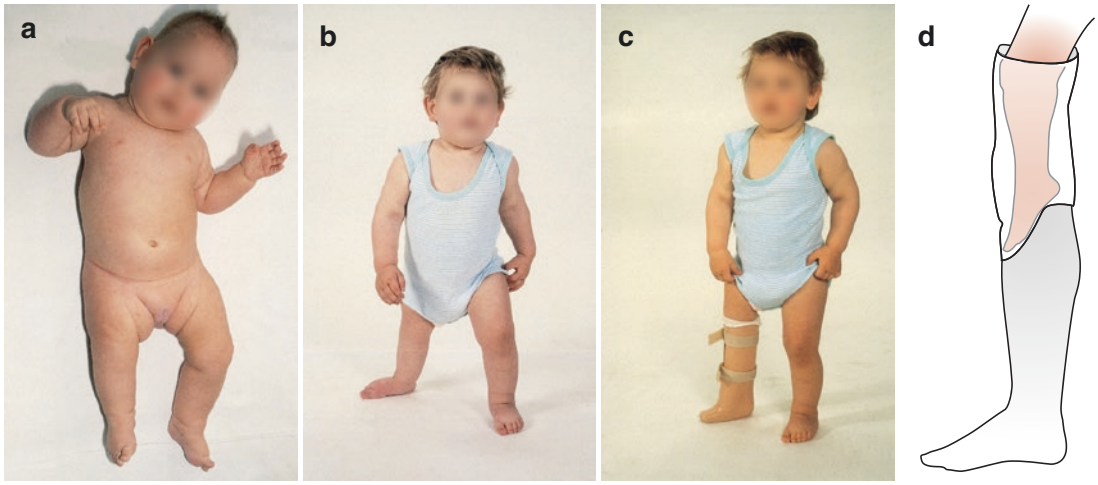



**Fig. 17.41** Congenitally short right femur. (**a**) At birth. (**b**) At 1.5 years of age. (**c**) Lengthening prosthesis. (**d**) The foot is positioned in the lower leg splint in equinus for cosmetic reasons

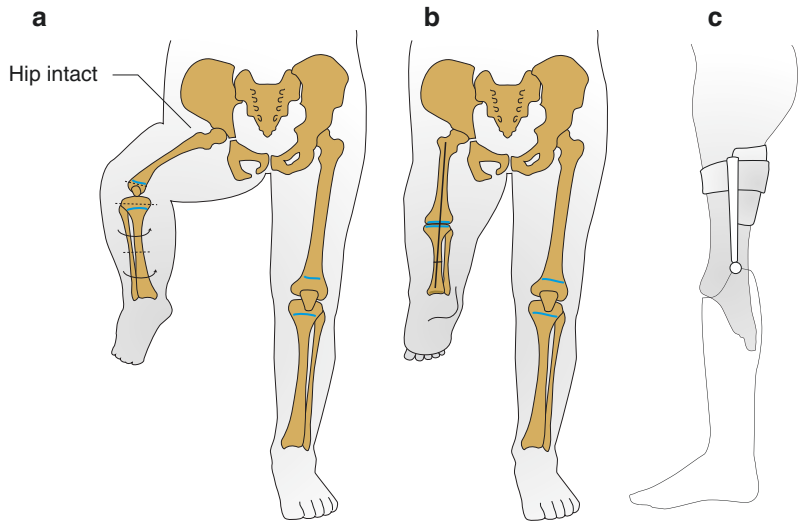



**Fig. 17.42** Internal rotation plasty according to van Nes for a congenital short femur. (**a**) In this procedure an arthrodesis of the knee is carried out and in the same operation the leg is rotated 180° after performing an osteotomy in the proximal part of the tibia. (**b**) The foot is now directed posteriorly. (**c**) After this a prosthesis can be prescribed in which the ankle joint functions as a knee (Redrawn from: Tachdjian MO. Tachdjian's paediatric orthopaedics. 2nd ed. Philadelphia: WB Saunders Company; 1990)

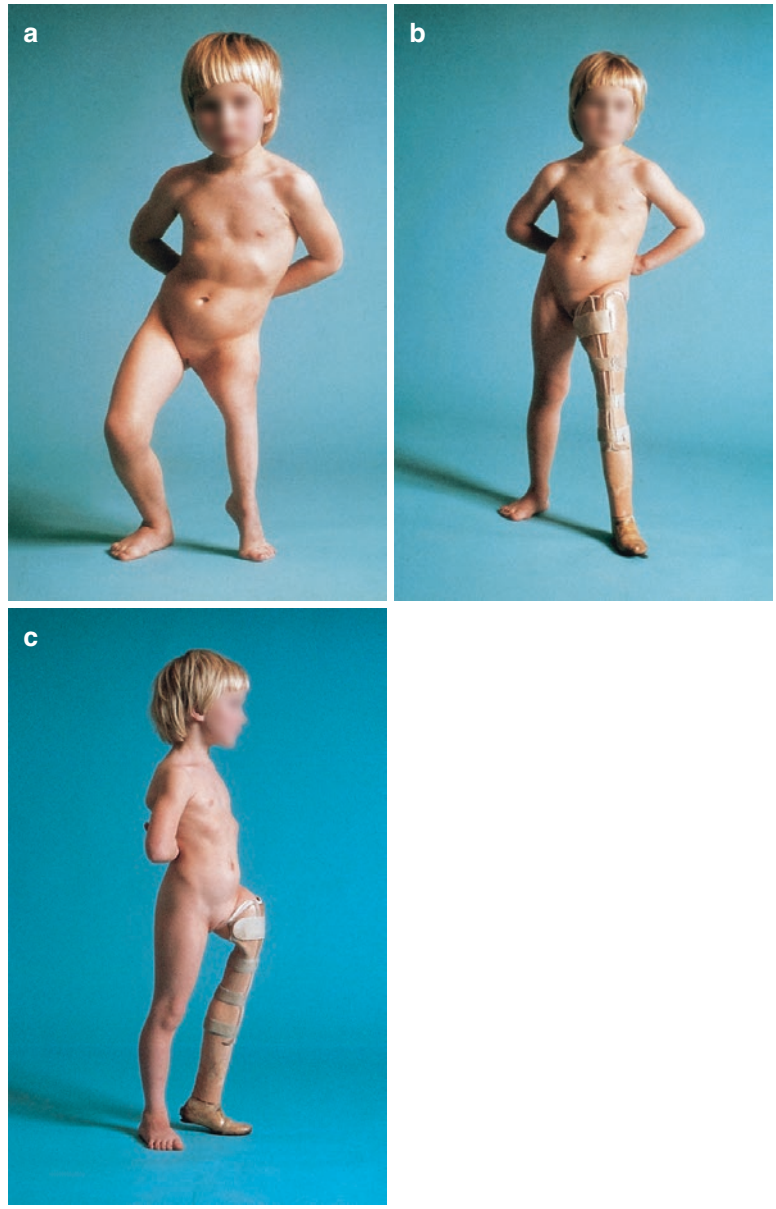



**Fig. 17.43** Child with a proximal focal femoral deficiency on the left side. (**a**) Without and (**b**) and (**c**) With a prosthesis. There is flexion in the proximal part of the femur

The leg is often in extreme external rotation. In order to reduce this a tenotomy of the sartorius muscle may be considered. There are no operative possibilities if only the distal part of the femur is present (Aitken IV). The appearance and the function of the prosthesis can be improved with several operative techniques if there is a substantial presence of the femoral part (Aitken I, II and III). A corrective osteotomy in the femur can be considered in the case of extreme flexion/abduction and external rotation in the thigh. A van Nes procedure may be considered if there is a stable hip (Aitken I). An arthrodesis of the knee with correction of the flexed position is advisable in the case of Aitken type II and III with a severe flexion contracture in the knee. The foot will sometimes be removed with a Syme amputation in order to allow the leg to fit into a nice cosmetic prosthesis. A Syme amputation is an amputation through the talocrural joint. The level of the amputation should be just above the contralateral knee joint in order to provide a good prosthetic function. The growth plates are left intact as much as possible (Fig. 17.44). Later on a possible

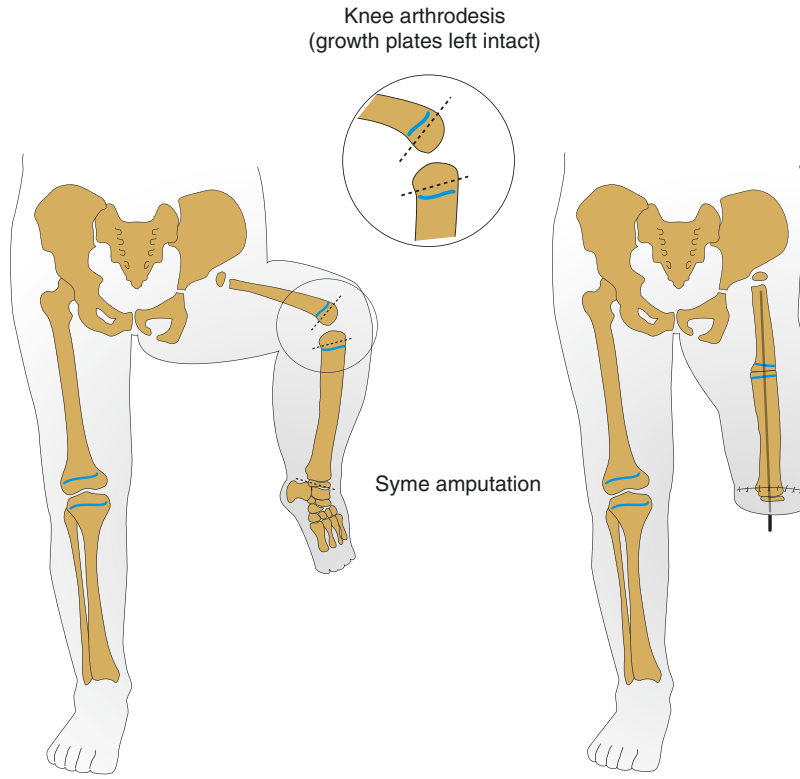



**Fig. 17.44** Knee arthrodesis and Syme amputation in a proximal focal femoral deficiency. The growth plates have been left intact (Redrawn from: Tachdjian MO. Tachdjian's paediatric orthopaedics. 2nd ed. Philadelphia: WB Saunders Company; 1990)

epiphysiodesis of one of the growth plates can be considered if the leg is still too long after the Syme amputation.

An arthrodesis of the proximal femoral stump onto the pelvis is contraindicated. This will otherwise cause back and knee problems in the future. Above all, a lengthening prosthesis in a mobile and unstable hip gives less problems than after an arthrodesis between the femur and pelvis.

In a bilateral proximal focal femoral deficiency the femurs are often equally short. These children walk at home with simple leg prostheses with tibial tuberosity support without extra lengthening. They usually use a wheelchair when outside.

## Abnormal Lower Leg and/or Foot

Complaint: there is a short lower leg and/or a foot deformity.

Assessment: apart from a short lower leg there is often a clubfoot with usually too many toes or a slight antecurvation in the shortened leg with a light valgus position, whereby the foot is in equinus and the outer two toes are as a rule absent or in which the middle toe or toes are absent or a foot in which one toe is absent.

Differential diagnosis:

Preaxial terminal and intercalary deficiency
- **tibial hypoplasia or aplasia**
  - type I
  - type II
  - type III
  - type IV
- **big toe aplasia**

Postaxial terminal and intercalary deficiency
- **fibular hypoplasia or aplasia**
  - type IA
  - type IB
  - type II
- **little toe aplasia**

Central deficiency
- **cleft foot (ectrodactylia, splitfoot, lobster-claw foot)**

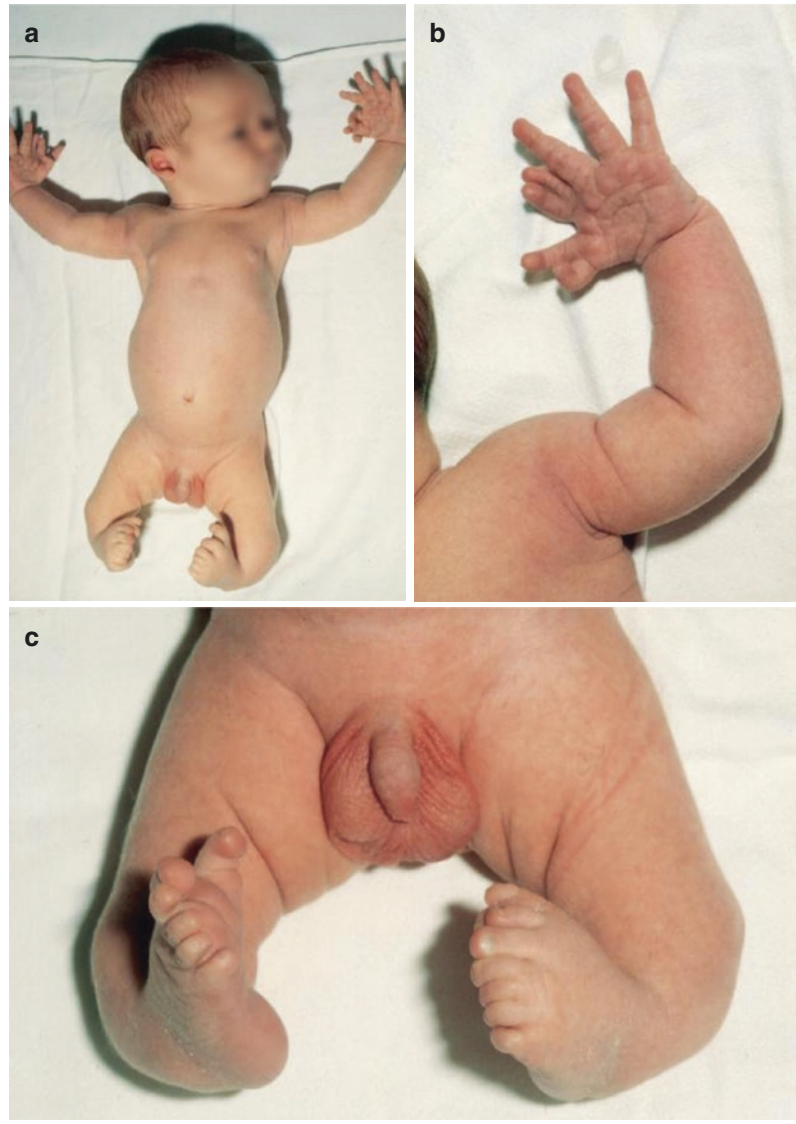



**Fig. 17.45** Bilateral tibial aplasia with polydactyly of the hands and the feet

Explanatory note: **paraxial terminal, paraxial intercalary and central deficiency.** In the classification of hypoplasia or aplasia of the tibia and fibula no distiction will be made between the presence of a complete foot or absence of a part of the foot. For a central deficiency we use the term cleft foot.

**Tibial hypoplasia and aplasia** This is a rare deformity which occurs in 1 in 1,000,000 life births. Multiple other deformities may be present and usually these are clubfeet and polydactyly of the hands and feet (Fig. 17.45). The deformity is divided into four types (Fig. 17.46).

*Type I* An X-ray taken directly after birth does not show a tibia (Fig. 17.47). This type is subdivided into two types.

*Type IA* In subtype IA the tibia is completely absent. The distal femoral epiphysis is at birth small and not visible on X-rays. There is no quadriceps function. There is a flexion contracture of the knee.

*Type IB* In this subtype the proximal part of the tibia is present but not ossified at birth. There is a normal ossification of the epiphysis in the distal part of the femur.

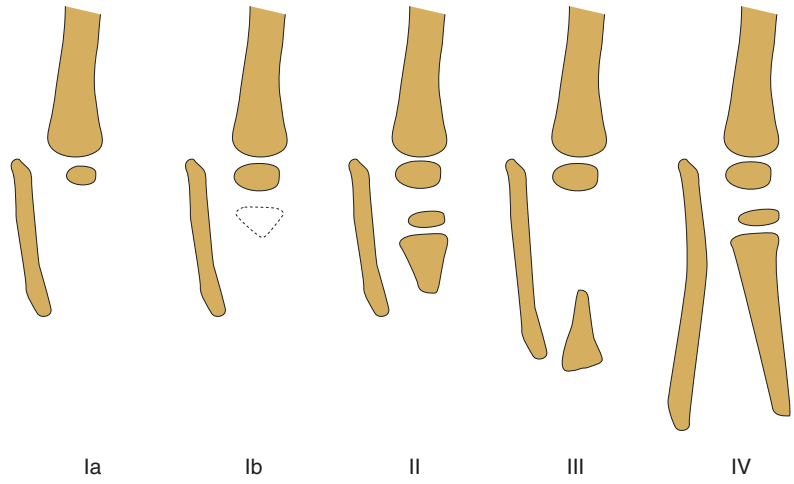



**Fig. 17.46** Radiological classification of tibial hypoplasia and aplasia modified by Jones, Barnes and Lloyd Roberts (1978). *Type IA*: tibia is completely absent. *Type IB*: the proximal part of the tibia is present but at birth is not yet ossified. *Type II*: the proximal part of the tibia is ossified directly after birth and can be visualized on X-rays. *Type III*: the proximal part of the tibia is absent, the distal part is present. *Type IV*: In this type the tibia is short and there is a tibiofibular diastasis in the distal part of the lower leg (Redrawn from: Jones E, Barnes J, Lloyd-Robers GC. Congenital aplasia and dysplasia of the tibia with intact fibula: classification and management. J Bone Joint Surg Br. 1978;60-B:31–9)

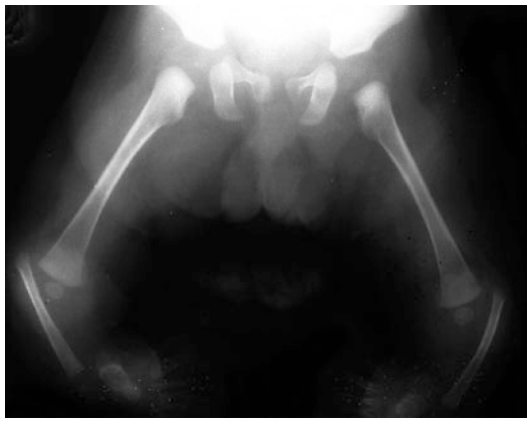


**Fig. 17.47** Bilateral tibial aplasia type IB. Same patient as in Fig. 17.45

*Type II* The proximal part of the tibia is ossified directly after birth and is therefore seen on X-rays. The distal part of the tibia is absent.

*Type III* The proximal part of the tibia is absent but the distal part is present. An unstable knee develops.

*Type IV* In this type the tibia is shortened and there is a diastasis between the tibia and fibula in the distal part of the lower leg. There is no ankle joint.

**Big toe aplasia** Absence of the big toe is only seen as a part of the Yunis Varon syndrome[1].

**Fibular hypoplasia and aplasia** An absence or partial absence of the fibula is the most common paraxial deficiency and occurs in 1 in 10,000 life births. The deformity can be divided into two types (Fig. 17.48). Type I is a fibula hypoplasia usually with a normal foot. In type II there is an aplasia or absence as a rule of the outer two or three rays. In both types there is a shortened tibia. The shortening is proportional to the degree of absence of the fibula. Consequently the shortening in type II is the most pronounced. In both types there is more or less in all cases a high patella localization (patella alta) with hypoplasia of the lateral femoral condyle in 75 % and in 50 % a genua valga.

[1] See Appendix.

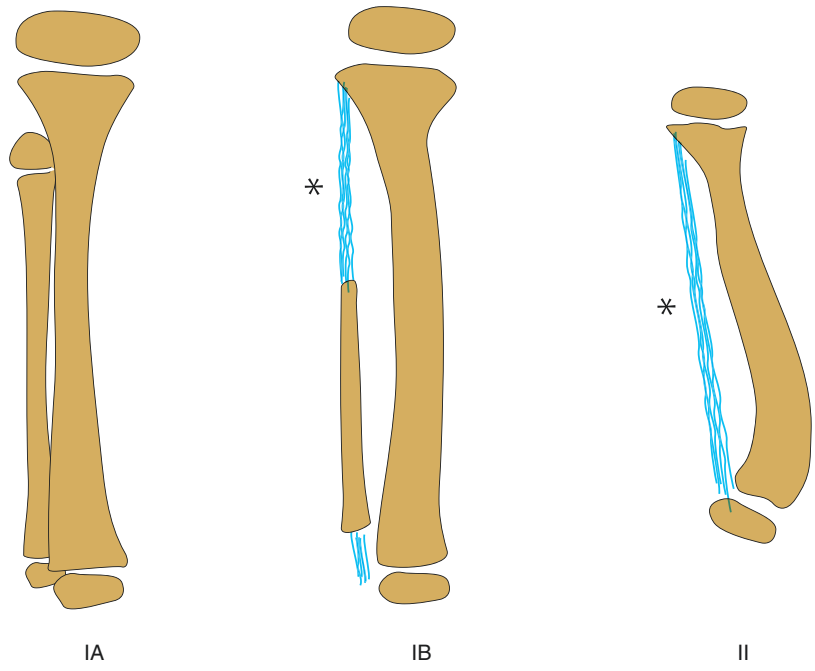



**Fig. 17.48** Classification of fibular hypoplasia and aplasia according to Achterman and Kalamchi (1979). *Type IA*: the fibula is shortened but the epiphysis is present in the proximal part and the distal part. *Type IB*: absence of the growth plates in the proximal and distal part of the fibula. *Type II*: the fibula is completely absent. * This shows a fibrous tissue remnant (Redrawn from: Achterman C, Kalamchi A. Congenital deficiency of the fibula. J Bone Joint Surg Br. 1979;61-B:133–7)

*Type I* Type I is unilateral. The fibula is shortened, the tibia is straight. As a rule there is no foot deformity. The lower leg is shortened and the ankle is in valgus to a greater or lesser extent (Fig. 17.49). This type is also divided into two subtypes.

*Type IA* In subtype IA the fibula is shortened but the epiphysis in the proximal and distal part are present.

*Type IB* In subtype IB the growth plates are absent in the proximal and distal part of the fibula. The fibula is consequently shorter than in subtype IA (30–50 % compared to normal).

*Type II* In type II the fibula is completely absent. There is a fibrous strand present as a remnant. The tibial shaft is not only severely shortened but may also have antecurvation and a light valgus with characteristic skin dimples at the level of the apex of the antecurvation and valgus deformity. The foot is in equinovalgus. The outward two or three rays of the foot are as a rule absent and there is often a tarsal coalition between talus and calcaneus. This type can also be bilateral (Fig. 17.50).

In most cases there is a shortened thigh in fibular hypoplasia and aplasia and in 15 % of cases there is a proximal focal femoral deficiency.

**Little toe aplasia** Absence of a small toe is practically always part of a hypoplasia or aplasia of the fibula.

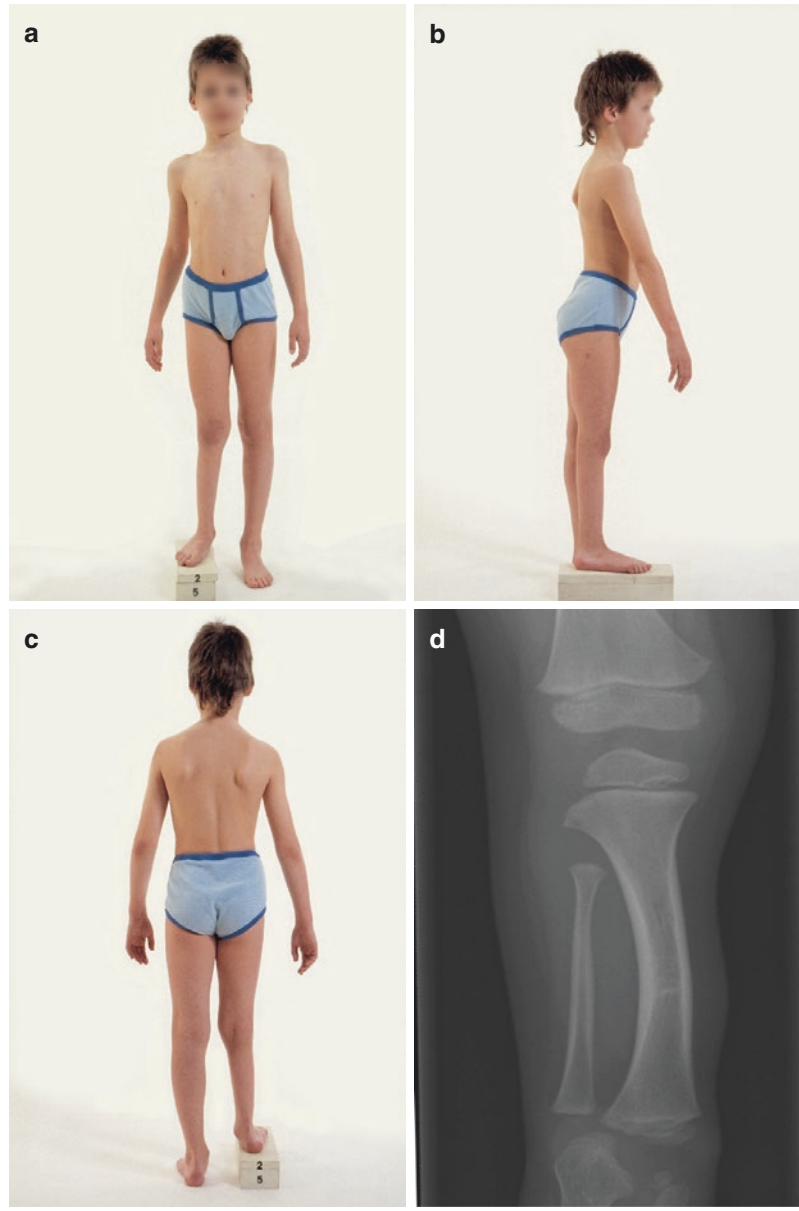



**Fig. 17.49** (**a**–**c**) Right fibular aplasia type IA. Shortening of the right lower leg. The right ankle is in valgus. (**d**) Anteroposterior X-ray of the right lower leg. Fibular aplasia type IA according to Achterman and Kalamchi

**Cleft foot** A cleft foot is seen in 1: 10,000 live births. Two or three central toes and their metatarsals are absent. The big toe on the medial side and the little or fourth and little toes on the lateral side appear like the claws of a lobster and are bent towards each other (Fig. 17.51). The deformity can be familial and is in that case bilateral and there will be lobster hands in 50 %. In non familial cases the deformity is mostly unilateral.

Supplementary assessment: X-rays of the whole lower extremity. On standard X-rays in a newborn with type I tibial aplasia it is not possible to see if the proximal part of the tibia is present or absent because the structures are cartilagenous and not yet ossified. An MRI is the best solution.

Primary care treatment: referral to a pediatric orthopedic surgeon and pediatric rehabilitation

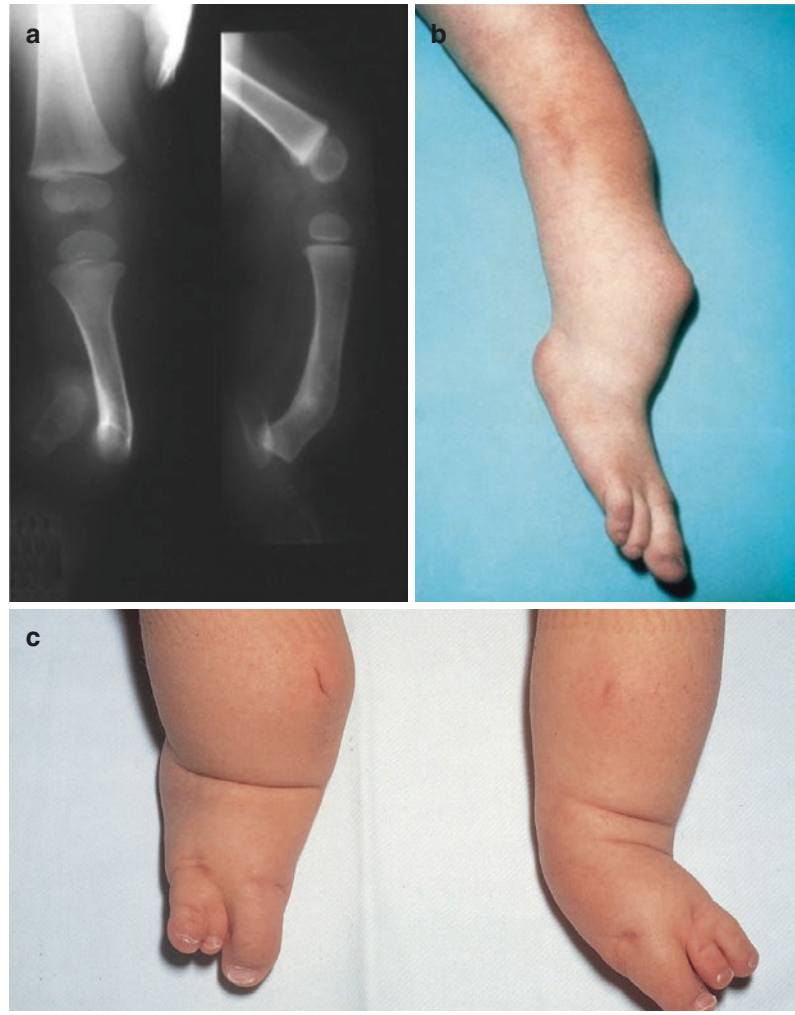



**Fig. 17.50** Bilateral fibular aplasia type II. (**a**) Antecurvation and valgus deformity in the right tibia. (**b**) The right foot is in equinovalgus. (**c**) The two outside rays of the feet are absent

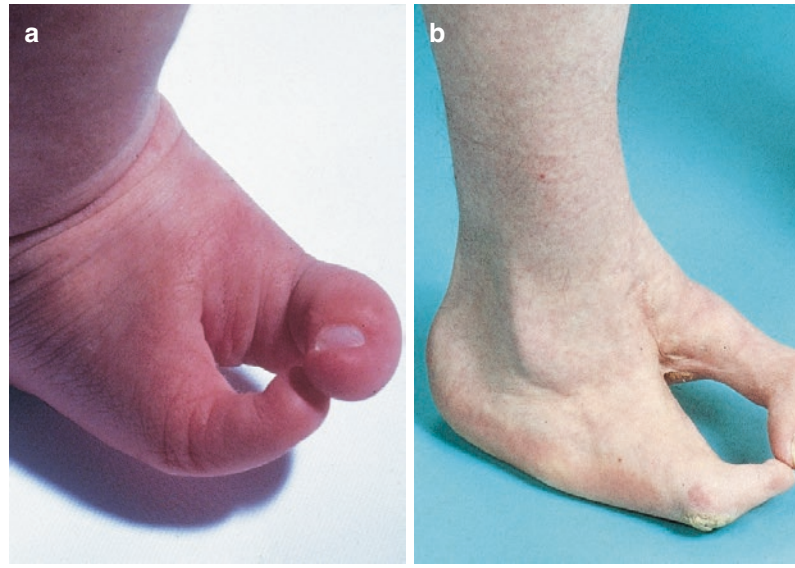



**Fig. 17.51** Cleft foot. (**a**) In a newborn. (**b**) In an adult

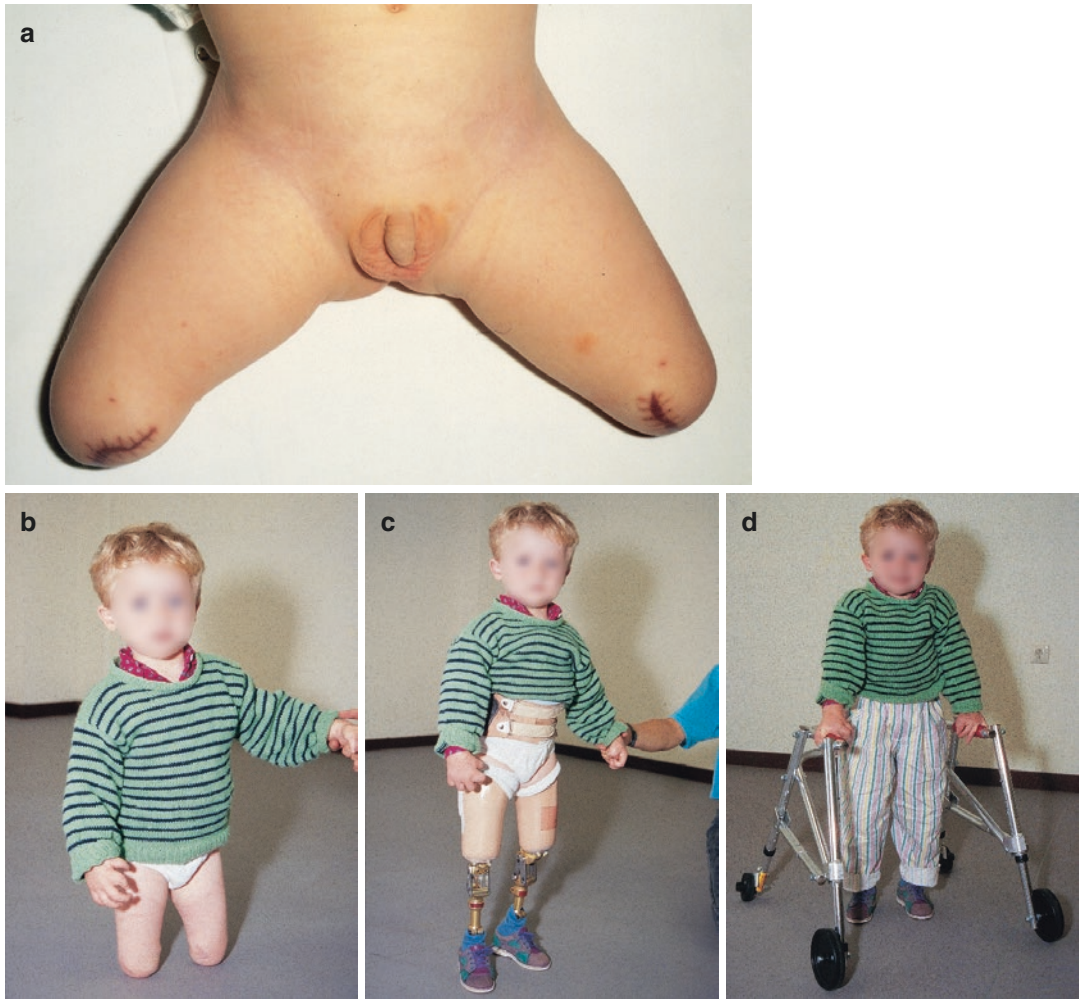



**Fig. 17.52** (**a**, **b**) Bilateral exarticulation at the level of the knee in type IA tibial aplasia. (**c**, **d**) Mobilization with knee exarticulation prostheses. Same patient as in Figs. 17.45 and 17.47

specialist. In the case of cleft feet the parents should be informed that the child can usually walk well and operations are usually unnecessary. Generally normal shoes can be worn which do not cause any problems when standing or walking.

When to refer: all paraxial deficiencies in the first 6 months of life.

Secondary care treatment: **tibial hypoplasia and aplasia.**

**Type IA** An exarticulation at the level of the knee is indicated in cases with complete absence of the tibia, preferably before the age of 10 months because the psychological aspects are less of a problem in an infant than in older children and the child can learn to walk with prostheses just as in normal children of the same age (Fig. 17.52).

**Type IB** The proximal part of the fibula is fixed to the proximal part of the tibia. This operation can only be carried out after the proximal part of the tibia has ossified. The foot will be amputated (Fig. 17.53). The child gets a lower leg prosthesis after this. A knee exarticulation is carried out if there is a serious flexion contracture in the knee.

**Type II** The same treatment as in type IB.

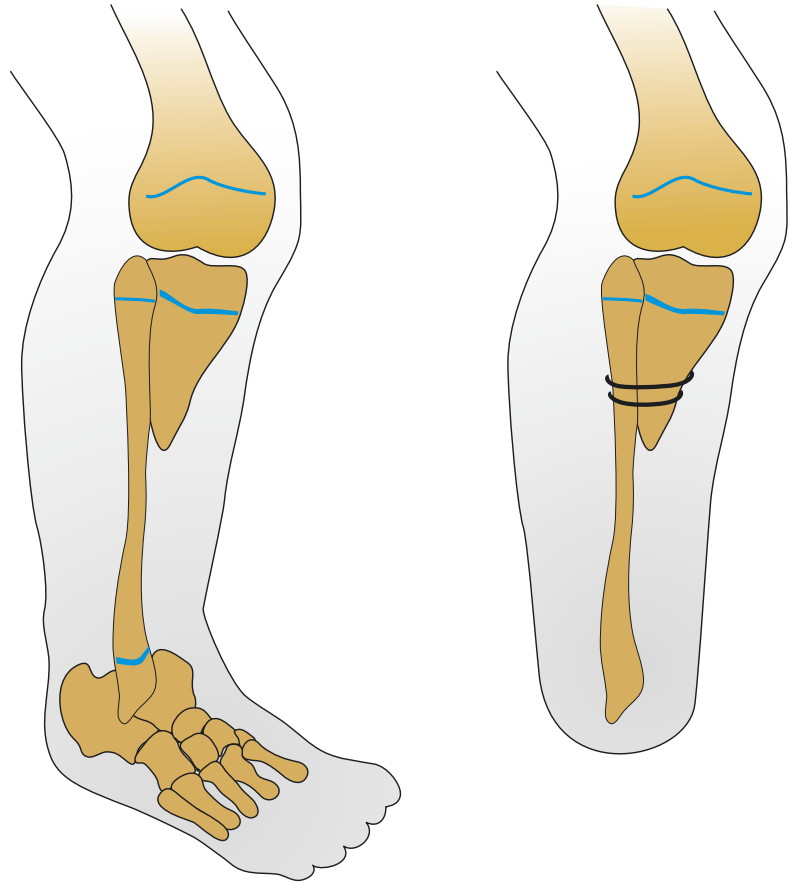


**Fig. 17.53** In type IB and II tibial hypoplasia. The proximal part of the fibula will be fixed to the proximal part of the tibia and a Syme amputation will be carried out

**Type III** There is little experience with this type. As a rule there is limited knee function. The distal part of the fibula will be fixed to the distal part of the tibia (Fig. 17.54). A lengthening prosthesis with an upper leg cuff and knee hinges is sufficient if the ankle function and the foot function are adequate. A Syme amputation will be carried out if there is insuf-ficient ankle and foot function, after which the patient gets a prosthesis. A Syme amputation is an amputation through the talocrural joint. An exarticulation at the level of the knee may be considered if there is no knee function in this type.

**Type IV** The most reasonable option is a Syme amputation if the ankle is absent.

**Fibular hypoplasia en aplasia. Type 1** Hereby the leg length difference and slight instability in

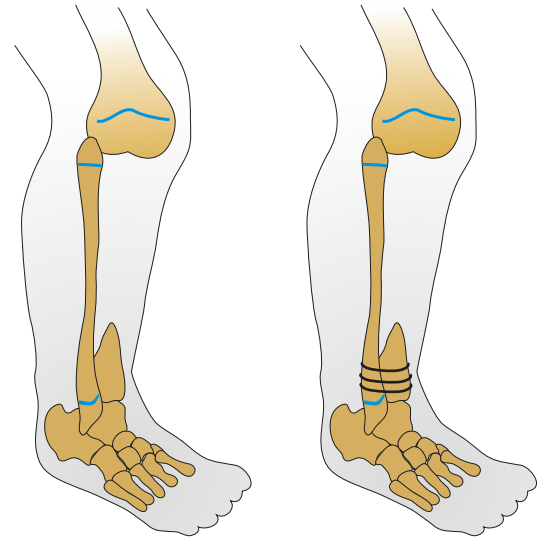


**Fig. 17.54** In a type III tibial hypoplasia one tries to preserve ankle function by fixing the fibula to the distal part of the tibia (Figures 17.54 and 17.55, redrawn from: Canale ST, Beaty JH. Operative pediatric orthopaedics. St. Louis: Mosby Year Book; 1991)

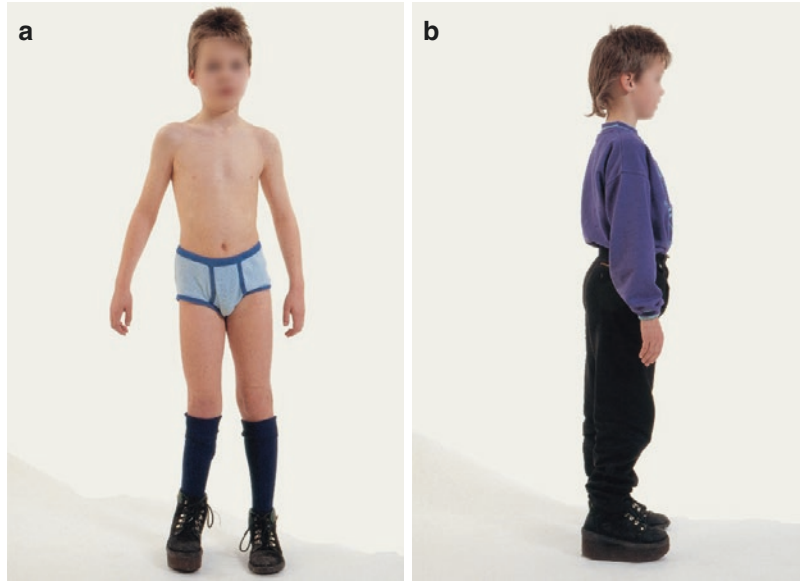



**Fig. 17.55** Shoe adaptation with heel sole elevation in right sided fibular hypoplasia type I

the ankle are compensated by a high shoe with heel and sole elevation (Fig. 17.55). An epiphysiodesis in the proximal part of the contralateral tibia and fibula can be considered when the child is older if the leg length difference is between 2 and 5 cm. If the leg length difference is more than 5 cm then the shortened leg can be operatively lengthened (see pp. 328 and 331).

**Type 1A** The valgus in the ankle in type 1A can be treated with a temporary epiphysiodesis of the medial malleolus or a supramalleolar varus osteotomy (Fig. 17.56).

**Type 1B** In type IB the equinovalgus position of the foot can be corrected by dividing the fibula connective tissue strand, lengthening the achilles tendon and reconstruction of the lateral malleolus according to Gruca (Fig. 17.57). If the leg length difference is more than 30 % of the normal leg the child can be provided with a leg lengthening orthosis with the foot in equinus (Fig. 17.58). In that case a Syme amputation can also be considered.

**Type II** A Syme amputation can be carried out in both unilateral and bilateral types at the age of 10 months. If the antecurvation is 40° or more, then a tibial correction osteotomy can be considered at the same time. An antecurvation of less than 40° usually corrects itself spontaneously as a rule with an acceptable position. The child can learn to walk with a lower leg prosthesis after this. A valgus position occurs in the knees in 50 % of cases. This can be corrected between 11 and 12 years of age by carrying out a temporary hemi-epiphysiodesis on the medial side of the growth plates in the distal part of the femur. The clamps will be removed when the knees are straight (Fig. 17.59). The length of the prostheses must of course be modified during growth. After a bilateral Syme amputation one should make sure that the prostheses have sufficient length so that the body proportions remain normal. After a bilateral Syme amputation children can walk small distances without prostheses.

In the past one tried to avoid amputations. However, it was almost impossible to keep the feet directly under the lower leg. The lower legs are generally severely shortened such that the body proportions are not normal and the child will not grow much taller than 150 cm (Fig. 17.60).

**Little toe aplasia** Orthopedic shoes.

**Ectrodactylia** Orthopedic shoes if necessary. One may consider approximating the seriously diverted rays in order to make it easier to modify the shoes.

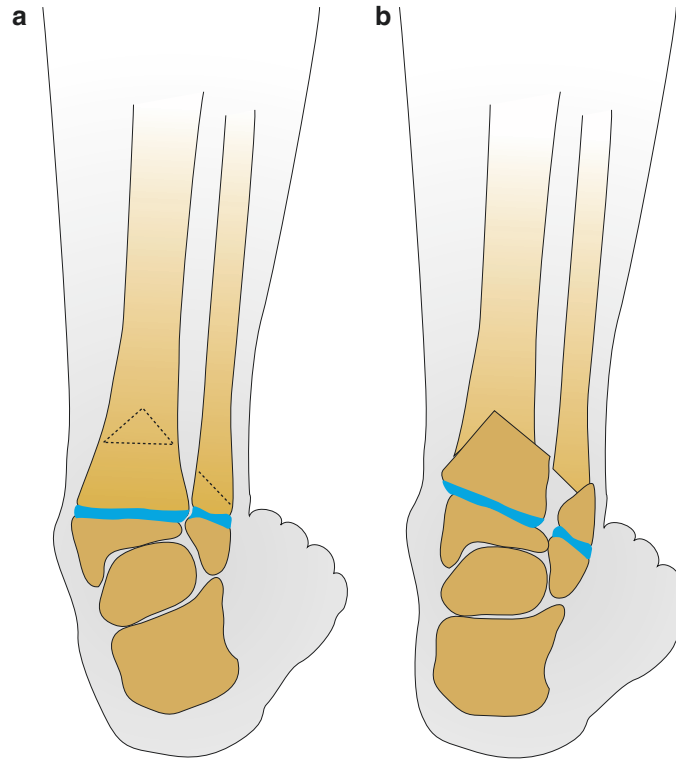



**Fig. 17.56** Supramalleolar correction osteotomy for the valgus deformity in the ankle

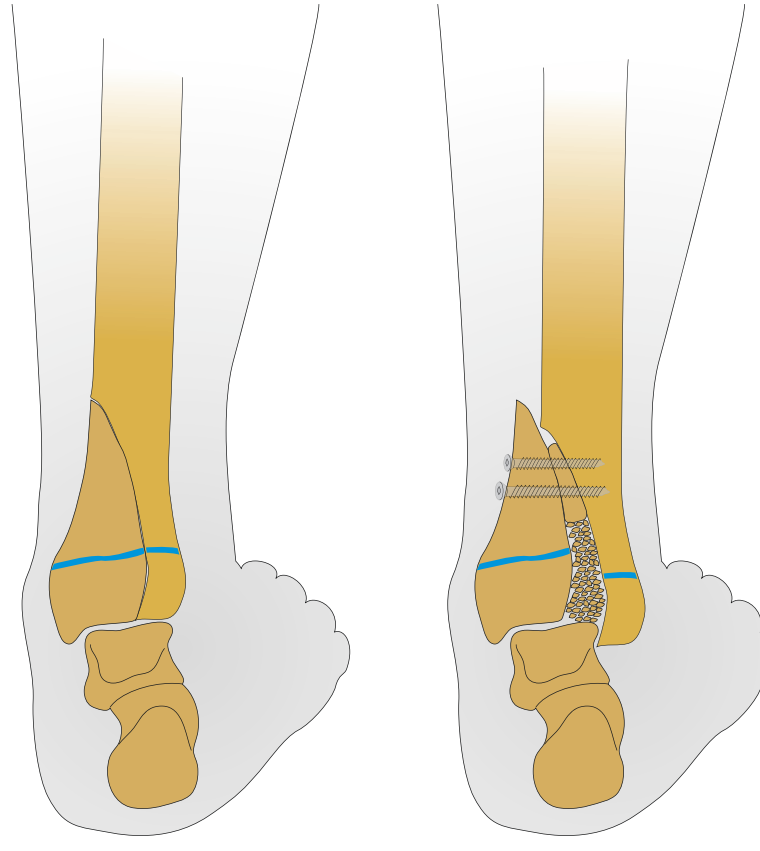


**Fig. 17.57** Gruca procedure (Redrawn from: Serafin J. A new operation for congenital absence of the fibula. Preliminary report. J Bone Joint Surg. Br. 1967;49:59–65)

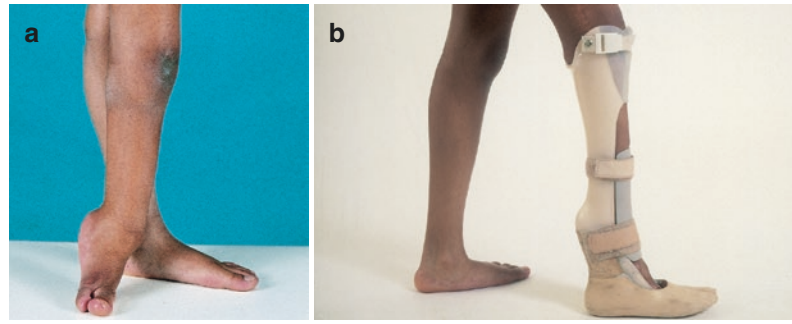



**Fig. 17.58** (**a**) Fibular aplasia. (**b**) Lengthening prosthesis with the foot in equinus in fibular aplasia

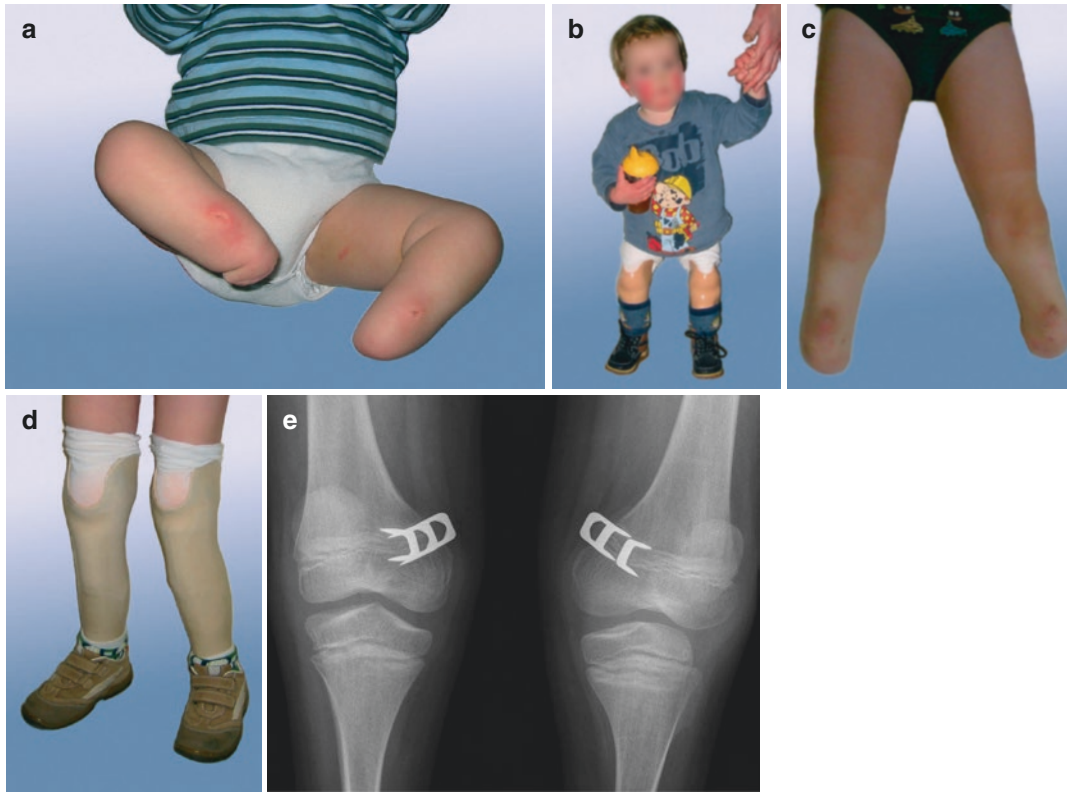



**Fig. 17.59** (**a**) Bilateral Syme amputation. (**b**) Prostheses after bilateral Syme amputation. (**c**, **d**) A valgus position develops in 50% of cases. (**e**) This can be corrected between 11 and 12 years of age by a temporary hemiepiphysiodesis on the medial side of the growth plates in the distal part of the femur. The clamps can be removed once the knees are straight

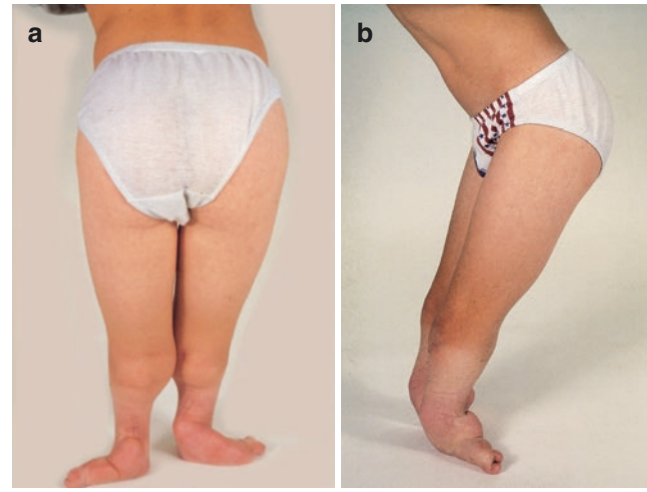



**Fig. 17.60** (**a**) and (**b**) Situation after attempting to reposition the feet directly under the lower legs in bilateral fibular aplasia. The lower legs are severely shortened. A bilateral Syme amputation was finally carried out

### Differential Diagnosis Complete or Partial Absence of a Leg

| **Absence of the whole or part of the leg** | **Transverse terminal deficiency** |
|---|---|
| Absence of the whole leg. | **Amelia** |
| Absence of the foot, lower leg and part of the upper leg. | **Incomplete amelia** |
| Absence of the foot and lower leg. | **Hemimelia** |
| Absence of the foot and part of the lower leg. | **Incomplete hemimelia** |
| Absence of the foot. | **Apodia** |
| Absence of all the toes and metatarsals. | **Complete adactylia** |
| Absence of all of the phalanges over the complete breadth of the foot. | **Complete aphalangia** |
| **Absence of the whole leg, upper or lower leg between the trunk and the foot, in which case the knee joint is always absent** | **Transverse intercalary deficiency (phocomelia)** |
| Absence of the thigh and lower leg. The foot is fixed to the pelvic girdle. | **Complete phocomelia** |
| Absence of the thigh. The lower leg is fixed to the pelvic girdle. | **Proximal phocomelia** |
| Absence of the lower leg. The foot is fixed onto the thigh. | **Distal phocomelia** |
| **Short upper leg. the knee joint is always present** | |

| | |
|---|---|
| The foot and the shortened upper leg are at a higher or more distal level than in the normal contralateral leg. | |
| The hip is present. | **Congenital short femur (femoral hypoplasia)** |
| The foot and the shortened thigh are at the level of the knee on the normal contralateral side. | |
| The proximal femoral part is deficient. | **Proximal focal femoral deficiency** |
| **Abnormal lower leg and/or foot** | **Paraxial terminal- and intercalary deficiency** |
| Absence or partial absence of the tibia. | **Tibial hypoplasia or aplasia** |
| The tibia is completely absent. | Type IA |
| Only the proximal part of the tibia is present and is not ossified at the time of birth. | Type IB |
| Only the proximal part of the tibia is present and is ossified at the time of birth. | Type II |
| The proximal part of the tibia is absent, but the distal part is present. | Type III |
| There is a tibiofibular diastasis in the distal part of the lower leg. There is no ankle joint. | Type IV |
| Absence of the big toe. | **Big toe aplasia** |
| Absence or partial absence of the fibula | **Fibular hypoplasia or aplasia** |
| The fibula is shortened but the proximal and distal epiphysis are present. The tibia is straight. | |
| Valgus position in the ankle. Normal foot. | Type IA |
| The fibula is shortened, the proximal and distal growth plates are absent. | |
| The tibia is straight. Valgus position in the ankle. Normal foot. | Type IB |
| The fibula is completely absent. Antecurvature and valgus position of the tibia. Equinovalgus position in the foot. | |
| The lateral 2 or 3 rays of the foot are absent. | Type II (this type can also be bilateral) |
| Absent toes | |
| Absence of the little toe. | **Little toe aplasia** |
| Cleft foot or absence of central toes | **Central deficiency** |
| Cleft foot or one or more absent central rays of the foot. | **Cleft foot (ectrodactylia, split foot, lobster claw foot)** |
| V-shaped defect. | Typical type |
| U-shaped defect. | Atypical type |

# Back

## Buddhist Posture

Complaint: directly after birth the legs are immobile. They lie in a Buddhist posture.

Assessment: on examination anomalies are found varying from only positional deformities of the feet and hip dislocations but also there can be a flexion abduction contracture in the hips and a flexion contracture of the knees up to 50° or 60° with paralysis and atrophy of the lower extremity which is complete below the knee (Fig. 17.61).

Differential diagnosis:

**Sacral agenesis (lumbosacral agenesis, caudal regression syndrome)**
- Type I
- Type II
- Type III
- Type IV

Explanatory note: **sacral agenesis.** The coccyx, sacrum and lumbar vertebrae can be completely or partially absent. The most usual term for a caudal absence of the vertebrae is sacral agenesis (a means without and genesis is occurrence in Greek). The terms vertebral agenesis, lumbosacral agenesis and caudal regression syndrome are also used. The anomalies that are found in sacral agenesis depend on the extent in which the vertebrae and accompanying motor nerves are absent.

Sacral agenesis is divided in four types (Fig. 17.62). These are very rare anomalies in which type II is the most frequent and type I the least frequent.

The incidence is 1–2.5 per 100,000 newborns.

*Type I (unilateral sacral agenesis)* The sacrum is partially or completely absent on one side. In this type the hips and knees are usually normal but sometimes there are postural deformities of the feet, i.e. clubfeet and a pes convex valgus (congenital talus verticalis). These children are able to walk.

*Type II (partial sacral agenesis)* The first sacral vertebra is present but the rest of the sacrum and the coccyx are absent. The underdeveloped (hypo-plastic) first sacral vertebra creates a stable joint with the iliac bones. There is a unilateral or bilateral hip dislocation as a result of the disturbed balance between the hip abductors and adductors whereby the hip abductors are weakened and the adductors are normal. Flexion

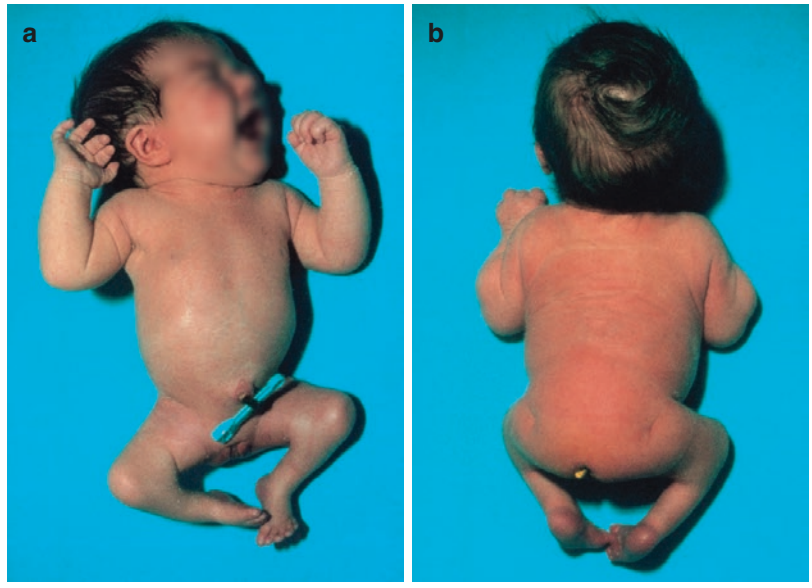



**Fig. 17.61** Sacral agenesis in a newborn

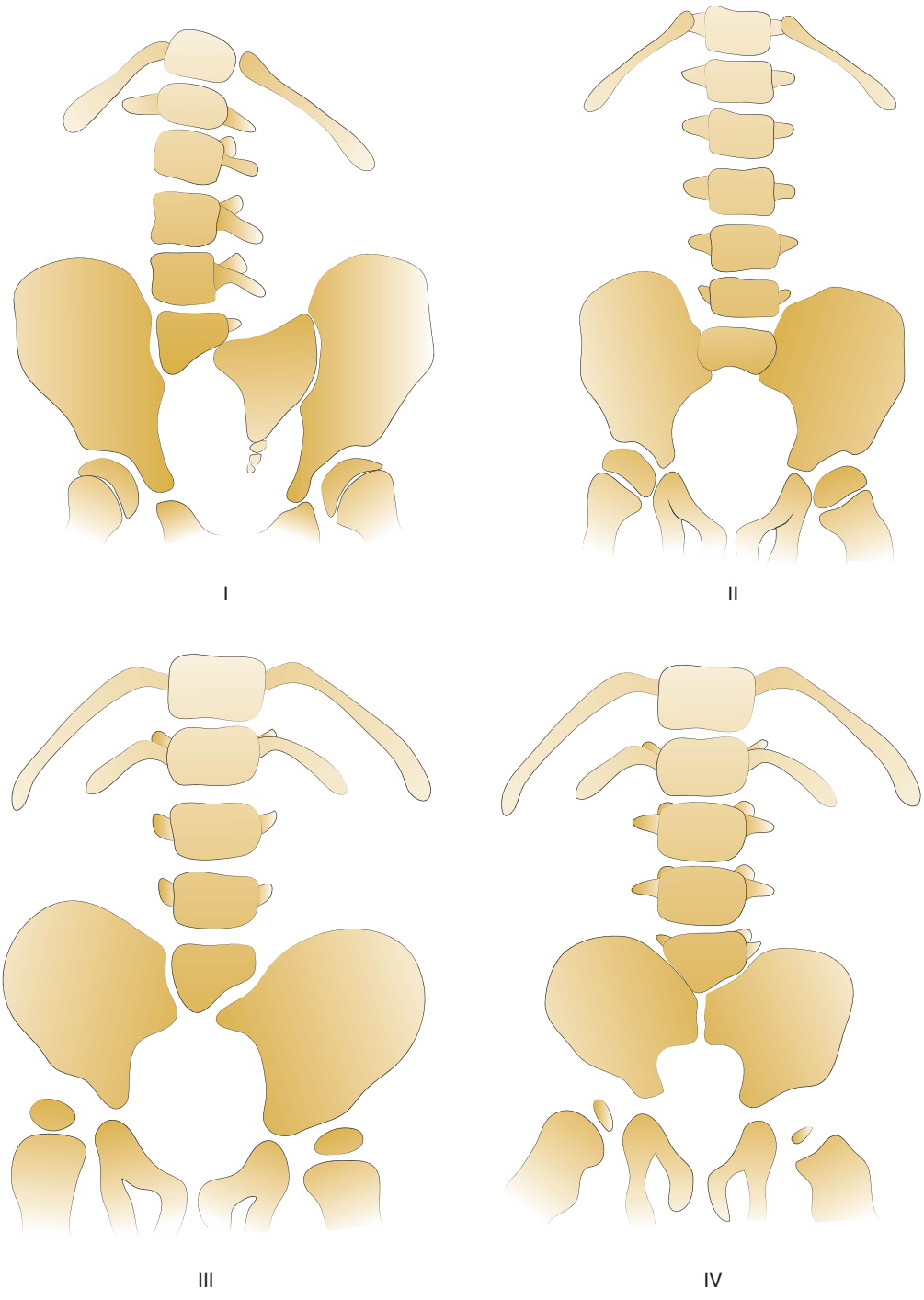



**Fig. 17.62** Sacral agenesis can be divided into four types (Renshaw 1978). *Type I*: unilateral sacral agenesis. The sacrum is partially or completely absent on one side. *Type II*: partial sacral agenesis. The first sacral vertebra is present but the rest of the sacrum and coccyx are absent. The underdeveloped (hypoplastic) first sacral vertebra has a stable joint with the iliac bones on both sides. *Type III*: lumbosacral agenesis. The coccyx, sacrum and a variable number of lumbar vertebrae are absent. The lowest lumbar vertebra articulates with the iliac bones. *Type IV*: lumbosacral agenesis with fusion or amphiarthrosis of the iliac bones (Redrawn from: Renshaw TS. Sacral agenesis. J Bone Joint Surg Am. 1978; 60-A:373–83)

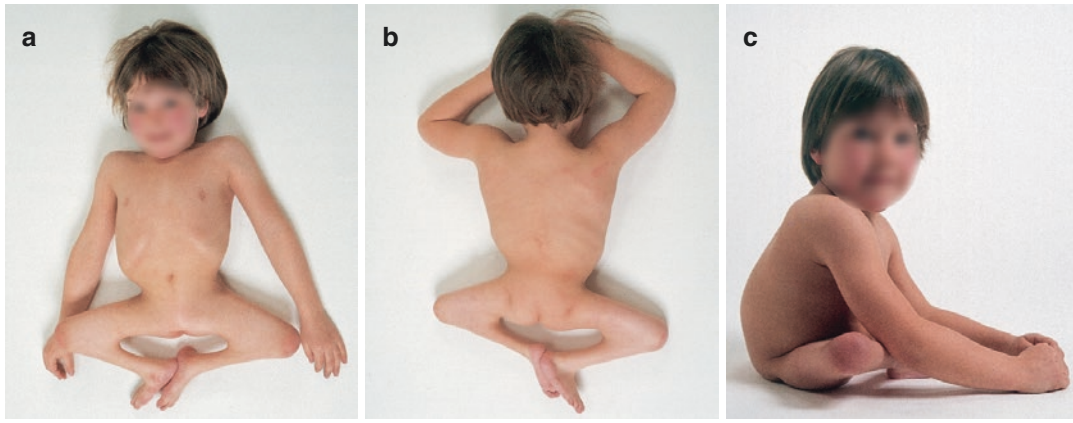



**Fig. 17.63** (**a**–**c**) Sacral agenesis type IV at 7 years of age (Buddhist posture)

contractures of the knees and the foot deformities are usually not prominent. There is reduced sensation below S1. Most children with a type II sacral agenesis can usually walk without support.

*Type III (lumbosacral agenesis)* The coccyx, the sacrum and a variable number of lumbar vertebrae are absent. The lowest lumbar vertebra articulates with the iliac bones. Hip dislocation, flexion contractures of the knees and postural deformities of the feet are present as a rule. There is no sensation below S1. Patients with a type III sacral agenesis are not capable of standing and walking without splints or crutches.

*Type IV (lumbosacral agenesis with fusion or amphiarthrosis of the iliac bones)* The coccyx and sacrum are completely absent.

The iliac bones are fused together or there is a joint between these bones (amphiarthrosis).

The child sits in a so-called Buddhist posture because of the flexion and abduction contracture of the hips and the flexion contracture in the knees up to 50° or 60° (Fig. 17.63). These children have problems with micturition and defecation. They are completely dependent on a wheelchair.

Normal muscle tissue is replaced with yellow fat, the tendons have a normal configuration but are thin if the lumbosacral vertebral column is completely absent. The blood vessels are very small and the motor nerves in the legs are absent. The sensory nerves in the legs are present but however, there is no sensation distal to S1.

Usually there is a stable connection between the spine and the pelvis in type I and II. This is not the case in types III and IV and in these cases it is possible that progressive kyphosis develops. Motor nerves may be absent, depending on the size of the defect. In types II, III and IV disturbances in sensation may be present distal to S1. A scoliosis occurs frequently. There is no correlation between the type of sacral agenesis and the incidence of scoliosis. Other anomalies can be present such as: hemivertebrae, spina bifida and Arnold Chiari syndrome. The Arnold Chiari syndrome is a situation in which the brainstem and cerebellar tonsils are displaced caudally with herniation through the foranum magnum and they usually have a hydrocephalus. A sacral agenesis can also be part of the VATER or VACTERL association[2].

Supplementary assessment: anteroposterior and lateral X-rays of the vertebral column, anteroposterior X-ray of the pelvis (Fig. 17.64) and an intravenous pyelogram.

Primary care treatment: none.

When to refer: a sacral agenesis must always be referred.

[2]See Appendix.

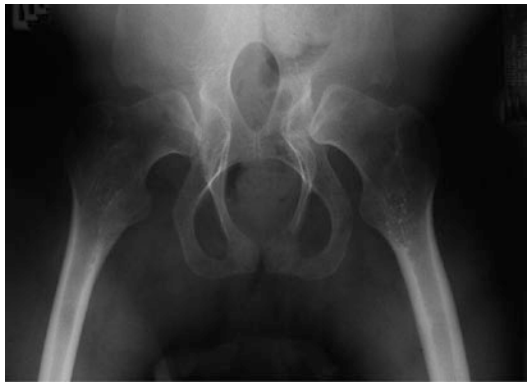


**Fig. 17.64** Sacral agenesis type IV. Fusion of the iliac bones

Secondary care treatment: **unilateral and partial sacral agenesis (type I and II).** In relation to starting to walk type I and type II have an excellent prognosis. Possible hip dislocations, contractures and foot deformities must be treated.

**Lumbosacral agenesis (type III and IV)** Walking is not possible in type III and IV. Sometimes the child is not able to sit because of instability between the vertebral column and the pelvis which is an indication for operative fixation of the vertebral column onto the pelvis.

**Differential Diagnosis in Sacral Agenesis (Lumbosacral Agenesis, Caudal Regression Syndrome)**

| | |
|---|---|
| Postural anomalies of the feet, otherwise apparently normal | **Type I: unilateral sacral agenesis** |
| Unilateral or bilateral hip dislocation and reduced sensation below S1 | **Type II: partial sacral agenesis** |
| Hip luxations, flexion contracture in the knees, postural anomalies in the feet and loss of sensation below S1 | **Type III: lumbosacral agenesis** |
| Buddhist posture and loss of sensation below S1 | **Type IV: lumbosacral agenesis with fusion or amphiarthrosis of the iliac bones** |

# Bone and Joint Infections 18

## Extremely Painful Immobile Extremity

Complaint: the child cannot move the involved extremity and has a lot of pain.

Assessment: a newborn obviously cannot point to where the pain is localized. What is conspicuous is that the involved extremity is not mobile and feels warmer around the focus of infection. If the bone or joint lies just under the skin, then there is also redness. There is severe local pressure pain at the level of the involved area. There can be an abnormally forced position in the adjacent or involved joint. An older child will refuse to use the arm or leg and will in the case of a leg certainly not stand on it or walk. Older children are usually febrile.

Differential diagnosis:

**acute osteomyelitis**
**septic arthritis** (**bacterial-**, **pyogenic-**, **suppurative arthritis**)

Explanatory note: **acute osteomyelitis and septic arthritis**. A distinction must be made between children younger than 9–18 months and older children.

A child younger than 9–18 months is mostly not ill and does not have a temperature. This is due to an under-developed defense system with as a rule gives hardly any reaction to the infection.

In newborns an acute osteomyelitis can occur in more locations at the same time. In those cases there is sufficient stimulation of the defense system and the child will be febrile.

Older children as a rule are ill and are febrile.

The WBC, BSE and CRP are as a rule normal in children younger than 9–18 months of age but are raised in older children. In acute osteomyelitis or septic arthritis in more areas these parameters will be raised not only in newborns but also in older children.

On examination an acute osteomyelitis cannot be distinguished from a septic arthritis. This is partly because a reactive hydrops in the adjacent joint can also be caused by an acute osteomyelitis. Furthermore, the involved or adjacent joint has an abnormal forced position in a septic arthritis and also often in an acute osteomyelitis. The hip will be in flexion, abduction and external rotation (Fig. 18.1) and the knee in 30° of flexion (Bonnet position). Joint aspiration and culture are often necessary to distinguish between an acute osteomyelitis and a septic arthritis. An untreated acute osteomyelitis will result in a chronic osteomyelitis and an untreated septic arthritis may result in a deformity of the joint, or even in resorption of the femoral head.

- Subacute osteomyelitis (Brodie abscess) (see pp. 222–226 and 228)
- Gonococcal arthritis (see p. 220)
- Bone and joint tuberculosis (see pp. 221, 222)
- Spondylodiscitis (see pp. 45, 48, 49, 51)
- Tuberculous spondylitis (see pp. 51, 52)


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2_18

## Incidence

An acute osteomyelitis or septic arthritis can occur at all ages but usually involves children under 10 years of age. It is more frequent in boys than in girls. The incidence is 5–20 per 100,000 children per year.

## Pathogenesis of Acute Osteomyelitis

A bone infection occurs because bacteria are spread hematologenously (bacteremia) or lymphogenously to the bone from a source of infection elsewhere in the body. These infections can be from the ear, throat, airways, intestines and urinary passages, sometimes skin infections after chickenpox or infection after a direct wound i.e. due to a nail in the foot and after diverse operations. The focus is usually not known. Bacteremia in children is a daily occurrence for example, there is a bacteremia in 50 % of cases after brushing the teeth.

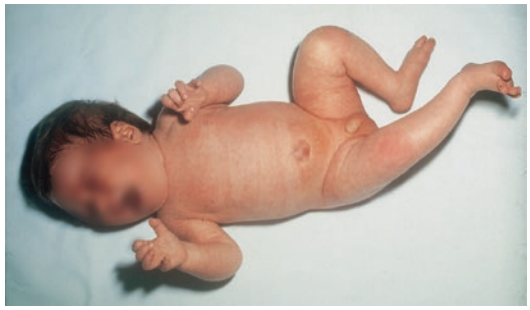


**Fig. 18.1** Newborn with an infected left hip joint. The hip is in flexion, abduction and external rotation

The parts of the metaphysis, particularly those close to the epiphyseal plate, have an excellent vascularisation. The nutrient artery that is responsible for the vascularisation splits up in the bone marrow into small arterioles which run towards the growth plate. Just before the growth plate they turn around and end up in a venous cavity (sinus), which drains into the marrow cavity (vascular loops). Infection starts where the arterioles turn around (Fig. 18.2). As a result of a reduction in blood circulation the bacteria accumulate here. In children an acute hematogenous osteomyelitis begins in the metaphysis, never in the epiphysis or diaphysis. Some authors suggest that an injury (i.e. a contusion) may play a role in acute hematogenous osteomyelitis in 30–40 %. In the first instance a cellulitis occurs. In the cellulitis stage clinical symptoms and signs may be present such as pain, redness, swelling, joint mobility loss and possibly fever and general malaise.

If an acute hematogenous osteomyelitis remains untreated, then a subperiosteal abscess

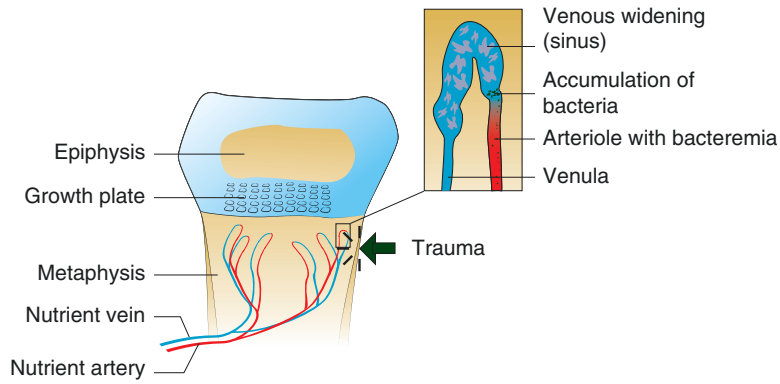



**Fig. 18.2** In otherwise normal children an acute hematogenous osteomyelitis occurs where the vascular loops are situated in the metaphysis. The bacteria accumulate here because of the reduction in blood circulation

will be present after several days. The periosteum will be raised up due to the accumulation of pus (Fig. 18.3).

## Pathogenesis of Septic Arthritis

- A septic or infectious arthritis often occurs hematogenously:
- As a result of a direct hematogenous infection of the synovium (joint capsule) or the synovial fluid (Fig. 18.4).
- In newborns an acute osteomyelitis that starts in the metaphysis can also spread into the epiphysis and then into the joint. In newborns the long bones do not have a bone nucleus in the epiphysis and the blood vessels run from the metaphysis into the epiphysis, the so-called transphyseal vessels. Through these vessels an infection can spread from the metaphysis into the epiphysis and finally into the joint. This causes total destruction of the growth plate and the epiphysis and finally the whole joint. After the appearance of the bony nucleus in the epiphysis transphyseal blood vessels disappear and the epiphysis and metaphysis have a separate blood supply. As a result an acute osteomyelitis in older chil-

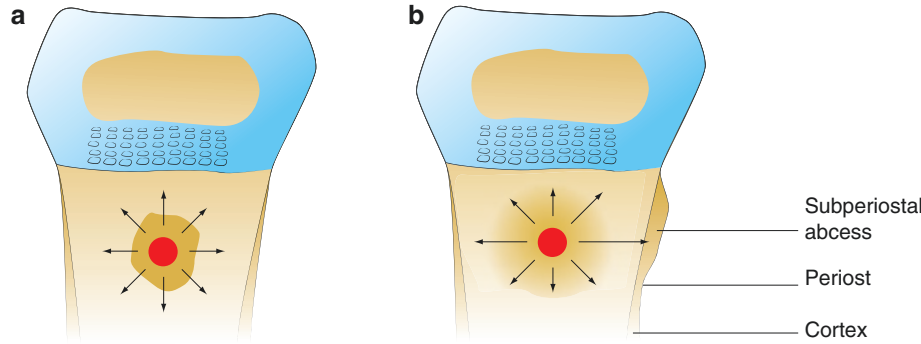



**Fig. 18.3** (**a**) An acute hematogenous osteomyelitis in the metaphysis. (**b**) If this is not treated a subperiosteal abscess will develop. The periosteum will be raised up as a result of the accumulation of pus

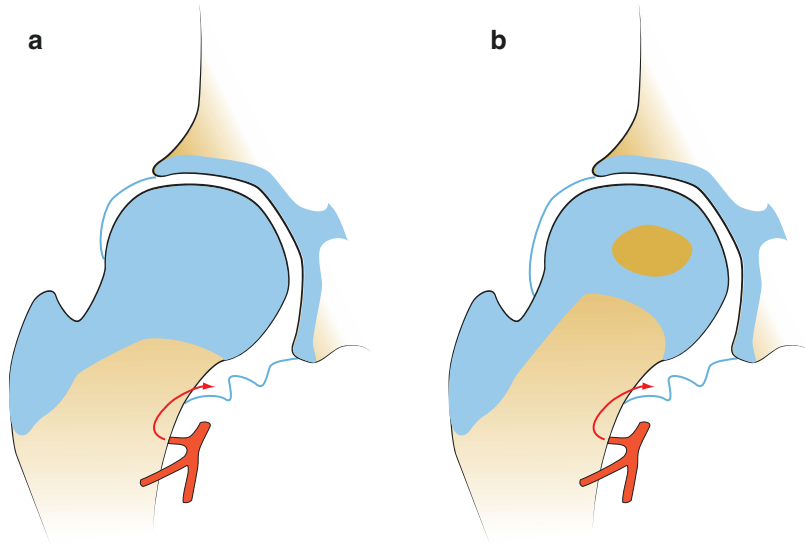



**Fig. 18.4** A septic arthritis can be the result of a direct infection of the synovium not only in newborns (**a**) but also in older children (**b**)

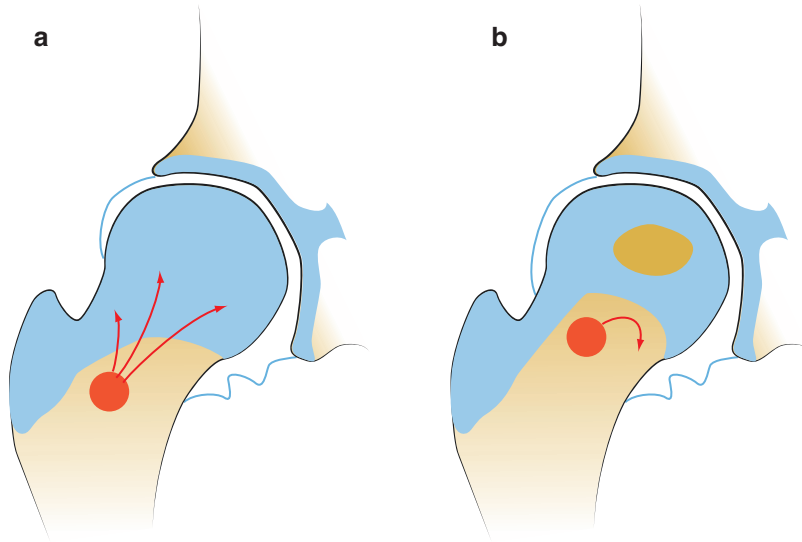



**Fig. 18.5** (**a**) In a newborn an acute hematogenous osteomyelitis in the metaphysis spreads by way of the transphyseal vessels to the epiphysis and further into the joint through a piece of bone which does not have an epiphyseal bone core. (**b**) After the appearance of the bone core in the epiphysis the transphyseal vessels disappear and the epiphysis and metaphysis have their own blood supply. In older children a metaphyseal infection as a rule does not spread from the growth plate to the epiphysis with the exception of meningococcal infections and tuberculosis

dren cannot spread from the metaphysis through the growth plate into the epiphysis (Fig. 18.6). Exceptions are in case of meningococcal sepsis and in delayed treatment of a serious acute osteomyelitis and in these cases the growth plates can be damaged.

- In joints in which the growth plate and a part of the metaphysis lie within the joint capsule, such as the glenohumeral joint, humeroradial joint, the hip joint and the talocrural joint, a metaphyseal infection can directly spread into the joint (Fig. 18.5).
- Apart from this a septic arthritis may be caused by local spread of an infection from adjacent tissues or contamination after an intra articular wound, puncture or operation on the joint (Fig. 18.7).

## Localization

The most common localizations in an acute osteomyelitis are the femur and the tibia (Table 18.1).

The knee is the most common localization for a septic or infectious arthritis followed by the hip and the ankle (Table 18.2).

## More Locations

**Neonates** In neonates an acute osteomyelitis or septic arthritis can occur in more locations all at the same time. This is the case in 40 %.

**Meningococcal infection** Luckily these infections are less frequent because of immunization. In a meningococcal (Neisseria meningitidis) infection the sepsis occurs within several hours with a meningitis. The symptoms are fever, headache, vomiting and meningeal irritation. There may be convulsions and there is often an otitis media. Meningeal irritation includes a stiff neck, a positive Brudzinski sign (by flexing the neck the knees are pulled up), a positive Kernig sign (when lying down the extended legs cannot be raised higher than 45°) and the "three foot

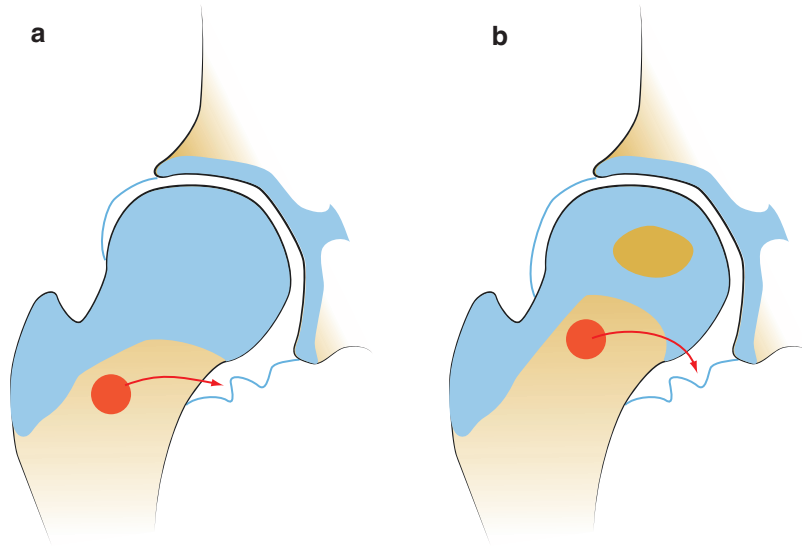



**Fig. 18.6** In joints where the growth plate and part of the metaphysis lie inside the joint capsule a metaphyseal infection can occur in the newborns (**a**) as well as older children (**b**) in which there is a direct extension into the joint

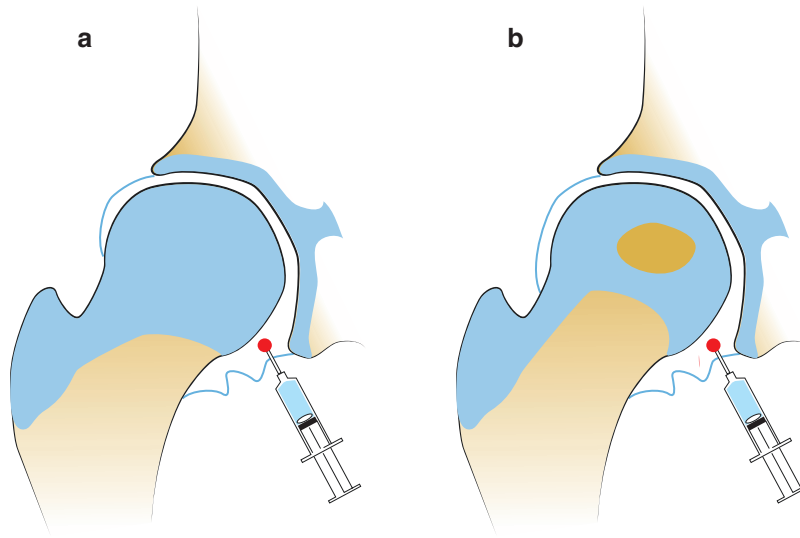



**Fig. 18.7** Apart from a bacteremia a septic arthritis can occur due to local spread from adjacent tissues or through contamination after an intra-articular wound, puncture or operation in newborns (**a**) or in older children (**b**)

phenomenon" (sitting straight up is only possible with the support of both arms that are placed behind the back). Compartment syndromes in extremities with skin necrosis, bone and joint infections can also occur. The infection is so aggressive that the growth plates can be involved in several locations (also in older children) with as a result bony bridges in the growth plates causing serious positional deformities and/or shortening.

## Pathogens

In a hematogenous type there must be an infectious process in other parts of the body. This can be i.e. an ear or throat infection or infection of the airways, urinary tract, intestinal infection, sometimes a skin infection after chickenpox or an infection after a direct wound such as with a nail in the foot, a puncture of a joint or after a surgical operation (Fig. 18.1). A bacterimia occurs from a focus else-

**Table 18.1** Distribution of acute osteomyelitis according to localization

| | |
|---|---|
| Femur | 28 % |
| Tibia | 24 % |
| Humerus | 12 % |
| Fibula | 5 % |
| Radius | 5 % |
| Calcaneus | 4 % |
| Phalanges | 4 % |
| Ulna | 2 % |
| Ilium | 2 % |
| Ischium | 2 % |
| Metatarsals | 2 % |
| Others | 10 % |

From: Jackson MA, Nelson JD. Etiology and medical management of acute suppurative bone and joint infections in pediatric patients. *J Pediatr Orthop*. 1982;2:313–23

**Table 18.2** Distribution of septic arthritis according to localization

| | |
|---|---|
| Knee | 41 % |
| Hip | 23 % |
| Ankle | 14 % |
| Elbow | 12 % |
| Wrist | 4 % |
| Shoulder | 4 % |
| Others | 2 % |

From: Jackson MA, Nelson JD. Etiology and medical management of acute suppurative bone and joint infections in pediatric patients. *J Pediatr Orthop*. 1982;2:313–23

where in the body whereby the bacteria are spread via the blood vessels to the bone or the joint. Usually no focus will be found in children. Bacterimia in children is a frequent daily occurring phenomenon. In 50 % of children a bacterimia is caused after brushing the teeth.

Staphylococcus aureus is the most frequent pathogen in bone and joint infections. Other bacteria involved vary with age.

**Neonates** Staphylococcus aureus and Streptococcus B are the most important pathogens in neonates.

Children from 4 weeks to 5 years of age. In this age group the infection is generally caused by Staphylococcus aureus and Streptococcus A. In a joint or bone infection caused by Haemophilus influenzae meningitis is present in 20 % of the cases. Haemophilus influenzae, a Gram-negative organism, has in many areas disappeared due to vaccinations. Infections by Kingella kingae and other Gram-negative organism are rising in perticular in infants.

Children older than 5 years of age. Staphylococcus aureus is also the most frequent pathogen in this age group. Salmonella bacteria are often involved in a septic arthritis or acute osteomyelitis in children who have sickle cell anemia.

**Prick wounds** An infection as a result of a prick wound, e.g. a child who has trod on a nail with a tennis shoe is usually caused by Pseudomonas aeruginosa. Within 4 days after the initial trauma there is so much pain that the patient cannot bear weight on the foot.

**Teenagers** In sexually active teenagers one should always consider Staphylococcus aureus and Neisseria gonorrhoeae (see p. 144).

Supplementary assessment: in acute osteomyelitis and also in a septic arthritis: the body temperature must be measured, the WBC, CRP and the BSE must be checked. A blood culture is positive in 50 % of cases. X-rays are not necessary in the first instance but may be useful in checking the skeleton prior to changes which occur later due to the infection. A double contour will be seen at the level of the metaphysis after 10–14 days in an inadequately treated acute osteomyelitis. This is due to a subperiosteal abscess (Fig. 18.8). In a septic arthritis of the glenohumeral or hip joint that has not been treated or inadequately treated, a distension occurs within the joint capsule due to accumulation of pus after 10–14 days with a subluxation of the head of the humerus or femur. A technetium scan shows in 80 % of cases a higher activity in an acute osteomyelitis and a septic arthritis. A gallium scan is more specific than a technetium scan but not more sensitive. An MRI in an acute osteomyelitis gives

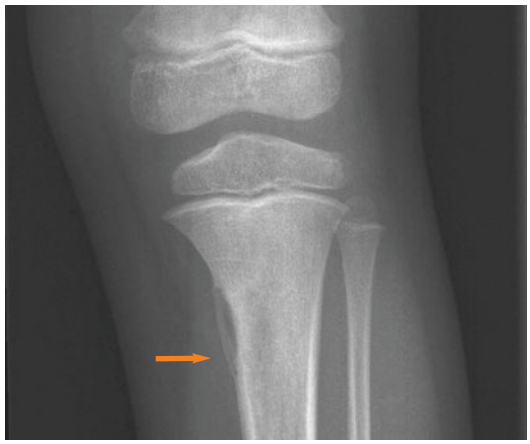


**Fig. 18.8** Anteroposterior X-ray of the left knee. Subperiosteal abscess on the medial side in the proximal part of the tibia (*arrow*)

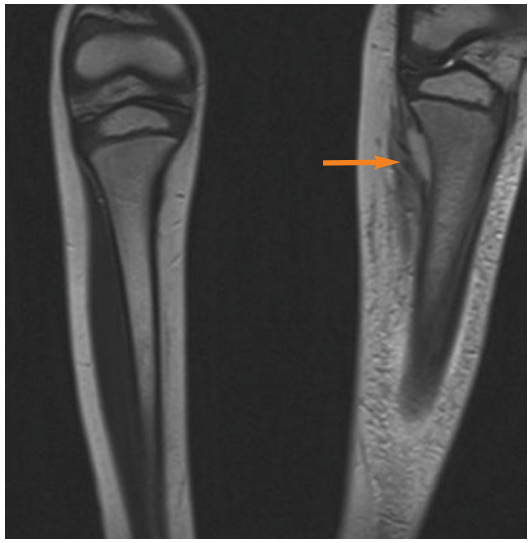


**Fig. 18.9** MRI of both lower legs. Subperiosteal abscess on the medial side in the proximal part of the tibia (*arrow*)

in the first instance no more information than an X-ray. An MRI is useful in order to visualize the extension in a subperiosteal abscess (Fig. 18.9). In a septic arthritis an MRI can only reveal fluid in the joint that can be seen more easily and simply on an ultrasound. The diagnosis osteomyelitis or septic arthritis can only be made with certainty after bone or joint aspiration and a positive culture (Table 18.3).

**Table 18.3** Criteria for the diagnosis of acute osteomyelitis and septic arthritis

| | |
|---|---|
| Possible | The appearance of a bone or joint infection are: pain, redness, swelling, mobility limitation of the involved or adjacent joint, possible fever and general malaise, and no other cause can be found for these complaints. |
| Probable | Positive blood culture with the appearance of bone or joint infection |
| Certain | Positive bone or joint aspiration |

Primary care treatment: in primary care one should particularly consider the possibility of an acute osteomyelitis or septic arthritis in children (younger than 9–18 months of age) who are not sick and are not febrile. The WBC, CRP and BSE are not raised. One should consider an acute osteomyelitis or septic arthritis if the limb is immobile (Table 18.4).

When to refer: children with a suspicion of acute osteomyelitis or septic arthritis must be referred as quickly as possible.

Secondary care treatment: **acute osteomyelitis**. The procedure is as follows: first of all an ultrasound of the joint will be taken, if there is no fluid in the joint then one should consider an acute osteomyelitis. Broad spectrum antibiotics will be given. Normally Staphylococcus aureus is sensitive to these antibiotics. If there is a good response within 48–72 h (the child has less pain, is less ill, the temperature is lower and the CRP is lower), then the treatment with the antibiotics can be continued and no further action is necessary. An aspiration of the bone should take place if there is no improvement within 48–72 h. In 90 % of cases the bone culture will be positive if bacteria are present even with systemic broad spectrum antibiotic therapy. If necessary the antibiotics can be changed according to the results on culture. There are no clear rules regarding the period of antibiotic therapy. In any event they must be continued until the CRP and BSE values are normal. As a rule two weeks of intrave-

**Table 18.4** Aspects of an acute osteomyelitis or septic arthritis in one or more locations in children younger than 9 months up to 1.5 years of age and in older children

| | Child younger than 9 months up to 1,5 years of age | | |
|---|---|---|---|
| | **(one location)** | **(more locations)** | Older child |
| General aspects of infection | Mostly absent | Febrile, general malaise | Normally febrile, general malaise |
| Pain | Severe | Severe | Severe |
| Functional lesion | Severe | Severe | Severe |
| WBC's | Mostly normal | Raised | Raised |
| BSE and CRP | Mostly normal | Raised | Raised |
| X-rays | Normal in acute phase | Normal in acute phase | Normal in acute phase |
| MRI | Normal in acute phase | Normal in acute phase | Normal in acute phase |
| Technetium- and galliumscan | 80 % positive | 80 % positive | 80 % positive |
| Blood cultures | 50 % positive | 50 % positive | 50 % positive |
| Pus on joint aspiration (in arthritis) | 70 % positive | 70 % positive | 70 % positive |
| Pus on bone cultures (in osteomyelitis) | 90 % positive | 90 % positive | 90 % positive |

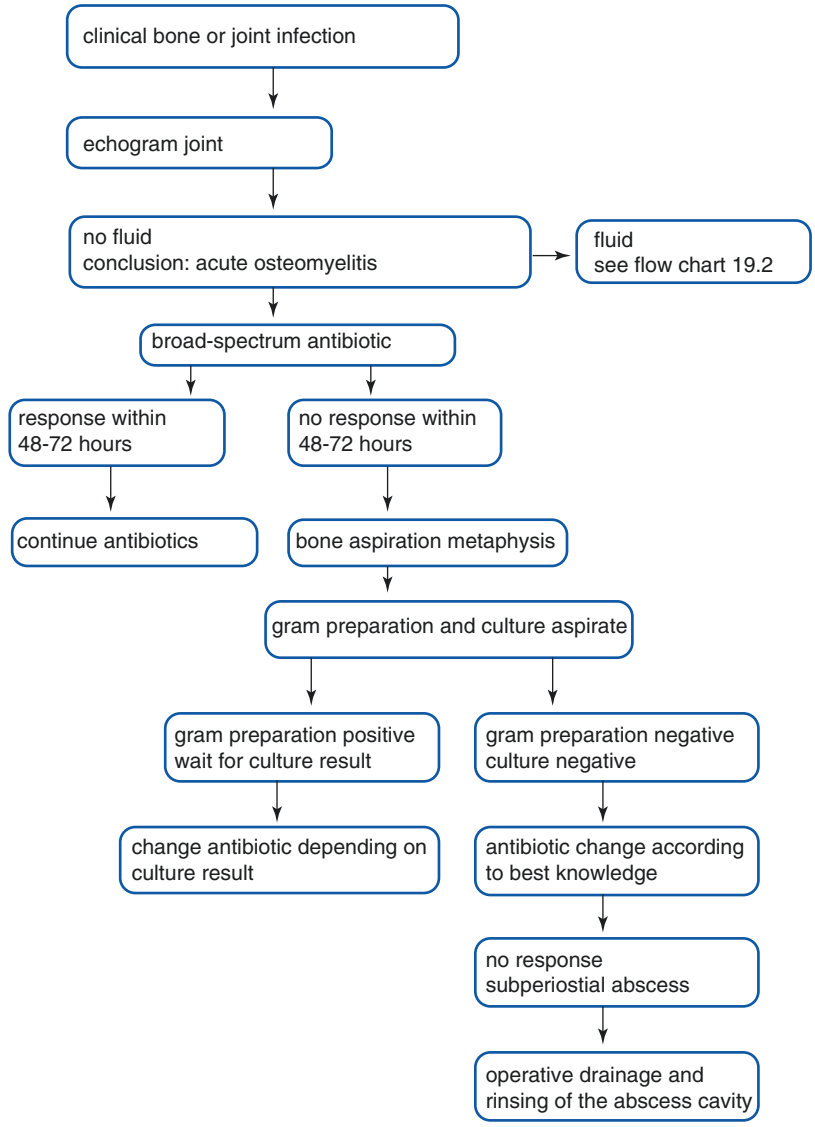
nous and after that, 4 weeks of oral antibiotics are given in acute hematogenous osteomyelitis (see Flow Chart 18.1).

**Septic arthritis** If on ultrasound fluid is present in the joint,then this must be aspirated under anesthetic. A gram preparation is made of a joint fluid including a bacterial culture. There is a septic arthritis if bacteria are seen on the gram preparation. Bacteria will be seen in 70 % of cases with a septic arthritis. If no bacteria are seen on the gram preparation then there is an acute osteomyelitis with a reactive hydrops in the adjacent joint. In that case it is also advisable to aspirate the metaphysis and check for bacteria on a gram preparation. If no bacteria are seen then one assumes that there is a septic arthritis. An acute septic arthritis of the hip joint is an indication for an emergency operation. An arthrotomy will be carried out in the case of a hip joint in which a window is made in the joint capsule on the anterior side so that the pus can be drained and the cavity can be rinsed out. A possible hip dislocation must also be reduced and the head of the femur can be kept in place with an abduction plaster or abduction orthosis (see Flow Chart 18.2). Other involved joints must also be drained. Arthroscopic rinsing is preferable if these are accessible such as in the knee, ankle, elbow and shoulder joint. In the other cases the joints will be punctured with a needle and rinsed. Rinsing the joint once is usually sufficient. The intervention can be repeated if this is not the case. Broad spectrum antibiotics will also be given in a septic arthritis. Staphylococcus aureus is usually sensitive to the antibiotics. Later on the antibiotic policy can be adjusted according to the results on culture. In the case of a septic arthritis the child will mostly be treated with one or more antibiotics during the first 4 weeks, 1 week of intravenous therapy and 3 weeks oral therapy. The CRP and the BSE must be normalized before stopping antibiotics.

## Late Complications

**Chronical osteomyelitis** Serious misalignment or shortening can occur in the involved bone because of damage to the growth plates (Fig. 18.10). There is a chance of a pathological fracture at the level of the osteomyelitis focus and these generally heal poorly.

If an acute hematogenous osteomyelitis remains untreated, then a subperiosteal abscess will be present after several days. The periosteum will be

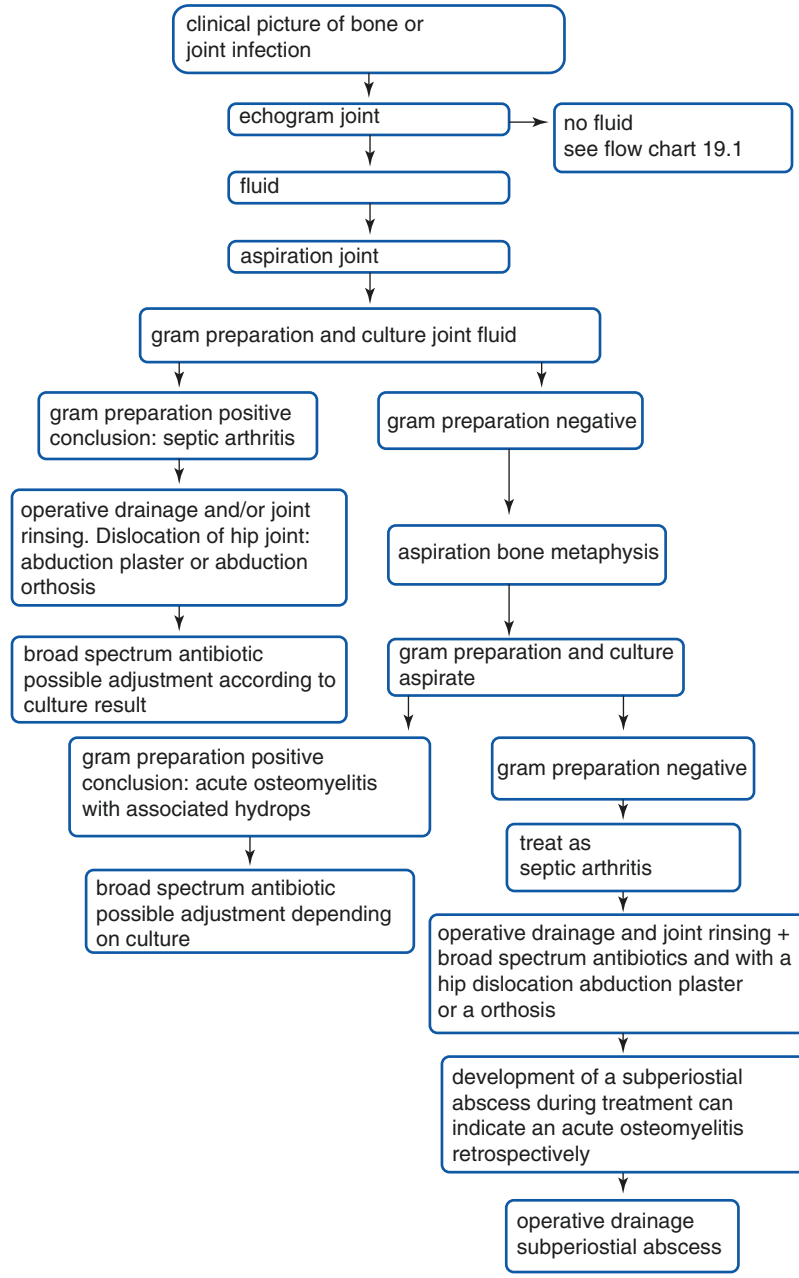



**Flow Chart 18.1** Author's procedure for the treatment of acute hematogenous osteomyelitis

raised up due to the accumulation of pus (Fig. 18.3). This subperiosteal abscess spreads over the whole metaphysis and diaphysis, causing thrombosis in the nutrient artery. As a result the necrotic piece of bone becomes a sequestrum.

The vascularisation of the periosteum remains intact and produces new bony tissue, the involucrum. The sequestrum often drains through a fistula. A cavity occurs, the cloaca, in the area of the sequestrum. A squamous cell carcinoma can develop if the fistula has been present for 15–20 years. A chronic osteomyelitis is not common in the western world but is commonly seen in developing countries.

In this case of chronical osteomyelitis one waits until the periosteum has formed over the complete length of the new bone tissue (the involucrum). At that time the ischemic piece of bone (sequestrum) and a possible fistula can be excised. In the case of a less extensive sequestrum this can be removed together with the involucrum (saucerization) (Fig. 18.11). The child has to be protected with antibiotics up until the




**Flow Chart 18.2** Author's procedure for the treatment of septic arthritis

time of the operation and for several weeks after this until the BSE and CRP are normal.

## Caffey Disease

A chronic osteomyelitis must not be confused with Caffey disease (infantile cortical hyperostosis) (Fig. 18.12). Caffey disease is characterized by a soft tissue swelling, subperiosteal new bone growth and cortical thickening of the involved pieces of bone and fever. The illness begins before the fifth month of life and is sometimes present at birth. A spontaneous recovery can be expected before 3 years of age. In rare cases it involves children older than 3 years of

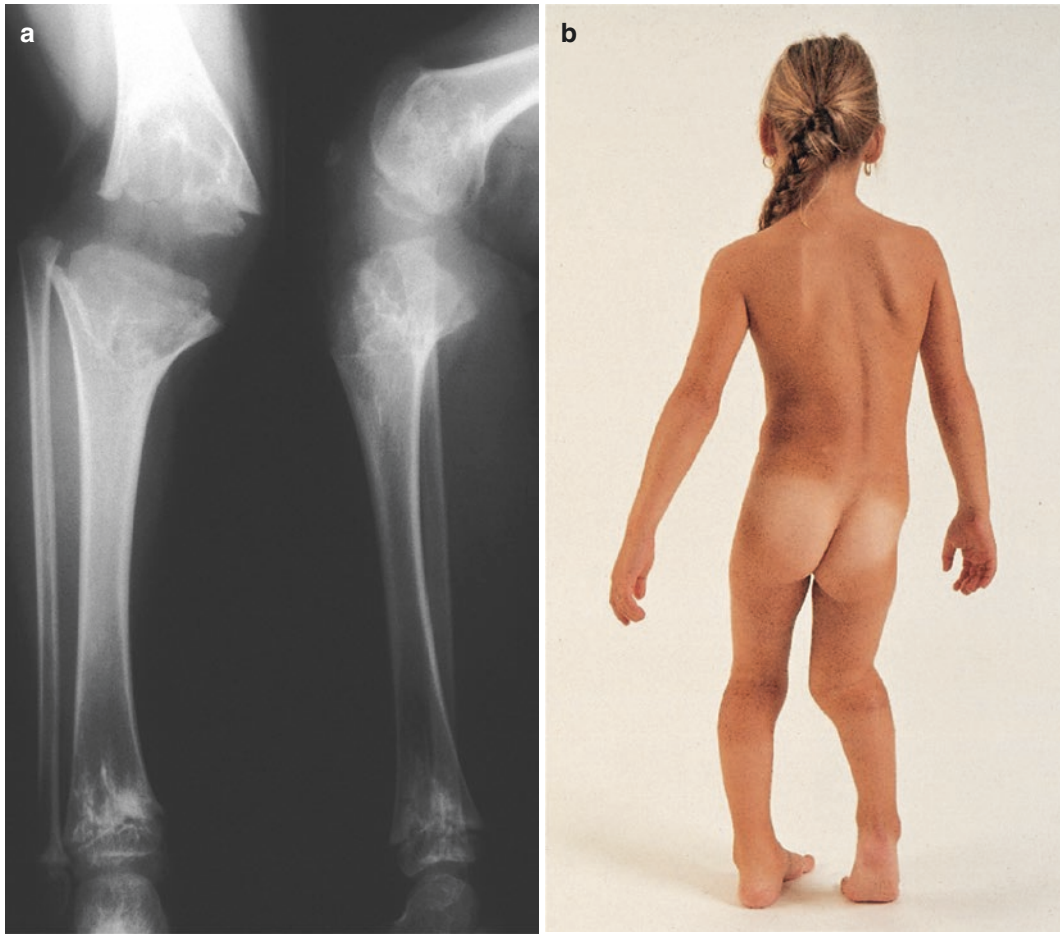



**Fig. 18.10** after meningococcal infection in a newborn. Various growth plates have been damaged and there are serious positional deformities and shortening

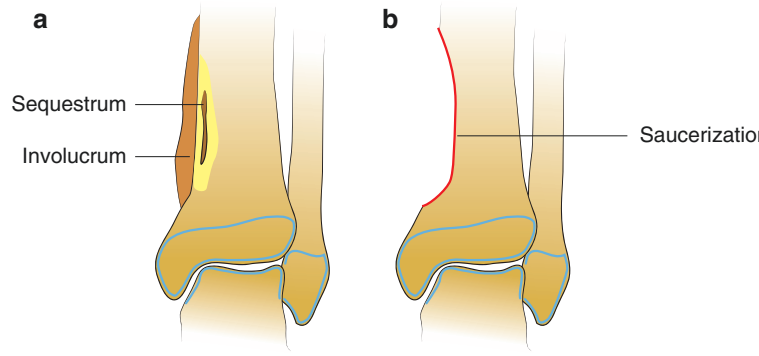



**Fig. 18.11** (**a**) Chronical infection in the distal part of the tibia with the formation of the sequestrum and involucrum. (**b**) The involucrum and the sequestrum are saucerized

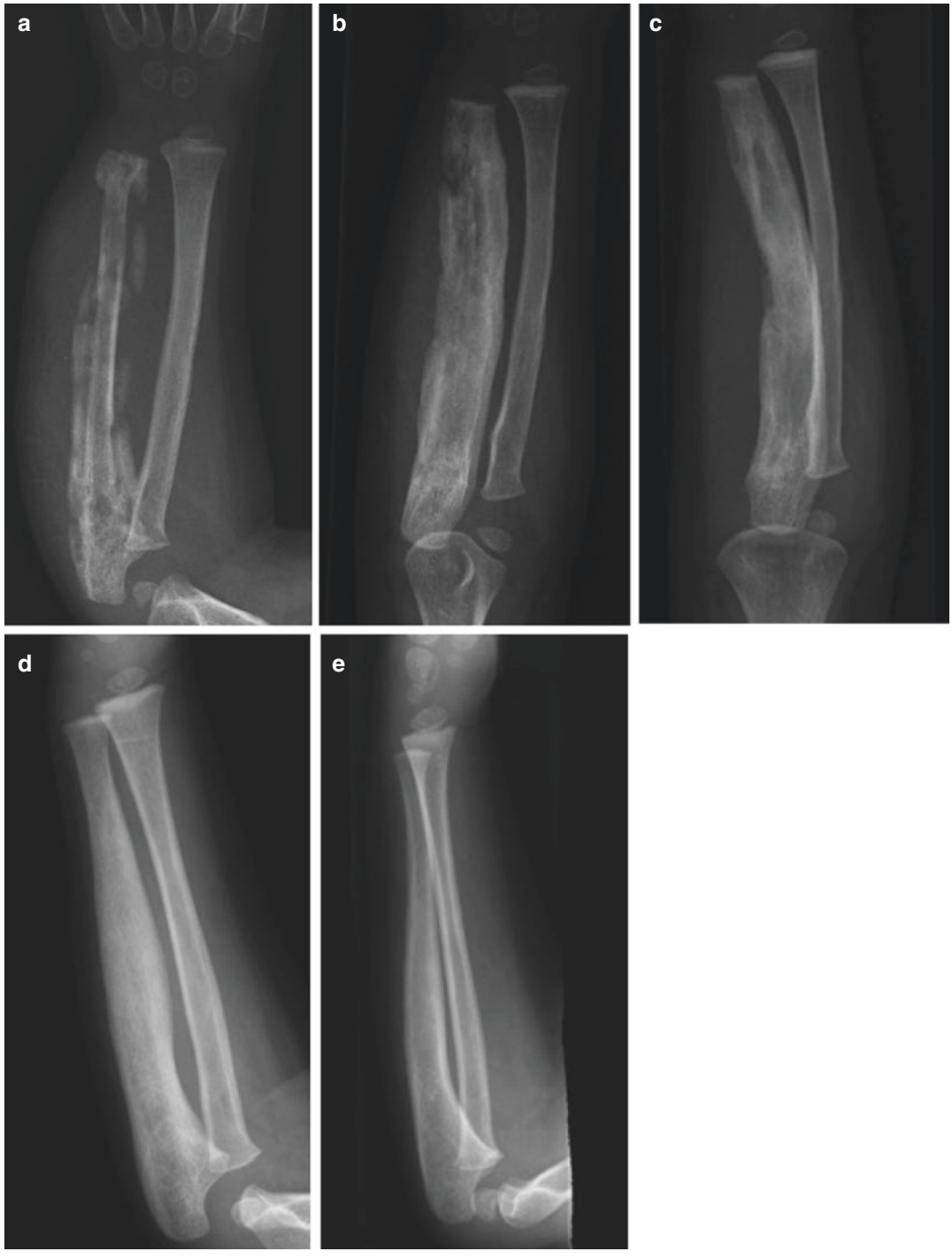



**Fig. 18.12** (**a**) A 3 year old girl with Caffey disease of the ulna. (**b**) Situation 4 weeks after establishing the diagnosis. (**c**) After 6 weeks. (**d**) After 8 months. (**e**) Situation after 2 years

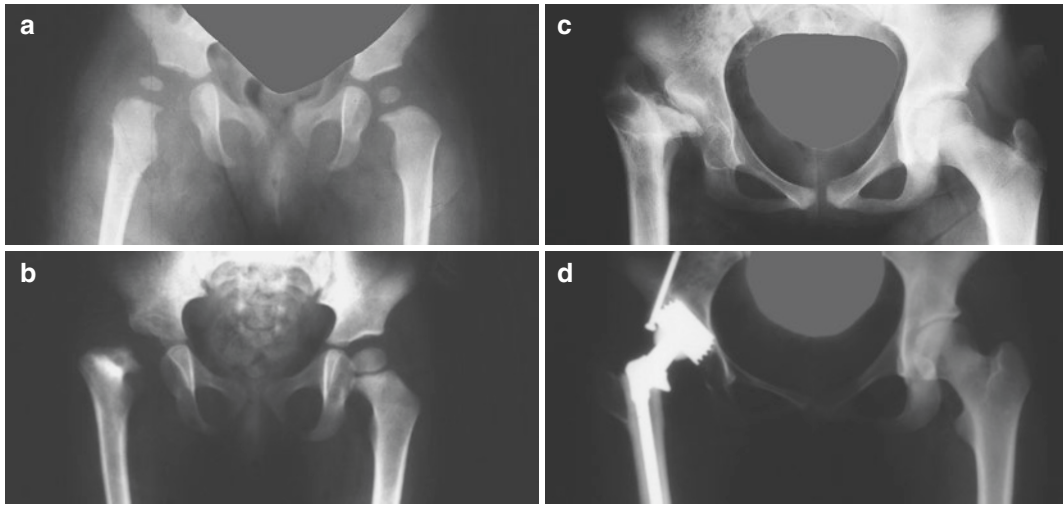



**Fig. 18.13** (**a**) Subluxation of the right hip as a result of septic arthritis. (**b**) A femoral head necrosis in the right hip has occurred as a result of either high pressure in the joint or toxins from the pathogen. (**c**) This has finally led to a serious deformity of the head of the femur, after which (**d**) an uncemented total hip arthroplasty has been carried out in this young adult

age. In a great many cases the mandible is involved as well as the ulna and tibia. The disease can also occur in the clavicles, the scapulae, the ribs and in rare cases the humerus, femur and fibula and is often confused with an osteomyelitis.

Chronical septic arthritis. Damage to the joint cartilage can occur with a joint infection. The damage occurs partly because of pressure caused by the bulk of the infectious process and partly because of the toxins produced by the pathogen. The growth plates can be damaged in joints where the epiphysis and the growth plate lie within the joint capsule.

In a humeroradial and a hip joint the increased pressure in the joint can lead to closure of the retinacular vessels which supply the radial head and the head of the femur causing an ischemic radial head or femoral head necrosis. The raised pressure in the joint can possibly cause dislocations in the hip and shoulder joint (Fig. 18.13).

**Differential Diagnosis: Bone and Joint Infections**

| **Extremely painful immobile extremity** | |
|---|---|
| Children younger than 1.5 years without fever. | **Acute osteomyelitis** |
| | **Septic arthritis** |
| Children younger than 1.5 years with fever. | **Acute osteomyelitis in more locations** |
| Children older than 1.5 years with fever. | **Acute osteomyelitis**<br>**Septic arthritis**<br>**More locations** |

# Addendum

Explanation as to why a child (just as in adults) when standing brings the upper part of the body above the painful involved hip.

Normal individuals standing with both legs on the ground have a center of gravity in the midline of the body at the level of the pelvis minor at the level of the sacrum. When walking the body weight presses alternately on the weight bearing leg and hip. This pressure is the result of body weight on one side and on the other the strength of the abductor muscles that are necessary to keep the pelvis in balance. This situation can be seen as a balance. In a normal walking pattern loading one leg is called balance **X**.

K is the body weight minus the weight of the loaded leg. The weight of one leg is about 15 % of the total body weight. M is the strength of the abductors necessary to keep the pelvis in balance. R is the sum of K + M.

B is the insertion point of the abductor muscles onto the greater trochanter. C is the center of the femoral head and D is the center of gravity of the body. We can now determine the balance. In an individual standing on one leg it is necessary to keep balance, which means $M \times BC = K \times CD$. The ratio of the short balance arm (BC) to the long balance arm (CD) is about 1:3 (in women 1:3.3 and in men 1:2.3). We can fill in the balance. $M \times 1 = K \times 3 M = 3K$. In other words: the abductors must have a strength three times the body weight (minus the weight of the loaded leg) in order to keep the pelvis in balance. The final pressure on the head of the femur(R) is the sum of the body weight minus the weight of the loaded leg (K) and the strength of the abductors (M):

$$R = 1K + M$$

$$R = 1 + 3K = 4K \left( \begin{array}{l} = 4 \times \text{the body} \\ \text{weight min us} \\ \text{the weith of the} \\ \text{loaded leg} \end{array} \right).$$

The question as to why a child with a painful hip brings the upper part of the body above the painful hip when standing can be explained as follows. What the child does in fact is to move the center of gravity of the body onto the involved hip with as a result a smaller balance arm CD. This situation is called balance **Y**.

In this example the distance from the center of the femoral head to the center of gravity of the body D″ is just as big as the distance from the insertion of the abductors B and the center of the femoral head C. The balance is then calculated as follows:

$$M \times BC = K \times CD''$$
$$M \times 1 = K \times 1$$
$$M = 1K$$

The final pressure on the femoral head in this example is as follows:

$$R = K + M$$

$$R = 1K + 1K = 2K \left( \begin{array}{l} = 2 \times \text{the body weight minus} \\ \text{the weight of the loaded leg} \end{array} \right).$$

If the child allows the upper part of the body to be displaced further in which case the center of


J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2

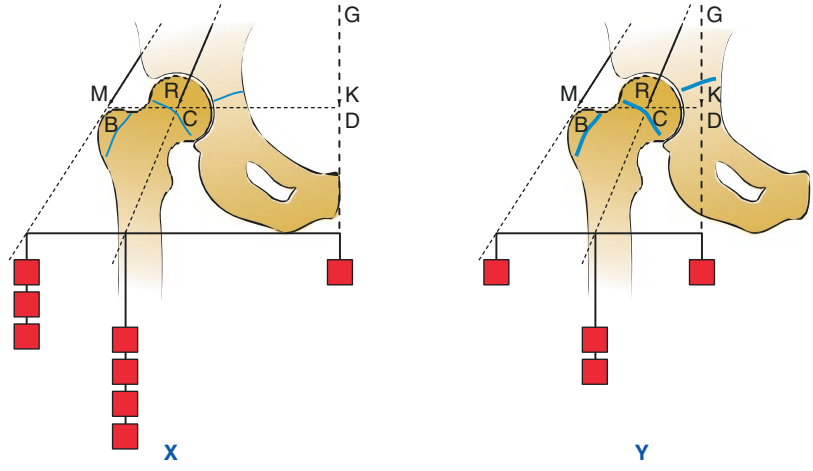
gravity D‴ coincides with the center of the femoral head and CD‴ is therefore 0 cm, then the balance is as follows:

$$M \times BC = K \times CD'''$$
$$M \times 1 = K \times 0$$
$$M = 0K$$
$$R = K + M$$
$$R = 1K + 0K = 1K (= 1 \times \text{the body weight minus the weight of the loaded leg}).$$

The abductors do not need to have more power in order to keep the pelvis in balance (if the child leans over further it will fall).

**X:** loading of the right hip when standing on the right leg with a normal gait. The pressure on the femoral head is four times the body weight minus the weight of the loaded leg

**Y**: loading on the right hip when standing on the right leg in which the upper part of the body leans over the right hip. The pressure on the hip joint is reduced as a result



# Appendix

In this appendix a number of syndromes mentioned in the text will be outlined. The most characteristic deformities are in **bold letters**.

## Achondroplasia

**Inheritance** Autosomal dominant, sporadic in 90 % of cases.

**Incidence** 1 in 15,000–25,000 births.

**Face and skull** **A conspicuous forehead and a saddle nose.** Skeleton: lumbar hyperlordosis. Lumbar scoliosis, flexion contractures of the elbows, short extremities with short and broad long bones. At birth the axes of the legs are usually normal but in **older children genua vara or valga** develops and sometimes a valgus deformity of the ankles.

**In general** Narrowed spinal canal whereby neurological symptoms are present such as pain and paraesthesias in the lower extremities.

**Survival prognosis** Normal.

## Acrocephalopolysyndactyly (Carpenter Type)

**Inheritance** Autosomal recessive.

**Incidence** Up to 1994 40 published cases.

**Face and skull** As a result of premature closure of the coronal suture on both sides there is as **tower shaped skull**. There is also hypertelorism, epicanthus, a flat nose, broad cheeks, a high palate, underdevelopment of the lower jaw (micrognathia) and low implanted ears.

**Skeleton** (brachy) **syndactyly** usually of the **middle finger and ring finger, an extra thumb or a shortened and broadened distal thumb phalanx**, genua valga, **duplication of the big toe** and syndactyly in the toes. Sometimes there is a radioulnar synostosis.

**In general** Mental retardation (75 %) and disturbance in sexual development (hypogenitalism or hypogonadism).

**Survival prognosis** Good.

## Acrocephalosyndactyly (Apert Syndrome)

**Inheritance** Autosomal dominant but also sporadic.

**Incidence** 1 in 45,000 births.

**Face and skull** There is a **tower shaped skull** as a result of premature closure of the coronal suture. There is also hypertelorism and there is also exophthalmus and a broad shortened nose.

**Skeleton** **Syndactyly of the fingers, mostly the central three in which three fingers share one nail. In the thumb the distal phalanx is short and broad**. There may also be syndactyly of the toes. Sometimes there is a synostosis of the radius

and ulna and a dislocation of the radial head. Occasionally a Klippel-Feil syndrome is also present.

**In general** As a result of premature closure of the coronal sutures most of these children are mentally retarded. Sometimes there is an atresia of the oesophagus and heart and renal anomalies.

**Survival prognosis** Good.

## Arthrogryposis Multiplex Congenita (Multiple Congenital Contractures)

**Inheritance** Sporadic.

**Incidence** 1 in 3,000 births.

**Face and skull** Not involved.

**Skeleton** In 40% only the lower extremities, in 10% only the upper extremities and in 50% the lower and upper extremities are involved. **Apart from immobile joints and contractures there is also a severe underdevelopment of the muscles around these joints**. In the upper extremities a classical picture is adduction/endorotation contracture of the shoulders, flexion and extension contractures of the elbows and flexion contractures in the wrist and fingers. As far as the lower extremities are concerned there are flexion contractures of the hips which sometimes are dislocated, hyperextension contractures of the knees which are sometimes dislocated and severe clubfeet.

**In general** Arthrogryposis multiplex congenita is a collective name for several hundred diseases and syndromes in which two or more joints are involved. In the English literature one speaks of multiple congenital contractures. Intrauterine movement is little or none in all types.

**Survival prognosis** Normal.

## Beckwith-Wiedemann Syndrome

**Inheritance** Autosomal dominant.

**Incidence** 7 in 10,000 births.

**Face A big tongue** (macroglossia) is most obvious.

**Skeleton** A type of **hemihypertrophy is present in 20%** of the children in which the whole half of the body is involved but also with crossed hypertrophy or hypotrophy in one extremity. **Great body height mostly obvious before the first year of life**. Apart from this, other skeletal deformities may be present including a scoliosis, hollow feet, polydactyly and a congenital radial head dislocation.

**In general** Enlargements of a number of organs such as the pancreas, and a navel hernia, hypoglycemia and neurological abnormalities.

**Survival prognosis** Good.

## Biemond II Syndrome

**Inheritance** Autosomal recessive.

**Incidence** 1 in 175,000 births.

**In general** The Biemond II syndrome has the same appearance as in the Laurence-Moon-Biedl-Bardet syndrome but only the eye deformities are different. In this syndrome there is **no iris (aniridia) or there is iris hypoplasia**.
See also Laurence-Moon-Biedl-Bardet syndrome.

## Camp(t)omelia Dysplasia (Cumming Dysplasia Syndrome)

**Inheritance** Not fully known.

**Incidence** 1 in 200,000 births.

**Face** Flat face, flap ears and micrognathia (small mandibula). Skeleton: **femurs, tibias and fibulas are curved anterolaterally. There are pretibial dimples** and possibly an equinus or clubfoot. Cervical kyphosis, broad short hands and syndactyly of the third and fourth toe.

**In general** Hydronephrosis (38 %), congenital heart anomalies (21 %).

**Survival prognosis** 77 % die directly after birth or at a young age.

## Conradi-Hünermann Syndrome (Chondrodysplasia Punctata)

**Inheritance** Autosomal dominant and recessive.

**Incidence** Not known.

**Face and skull** Hypertelorism. A congenital cataract is present in 70 % of recessive cases and in 20 % of dominant cases. There is a large forehead, a saddle nose and little hair growth.

**Skeleton** Scoliosis, **contractures of the elbows, hips and knees**. Cervical kyphosis.

**In general** Mental retardation (50 %). **At 4 years of age stippled calcifications seen on the X-rays have disappeared**. Survival prognosis: in the dominant type usually normal. In de recessive type half of the children die in the first year of life and there are very few who reach adulthood.

## Cornelia De Lange Syndrome

**Inheritance** Autosomal dominant.

**Incidence** 1 in 10,000 births.

**Face and skull** Patients have a characteristic face: **long continuous eyebrows, long curly eyelashes, a turned-up nose and a carp mouth**.

**Skeleton** The elbow can be underdeveloped with a dislocation of the radial head. There is only one palmar crease and there may be shortened phalanges of the fingers and toes and **a short, broad metacarpal I and syndactyly of the second and third toes**.

**In general** Mental retardation and a retarded growth.

**Survival prognosis** Good.

## Cranio-Carpo-Tarsal Syndrome (Freemans-Sheldon Syndrome or Whistling-Face Syndrome)

**Inheritance** Usually sporadic but can also be autosomal dominant or recessive.

**Incidence** Up to 2002 80 cases were described in literature. Face and skull: small mouth, **facial expression of someone who is whistling**. H-shaped groove in the chin. High palate with a small tongue.

**Skeleton** Ulnar deviation of the hands, flexion contractures in the fingers, no thumb opposition, clubfeet.

**In general** Frequent aspiration pneumonias.

**Survival prognosis** Some of the children die as a result of aspiration pneumonias.

## Diastrophic Dwarfism

**Inheritance** Autosomal recessive.

**Incidence** 2 in 1,000,000 births.

**Face and skull** Cystic swelling in the ears in children. This causes cauliflower ears in adulthood.

**Skeleton Dwarfed growth with short extremities. There is a shortened and broadened**

**metacarpal I and a hitch-hiker** thumb, flexion contractures or dislocations of the knees and hips and in many cases foot anomalies such as metatarsus adductus and clubfeet. At birth the back appears to be normal but they frequently develop a scoliosis or kyphoscoliosis.

**In general** The height in adulthood is between 80 and 140 cm. Intelligence is normal.

**Survival prognosis** 25 % die during the first year of life.

## Down Syndrome (Trisomy 21)

**Inheritance** Sporadic (94 %).

**Incidence** 1 in 660 births.

**Face and skull** Short neck, **brachycephaly, straight hair, vertical skinfold over the medial canthus (epicanthus), tongue with fissures, low implanted small ears**.

**Skeleton** Short metacarpals, brachydactyly, clinodactyly, short and deformed middle phalanx little finger (60 %), a single transverse palmar crease **(simian crease) (55 %)**, metatarsus primus varus (90 %). Atlanto, occipital or atlanto-axial subluxation, **developmental dysplasia of the hip (70 %)**.

**In general** Heart anomalies (40 %), usually a ventricular septal defect, hypermobile joints (90 %), mental retardation (100 %), small stature (adult men on average about 155 cm and women 145 cm).

**Survival prognosis** The prognosis is good if there are no heart anomalies. The survival chance as a result of airway infections and heart anomalies has improved with the introduction of antibiotics and heart surgery. Average life expectancy is 60 years.

## Ehlers-Danlos Syndrome

**Inheritance** Dependent on the type: x-chromosome recessive, autosomal dominant.

**Incidence** 1 in 10,000 births.

**Face and skull** Bilateral epicanthus. Patients can reach the nose with the tip of their tongues. The ears can be stretched out.

**Skeleton** (lordo) scoliosis, hypermobile joints with genua recurvata and in 25 % of the cases hip dislocations. Furthermore possible dislocations of shoulder, elbow and patella, hyperkyphosis, scoliosis and flatfeet. The number of creases in the handpalm is more than normal.

**In general** It involves an inherited connective tissue disease with increased elasticity of connective tissue leading to an **extremely elastic skin, hypermobile joints and fragility of blood vessels and organs that can easily bleed**. The condition has an extensive spectrum in which the degree of hyperelasticity of the skin, the hypermobility of the joints and the fragility of the vessels and organs varies considerably. Up until 1997 10 or 11 types were known. Later on another classification has been made in which there are six basic types and three rare types. The classical types have hyperelastic and fragile skin and the hypermobile types have hypermobile joints. These two types have 90 % prevalence.

**Survival prognosis** Dependent on the blood vessel fragility.

## Ellis-van Creveld Syndrome (Chondro Ectodermal Dysplasia)

**Inheritance** Autosomal recessive.

**Incidence** 1 in 100,000 births. Is relatively frequent in the Amish population.

**Face and skull Frugal scalp hair growth including the eyebrows with anomalies of the nails and teeth**.

**Skeleton** This syndrome is also characterized by dwarfism, small chest, an **extra little finger**, an extra little toe, fusion of the capitate and hamate bones (7 %) or other **carpal fusions** (71 %).

**In general** Congenital heart anomalies.

**Survival prognosis** 50 % die in the first years of life.

## Fanconi Anemia

**Inheritance** Autosomal recessive.

**Incidence** 1 in 350,000 births.

**Face and skull** Small head, visual disturbances in which objects appear smaller than normal (microphthalmia), strabismus, epicanthus, blue sclerae, dysplastic ears and deafness.

**Skeleton** Radial hypoplasia or aplasia, hypoplasia of the shoulder and chest muscles, triphalangeal thumb and an extra thumb or a shortened and small metacarpal I.

**In general** There is **a pancytopenia in combination with a radial hypoplasia or aplasia**. The pancytopenia is discovered at an average age of 8 years and varies from birth up to 20 years of age. The child is susceptable to infections and hemorrhages. Some-times café au lait areas are seen in the skin.

**Survival prognosis** 35 % mortality before 10 years of age.

## Goltz Syndrome (Goltz Gorlin Syndrome or Focal Dermal Hypoplasia)

**Inheritance** In 95 % of cases sporadic, in which 95 % are girls and in 5 % there is an x-chromosome dominant trait.

**Incidence** Up until 1994 more than 200 cases have been published.

**Face and skull** Frugal scalp hair, strabismus, split iris, irregular pupils, papillomas of the lips, tooth deformities, pointed chin, teleangiectasias in the atrophic cheek skin.

**Skeleton syndactyly of fingers and toes and an extra little finger**.

**In general Skin anomalies** such as epitheleal swellings of the lips and genitals, atrophy and pigmentation of the skin.

**Survival prognosis** Good.

## Hand, Foot and Uterus Syndrome

**Inheritance** Autosomal dominant.

**Incidence** Eight families have been described up until 1993.

**Skeleton The first metacarpal is short and broad and the same with the first metatarsal.** The little fingers are short and have a clinodactyly. There may also be a synostosis between the trapezium and scaphoid bone and between the medial cuneiform bone and the navicular bone.

**In general** Urogenital anomalies.

**Survival prognosis** Good.

## Holt-Oram Syndrome (Heart-Hand Syndrome)

**Inheritance** Autosomal dominant.

**Incidence** Up until 1991 more than 200 cases have been described.

**Skeleton There are only anomalies in the upper extremity**. The anomalies in the upper extremity are asymmetrical and these are worse on the left side than on the right side. Anomalies in the upper extremities: a shortened and small first metacarpal, cleft hand, triphalangeal thumb, an extra thumb, phocomelia, absent scaphoid, duplication of the scaphoid or synostosis between the scaphoid and other carpal bones, synostosis of the forearm, radial hypoplasia or aplasia and hypoplastic shoulder muscles.

**In general** Heart anomalies. These are ventricular septal defects or an atrial septal defect.

**Survival prognosis** Depends on the heart anomaly.

## Hunter Syndrome (Mucopolysaccharidosis Type II)

**Inheritance** X-chromosome recessive (therefore only boys).

**Incidence** 6 in 100,000 births.

**In general** Hunter syndrome **appears similar to Hurler syndrome. However, there are no cloudy corneas** and the inheritance is different. Hunter syndrome manifests itself between 2 and 4 years of age. **Dermatan and heparin sulphate levels are greatly raised in the urine**.

**Survival prognosis** They die as a rule before 20 years of age because of obstruction to the coronary vessels and/or broncopneumonias.

## Hurler Syndrome (Mucopolysaccharidosis Type I)

**Inheritance** Autosomal recessive.

**Incidence** 1 in 100,000 births.

**Face** Crude facial expression, hypertelorism, saddle nose, cloudy corneas and glaucoma.

**Skeleton** D**warf growth**, short neck, **kyphosis, hip dislocations, irregular femoral head epiphysis and poorly developed acetabulae, short robust hands**.

**In general** Mental retardation, thickened coronary vessels and heart valves. Chronic infection of the nasal mucosa and upper airways causing deafness and bronchopneumonias. Hurler syndrome becomes obvious between 1 and 2 years of age. Dermatan sulphate is highly raised in the urine.

**Survival prognosis** These children die mostly before 10 years of age due to obstruction of the coronary vessels and/or broncho pneumonias.

## Jeune Syndrome

**Inheritance** Autosomal recessive.

**Incidence** 1 in 100,000 births.

**In general** The anomalies are the same as in Ellis-van Creveld syndrome, the difference is that **the chest is much smaller than in the Ellis-van Creveld syndrome**.

## Juberg-Hayward Syndrome (Orocraniodigital Syndrome)

**Inheritance** Autosomal recessive.

**Incidence** 12 cases have been described in literature up to 2002.

**Skull and face** **Cleft lip and palate**.

**Skeleton** **The first metacarpal is short and small, hypoplasia or aplasia of the thumb**, radial hypoplasia or aplasia, radial head subluxation, humeroradial synostosis and a radioulnar synostosis.

**Survival prognosis** Good.

## Klinefelter Syndrome (Chromosome XXY Syndrome)

**Inheritance** Faulty division of the chromosomes.

**Incidence** 1 in 500 boys.

**Face and skull** Not abhorrent.

**Skeleton** Long thin extremities in which the distance between the head and the pubis is less than

the distance between the pubis and the heel. Normally these are equal. The anomaly is at the beginning not obvious. The diagnosis is usually made during puberty. An orthopaedic anomaly that relatively frequently occurs is a **radioulnar synostosis**.

**In general** **Hypogenitalism and hypogonadism** with absent spermatogenesis and gynaecomastia. There is mental retardation in 20 % of cases.

**Survival prognosis** Good.

## Klippel-Feil Syndrome (Congenital Brevicollis)

**Inheritance** In a few cases it is autosomal dominant but in most cases it is not inheritable.

**Incidence** 1 in 42,000 births.

**Skeleton** The anomaly is characterized by a **short neck**. The short neck is caused by vertebrae which have fused together (block vertebra) or are absent. Besides this there may be unilateral (unsegmentend) bony connections, hemivertebrae, absence of posterior elements (arches and spinal processess), a basilar impression, an atlanto-occipital fusion, an atlanto-axial subluxation and dens hypoplasia. There is a torticollis in 20 % of cases and in 60 % a thoracic type of scoliosis as a result of hemivertebrae and unilateral unsegmented bony connections. There is a raised shoulder blade in 30 % of cases, and this is known as a Sprengel deformity.

**In general** It involves girls in 65 % of cases. There are heart and kidney anomalies in 25 % of cases, in 30 % deafness and in 5–10 % a cleft palate. Neurological complications are possible in adulthood because of compression of the spinal nerves.

**Survival prognosis** Good.

## Klippel-Trenaunay-Weber Syndrome (Angio-Osteohypertrophy)

**Inheritance** Sporadic.

**Incidence** 900 cases have been described up until 1994.

**Face and skull** Not anomalous.

**Skeleton** Dystrophy of the soft tissues and skeleton. Sometimes there is a scoliosis, syndactyly, polydactyly, clinodactyly or macrodactyly.

**In general** The syndrome is characterized by a combination of **cutaneous hemangiomas, varicosities and hemihypertrophy of the soft tissues and skeleton**. Vascular anomalies belong to the so-called congenital angiodysplasias and usually involve venous anomalies. Arteriovenous fistulas, lymphangiomas and lymphoedema can also be present. The severity of the varicose vessels varies but is obvious around 12 years of age. Clinically significant arterio-venous shunting can finally occur. The anomalies are located in the lower extremity in 95 % of cases but can be present in other areas of the body, sometimes in several locations. These anomalies can only lead to hemorrhage in the digestive tract. Asymmetry is often most evident in the leg but the leg length difference is relatively less impressive. The children have a relative mild form in 50 % of cases but about 50 % of the patients develop thromboembolic problems.

**Survival prognosis** Normal in most cases.

## Larsen Syndrome

**Inheritance** Autosomal dominant or sporadic.

**Incidence** 120 cases have been described up to 1994.

**Face and skull** Conspicuous forehead, **saddle nose** and hypertelorism. There can also be a cleft

palate and hydrocephalus. Skeleton: **frequent inborn dislocations of wrists, elbows, hips and knees.** Less frequent: club feet, cervical kyphosis and short metacarpals.

**In general** **Short fingernails**.

**Survival prognosis** Good.

## Laurence-Moon-Biedl-Bardet Syndrome

**Inheritance** Autosomal recessive.

**Incidence** 1 in 175,000 births.

**Face and skull** **Retina degeneration (retinitis pigmentosa) is present in 70 % of cases**.

**Skeleton** **There is an extra little finger present in 75 %** of cases. Sometimes there is a syndactyly.

**In general** Obesity and mental retardation. Kidney anomalies and underdevelopment of the genitalia in 60 % of men and in 25 % of women.

**Survival prognosis** 35 % die early in life.

## Léri-Weill Disease (Dyschondrosteosis)

**Inheritance** Pseudoautosomal dominant.

**Incidence** Not known.

**Face** No anomaly.

**Skeleton** Most consistent is the presence of a **Madelung deformity, as a rule bilateral**. As a result of short forearms and short lower legs (mesomelia) there is a **disproportional dwarfism**. Other skeletal anomalies can be short hands and feet (shortening of the fourth metacarpal and the metatarsals). Limited mobility of the shoulders or elbows or both. Dislocation of the elbow, genu valgum.

**In general** The final adult height is 137 up to 152 cm.

**Survival prognosis** Good.

## Mafucci Syndrome

**Inheritance** Sporadic.

**Incidence** 180 cases have been published up until 2004.

**Face and skull** No anomaly.

**Skeleton** **Dwarfism, enchondromata**, leg length difference, scoliosis and pathological fractures.

**In general** Subcutaneous **hemangiomas are seen in 25 % of the children at birth and in 75 % before the fifth year of life**. The enchondromata and the subcutaneous hemangiomas are particularly unilateral and involve one extremity.

**Survival prognosis** 20–30 % chance of developing a chondrsarcoma, usually after the age of 40 years. Other tumors are also possible.

## Marfan Syndrome

**Inheritance** Autosomal dominant, 25–30 % sporadic.

**Incidence** 1 in 100,000 births.

**Skeleton** The patients have a characteristic appearance **with disproportional long, thin extremities with small fingers (arachnodactyly)**. There is joint laxity and a high position of the patellae (patella alta) with subluxations, genua recurvata and flat feet. 50 % of cases have a (lordo) scoliosis. Other back anomalies: lumbar kyphosis, flat back and a spondylolisthesis particularly at the level of L5-S1. There is a **pectus excavatum in 70 %** of cases. Apart from that diverse other anomalies can

develop in the skeleton, such as acetabular protrusions.

**In general** One can make the diagnosis Marfan if two or more of the four following aspects are present: skeletal deformities, lens dislocation, anomalies of the cardiovascular system and a positive family history. There is a dislocation of the lens in **80 % of cases, a so-called ectopiaor luxatio lentis**. Two thirds of the children have an anomaly of the heart and blood vessels of which the most characteristic is **dilatation of the aorta** but anomalies in the mitral valve are frequent.

**Survival prognosis** Average age at death: 45 years.

## Möbius Syndrome

**Inheritance** Not inheritable (sporadic).

**Incidence** More than 200 cases described in the literature up until 1990.

**Face and skull** Characteristic is a paralysis of the facial nerve (cranial nerve VII) **whereby there is no facial expression** and paralysis of the abducens nerve (cranial nerve VI) which causes a strabismus. Other cranial nerves can also be paralytic, e.g. the hypoglossal nerve (cranial nerve XII) which occurs most frequently with as a result paralysis of the intrinsic muscles of the tongue with as a result difficulty in speaking and eating. Skeleton: **half of the children have club feet** and at least a third have hand deformities varying from underdeveloped nails or even complete absence of the hand. Poland syndrome is an accompanying phenomenon in 15 % of cases.

**In general** 10–15 % of patients are mentally retarded. A real Möbius syndrome, involves paralysis of the cranial nerves VI and VII in combination with other cranial nerve paralyses, Poland syndrome and congenital anomalies of the hands and feet.

**Survival prognosis** Good.

## Morquio Syndrome (Mucopolysaccharidosis Type IV)

**Inheritance** Autosomal recessive.

**Incidence** 1 in 100,000 births.

**Face and skull** Normal.

**Skeleton** As a result of flat vertebral bodies (platyspondyly) there is dwarfism of the shortened trunk. Apart from the kyphosis there can also be a progressive (kypho)scoliosis and atlantoaxial instability as a result of hypoplasia or absent dens. One of the most impressive appearances is the 90° angle between the manubrium and the sternum **(pectus carinatum). The femoral head epiphyses are extremely irregular. The acetabula are poorly developed. Genua valga and short heavy hands**.

**In general** Normal intelligence. Inborn anomalies of the heart valves. Morquio syndrome is usually diagnosed between 1 and 3 years of age. High levels of keratin sulfate in the urine. Survival prognosis: frequently die before 20 years of age as a result of airway infections or heart problems.

## Mucopolysaccharidosis

**Inheritance** Seven types have been described. All of these with exception of the Hunter syndrome are autosomal recessively inherited. Hunter syndrome occurs only in boys. The most well known types are Hunter syndrome, Hurler syndrome and Morquio syndrome.

**Skeleton** In all types of mucopolysaccharidosis there is **dwarfism and skeletal anomalies involving the vertebral column, hips and hands**.

**In general** The largest group have a lysosomal storage disease. This can be determined on the grounds of the release of mucopolysaccharides. This usually particularly involves **heparin-, der-**

**matinand keratin sulphate, which are present in high concentrations in the urine**.

**Survival prognosis** Usually die before 20 years of age as a result of respiratory infections or heart problems.

## Multiple Epiphyseal Dysplasia

**Inheritance** Autosomal dominant.

**Face and skull** Normal.

**Incidence** 9 in 100,000 births.

**Skeleton** After the appearance of an **ossification centre the epiphyses of the long cortical bones are flattened which gives rise to painful hips, knees and ankles, short metacarpals, with short small squatty fingers**. Genua vara or valga.

**In general** The hip deformities are always bilateral in contrast to the situation in Perthes disease. Frequently occurring back pain and slowly progressive stiffness in the joints occurs in children. There is arthritis generally before the age of 30. Children are small. Final height prognosis: 145–170 cm.

**Survival prognosis** Normal.

## Myositis Ossificans Multiplex Progressiva (Progressive Myositis Ossificans or Fibrodysplasia Ossificans Progressiva)

**Inheritance** Autosomal dominant and sporadic.

**Skeleton** This involves a progressive ossification of fascias, aponeuroses, ligaments, tendons and connective tissue. The muscles are normal. The best name is in fact fibrodysplasia ossificans progressiva. Apart from the progressive ossification there is also a **shortened and broadened first metacarpal and a short and broadened first metatarsal.** Progressive calcification and thereafter ossification develops usually before 10 years of age but can develop from birth up to 20 years of age. The swellings begin usually in the neck and at the level of the shoulder. There is a limited mobility in the shoulders, hips and knees. This all leads to serious invalidity.

**In general** There is no special treatment. Removal of the extra ossal bone gives disappointing results. Radiotherapy makes the situation worse.

**Survival prognosis** Good.

## Nail Patella Syndrome (Onycho-Osteodysplasia)

**Inheritance** Autosomal dominant.

**Incidence** 1 in 50,000 births.

**Face and skull** Abnormal eyes: ptosis, cataract, iris anomalies with amongst other aspects a colour difference of both irises (heterochromia).

**Skeleton** There are **absent or underdeveloped patellae in 60–90 % of cases** with hypoplasia of the lateral femoral condyle. The underdeveloped patellae lead often to recurrent dislocations in the patellofemoral joints. In 60–90 % of cases there is also a **deformity of the elbow joints** (radial head aplasia, aplasia of the capitellum, elbow dislocation) and less mobility in proand supination. There are horn-like anomalies on the outside and in the center of the right and left iliac bones in 2/3 of cases. Pectus carinatum, scoliosis, underdeveloped scapulae, clinodactyly and camptodactyly may also be present.

**In general** A**bnormal nails, the nails of the fingers are more frequently involved than the toe nails**. The thumb is usually the most involved, particular on the ulnar side and the index and other fingers are less involved. There are renal anomalies in 30 %.

**Survival prognosis** If there are no renal anomalies, then the prognosis is good. 10 % of the patients between 5 and 25 years of age die if there are renal anomalies.

## Neurofibromatosis (Von Recklinghausen Disease)

**Inheritance** Autosomal dominant or sporadic (50 %).

**Incidence** 1 and 3000 children. There are five different types of neurofibromatosis. In 85 % of cases there is a Von Recklinghausen NF1 disease.

**Face and skull** Apart from possible fibromas under the skin, no anomalies.

**Skeleton** In 70 % a kyphoscoliosis, in 7 % a tibial pseudarthrosis, asymmetrical overgrowth of the lower extremities and macrodactyly. Cervical kyphosis.

**In general** **The diagnosis can be made with certainty if the patient has at least two of the seven following characteristic features**

- six or more café-au-lait areas larger than 5 mm prior to puberty or larger than 15 mm after this time,
- two or more neurofibromas or a plexiform neurofibroma,
- diffuse freckles in skin creases,
- specific primary bony defect, such as a tibial pseudarthrosis,
- optic nerve glioma,
- two or more Lisch nodules in the iris,
- first degree family member with an NF1.

The chances of malignancy in neurofibromas is 1–5 % and less frequently occuring tumors are in the optical and the acoustic nerve. Furthermore there is a chance of hypertension, early puberty and in 40 % of cases a low IQ and in 2–5 % mental retardation.

**Survival prognosis** Normal.

## Ollier Disease (Enchondromatosis)

**Inheritance** Sporadic. Familial only described in six cases.

**Incidence** Unknown. Personal experience: three cases.

**Face and skull** Not abnormal.

**Skeleton** **Multiple enchondromas**. There is a mild and severe type of Ollier disease. There are enchondromas in several metaphyses in the cortical long bones in the upper and lower extremities and metacarpals. Conspicuous is the fact that the deformity often appears in both extremities but is asymmetrical. There can be a very mild type in one extremity and in the other a severe type with as a result a difference in leg length but also axial deformities. Enchondromas stop growing at puberty after which a certain amount of remodelling occurs.

**In general** Ovarian tumors are frequent.

**Survival prognosis** Malignant transformation in about 30 % (chondrosarcomas).

## Orofacial Digital Syndrome Type I and Type II

**Inheritance** Type I: x-chromosome dominant. Type II: autosomal recessive.

**Incidence** Type I: 1 in 45,000 births. Type II: 1 in 300,000 births.

**Face and skull** **There is a cleft lip, cleft tongue, small eyeballs (microphtalmia), eyes close together and a pinched nose**. Skeleton: **brachydactyly and syndactyly of fingers and toes and an extra little finger**.

**In general** There is a difference between types I and II. In type I there is baldness (alopecia) and multiple hyperplastic frenula. In type II these characteristics are absent.

**Survival prognosis** Type I only in girls, boys with this type do not survive.

## Osteogenesis Imperfecta

**Inheritance** Type I: autosomal dominant. Type II: sporadic, in 5 % autosomal recessive. Type III: autosomal recessive. Type IV: autosomal dominant.

**Incidence** 1 in 20,000 births.

**Face and skull** Broad skull, projecting axillas, triangular face and developmental disturbances in the teeth.

**Skeleton** **Type I: dark blue sclerae**, generalized bony fragility and deafness (otosclerose). **10 % of these children have fractures at birth** and most of them before 5 years of age. A kyphosis of the back occurs with the passage of time. **Otosclerosis** is present at 50 years of age and is present in nearly 100 % of cases. There is an opalescence of the teeth.

**Type II: extreme bone fragility. Intrauterine fractures very frequently present**. The long cortical bones are completely misformed. **These children die in the perinatal period or shortly after**.

**Type III: progressive deformation of the long cortical bones and the back**. Multiple fractures. **The sclerae are blue at the time of birth but become white during growth**.

**Type IV: this is the mildest type. Fractures only occur in the first years of life**. Small posture. Sometimes the long cortical bones are bent and a kyphoscoliosis can occur in adulthood. Sometimes there is dentogenesis imperfecta.

**In general** There is a collagen structural anomaly as a result of abnormal osteoblasts that are not capable of building up a good quality organic matrix. The occurrence of fractures varies. In type II intrauterine fractures can occur. Fractures occur in type IV only in the first few years of life and become less as the child gets older. In adulthood there is no increased occurrence of fractures. Fractures occur more frequently in the lower than in the upper extremities. This is completely the opposite in normal children. Children often have a small posture. There is hypermobility. There is a kyphoscoliosis in 20–40 % of cases. The large cortical bones are short and broad with a very thin cortex on X-ray. The fractures heal normally.

**Survival prognosis** Type I and IV have a normal life expectancy. Adults with type III often die in the fourth or fifth decade as a result of heart valve defects. In type II brain injuries occur because of the deformation of the skull which leads to death in the perinatal period or shortly after.

## Poland Syndrome

**Inheritance** Sporadic and seldom autosomal dominant.

**Incidence** 1 in 32,000 births, 70 % are boys.

**Skeleton** There is a **unilateral absence of the pectoralis minor muscle and the sternal part of the pectoralis major**. As a rule there are also hand anomalies on the same side such as absence of fingers in combination with a small hand (hypoplasia), syndactyly and too short fingers because of absence of one of the phalanges in all fingers (brachydactyly). The combination of hypoplasia of the hand, syndactyly and brachydactyly is called symbrachydactyly. There may possibly be other deformities such as absence of forearm flexors, underdevelopment of the whole arm, Klippel-Feil syndrome (short neck), Möbius syndrome (paralysis of the facial muscles), Sprengel deformity and a pectus excavatum.

**Survival prognosis** Good.

## Proteus Syndrome

**Inheritance** Mostly sporadic, sometimes autosomal dominant.

**Incidence** In 2001 more than 200 cases had been published.

**Face and skull** Saddle nose, microcephaly.

**Skeleton** Unilateral or local hypertrophy of more types of tissue, which lead to **gigantism of the**

**trunk or limbs**. There are serious positional deformities in the legs, such as knock knees in combination with gigantism. Disproportional overgrowth, mostly asymmetrical in the extremities or only involving the hands and feet (macrodactyly).

**In general** Progressive hypertrophy. Connective tissue naevi are particularly seen on the footsole and look a little bit like a brain. Apart from that other frequent tumors are ovarian tumors and diverse vascular malformations.

**Survival prognosis** Usually normal.

## Pseudohypoparathyroidism (Albright's Hereditary Osteodystrophy)

**Inheritance** Sporadic.

**Incidence** 20 cases published up to 1997.

**Face and skull** Prominent forehead in small children and circular face, in older children apart from a prominent forehead and a circular face, ptosis of the upper eyelids and a saddle nose.

**Skeleton Brachydactyly of the fourth and fifth metacarpal** and less involved third, second and first metacarpal. Brachydactyly of the toes with broad nails. Other skeletal deformities can be a ulnar hypoplasia and genu valgum.

**In general** Mental retardation in more than 50 % of cases. Calcifications in the soft tissues around the skull and extremities.

**Survival prognosis** Normal.

## Rothmund-Thomson Syndrome (Congenital Poikilodermatosis)

**Inheritance** Autosomal recessive.

**Incidence** Up to 2001 there were about 200 cases described in the literature, particularly girls.

**Face and skull** Some of the children have frugal hair growth, not only in the scalp but also in the eyebrows and the eyelashes. Some of them are completely bald. There are often dental deformities. Some of the patients have a cloudiness in front of or behind the lens (zonular cataract). This cloudiness occurs usually around the age of 5 years.

**Skeleton** Bodily deformities can be: dwarfism, scoliosis, brachydactyly, syndactyly, a cleft hand, hypoplasia or aplasia of the radius or ulna and hypoplasia or aplasia of the thumb.

**In general** Characteristic are the **skin anomalies that usually occur in the first 3–6 months of life and begin with an erythema in the face, later on in the buttocks and the extremities and finally over the whole body**. It begins first of all as an erythema and later on gets a marble appearance. Apart from that there may be depigmentation, hyperpigmentation and wide capillaries in the skin (teleangiectasia). Skin cancer can occur in adulthood.

**Survival prognosis** Good.

## Rubinstein-Taybi Syndrome (Otopalatodigital Syndrome)

**Inheritance** Not completely clear. Anomalies in boys are worse than in girls.

**Incidence** More than 100 cases have been described.

**Face and skull** Characteristic face with a broad nose, a **cleft palate** and abnormal dentition.

**Skeleton** A **broad distal phalanx in the thumbs and fingers**, short fingers, syndactyly, clinodactyly, broad shortened big toe, pectus excavatum and elbow deformity.

**In general** **Short posture**, mental retardation and **deafness**.

**Survival prognosis** Good.

## Russell-Silver Syndrome

**Inheritance** Sporadic.

**Incidence** About 200 cases have been reported up to 1992.

**Face** **Characteristic triangular face** with thin upper lip.

**Skeleton** In 80 % hemihypotrophy whereby the involved side is on average 2–6 cm shorter. Small adults (<150 cm tall). **Broad thumbs and big toes**, possible clinodactyly or camptodactyly of the little fingers.

**In general** Urogenital anomalies.

**Survival prognosis** Normal.

## Spondylo-Epiphyseal Dysplasia

**Inheritance** Autosomal dominant.

**Incidence** 1 in 100,000 births.

**Face and skull** Flat face.

**Skeleton** The most characteristic anomalies are localized in the vertebral column. There are **wedge shaped and flattened vertebral bodies (platyspondylia)**. As a result of that there is a short neck and a short trunk, kyphoscoliosis and an accentuated lumbar lordosis. Hypoplasia of the odontoid process (dens). C1-C2 subluxation. Apart from the vertebral column the hips and shoulders can also be involved. **The epiphyses in the proximal part of the humerus and femur are irregular and deformed**.

**In general** Dwarfism but not always. Quickly tired, poor muscle strength, strong chance of arthritis in the hips and shoulders.

**Survival prognosis** Normal.

## TAR Syndrome

**Inheritance** Autosomal recessive.

**Incidence** About 200 cases have been reported in literature up to 2003.

**Skeleton** There is always a **bilateral aplasia of the radius**, and possible deformities in the humerus and ulna. Apart from these there may be anomalies in the lower extremities such as subluxations, rotational disturbances and hypoor aplasia of the lower extremities.

**In general** The T stands for **Thrombocytopenia** and AR for Aplasia of the Radius. The syndrome is characterized by thrombocytopenia. 40 % of the children have hemorrhages at a young age which disappear with the passage of time. Thrombocytopenia is mostly resolved spontaneously by the age of 4 or 5. Apart from thrombocytopenia other anomalies may be present such as tetralogy of Fallot or an atrial septal defect. Fallot tetralogy involves a defect in the ventricular septum, dextro position of the aorta, pulmonary artery stenosis and hypertrophy of the right ventricle. Sometimes there is a Meckel diverticulum.

**Survival prognosis** 40 % die early in life.

## Trisomy 13 (Patau Syndrome)

**Inheritance** Sporadic.

**Incidence** 1 in 6,000 life births.

**Face and skull** **Microcephaly, posteriorly directed forehead, microphthalmia, cleft lip, cleft palate and a broad nose**.

**Skeleton** Radial hypoplasia or aplasia, syndactyly, camptodactyly, an **extra little finger and an extra small toe**.

**In general** Congenital heart and kidney anomalies.

**Survival prognosis** About half die in the first month of life, 75 % within 6 months and less than 5 % will become older than 1 year. The oldest patient described in literature died at the age of 35.

## Trisomy 18

**Inheritance** Sporadic.

**Incidence** 1 in 6,000 live births. It occurs three times more commonly in girls than boys.

**Face and skull** **Cleft lip, cleft palate and an underdeveloped mandible (micrognathia)**.

**Skeleton** Anomalies can be: thumb hypoplasia and aplasia, syndactyly, polydactyly, cleft hand, hypoplasia or aplasia of the radius, phocomelia, hip dislocations, hemivertebrae, scoliosis, club feet, pes convex valgus and an extra small toe.

**In general** Congenital heart and kidney anomalies.

**Survival prognosis** About one third of these children die in the first month of life, about 50 % within 2 months after birth and less than 10 % live to be older than 1 year of age.

## VATER- or VACTERL-Association

**Inheritance** Sporadic.

**Incidence** 3 in 20,000 live births.

**Skeleton** 60 % of the children have a **skeletal anomaly that is mosty localized in the vertebral column with segmentation disturbance and sacral agenesis (absent sacrum)**. The anomalies in the vertebral column are two times more frequent than deformities of the extremities such as **radial hypoplasia or aplasia**.

**In general** (V = Vertebral, A = Anal, T = Trachea, E = (o)Esophageal, R = Renal). The syndrome is known nowadays as the VACTERL-association in which the C stands for Cardiac anomalies and the L for anomalies of the Limbs. Usually in the VATER-association three of the five components are present but in the VACTERL-association four of the seven components. In 60 % of the children there is an **anus anomaly** (mostly an anus atresia), 85 % of the children have **tracheo-oesophageal fistulas**, 2/3 of the children have renal anomalies in which 40 % have a unilateral renal aplasia. **Cardiac anomalies** occur in 60 % and in 2/3 of these cases there is a ventricular septal defect.

**Survival prognosis** 75 % of these children die early in life.

## Yunis Varon Syndrome

**Inheritance** Autosomal recessive.

**Incidence** 18 cases have been reported.

**Face and skull** Defective growth of the bones of the skull. Hypotelorism, dysplastic ears, sparse hairs, absent eyelashes and eyebrows. Thin lower lip, cupid bow upper lip and narrow high arched palate.

**Skeleton** Hypoplasia or aplasia of thumbs and big toes or distal aphalangia, complete or partially absent clavicles. Bilateral hip dislocation.

**In general** Prenatal and postnatal growth deficiency.

**Survival prognosis** These children usually die in the first year of life.

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# Index




J.D. Visser, *Pediatric Orthopedics*, DOI 10.1007/978-3-319-40178-2