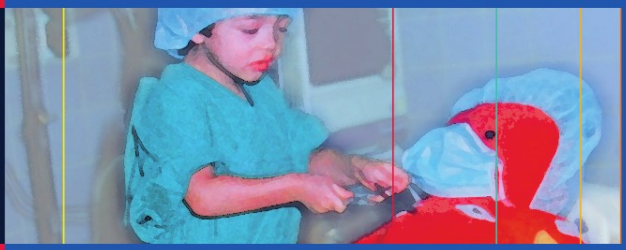
Zacharias Zachariou
Editor



Pediatric Surgery Digest

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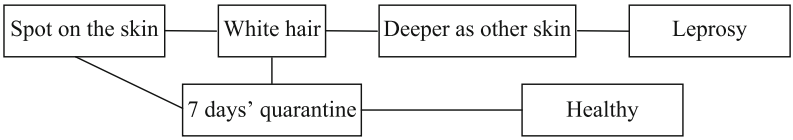
Preface

A discipline for childcare must remain young, responsive, and flexible. This flexibility has to be sustained in order for pediatric surgeons to establish their position, which lies in the fields of both pediatrics and surgery. The concept of this book is to break down barriers and to enable young physicians to look up everyday questions concerning diagnostic measures, therapeutic regimes, postsurgical care, and prognostic values during rounds, in collaboration with pediatricians. It also offers a solid and quick overview of the European Pediatric Surgical curriculum.

And the priest shall look on the plague in the skin and when the hair in the plague is turned white, and the plague in sight be deeper than the skin of his flesh, it is a plague of leprosy: and the priest shall look on him and pronounce him unclean. If the bright spot be white in the skin of his flesh, and in sight be not deeper than the skin, and the hair thereof be not turned white; then the priest shall shut up him, that hath the plague seven days.

Leviticus 13, 3–4

This bible quotation brings a step-by-step solution to a problem. Leprosy is excluded or confirmed following the guidelines in this formula, as for each condition an individual proposal is suggested. An algorithm is set up using the words "when," "it is," "if," "then ," etc. However, the mass of words is confusing to the reader, who constantly has to refer to prior information in the text. The essentials in this communication icon are lost due to the lack of abridgement. By enlarging the "blank areas" the specific information is enhanced, providing the same information much quicker in the overview.



The user follows just the path that indicates the actual findings, omitting those that are of no interest in that specific case. Time saving is not the only benefit of this approach: much more information can be concentrated as well in this concise form of text.

Despite the fact that this book cannot substitute for a tutorial-type textbook, it can be utilized well by someone who is proficient in the fundamentals of pediatric surgery. It is divided into four parts. Following the introductory Part I, Part II deals with pre- and postoperative guidelines, surgical principles, and emergencies in childhood. Part III deals with diagnosis, therapy, and the prognosis of pediatric surgical diseases, while Part IV reflects normal laboratory values and the dosage regimens of pharmaceuticals in childhood. It may be that a reader thinks that the book would benefit from the addition of further appendices, in which case the editor would welcome their suggestions, and gaps can then be filled in a future edition.

I wish to thank all the distinguished authors, from nearly all the European countries, for their contributions in order to promote pediatric surgical knowledge. A lot of the figures were kindly placed at our disposal by Professors Prem Puri and Michael Höllwarth, from Professor Ahmed Hadidi and Professor Klaas Bax. For their skillful secretarial work I would like to thank Ursula Gueder and Remus Vezan. Special thanks also go to Dr. Vera Pedersen for proofreading this manuscript, considering both grammatical and medical aspects. Finally I would like to thank the editorial staff of Springer-Verlag, especially Mrs. Gabriele Schröder and Mrs. Stephanie Benko, for their valuable support.

Berne, June 2008
Zacharias Zachariou

Synopsis

The contents of this manual offer a praxis-orientated and up-to date overview of all subjects in paediatric surgery according to the European Paediatric Surgical Curriculum. Additionally interdisciplinary aspects concerning pediatrics, adult surgery, obstetrics and other disciplines working with children were considered.

The tables presented enable quick access to indications for the operative and conservative therapy with schematic step-by-step illustrations of nearly all surgical procedures. Practical information on child-adapted dosages for pharmacotherapy as well as age-specific injury patterns from scalding in young children to spleen rupture in teenaged vehicle users are also taken into consideration. The surgical role in modern pediatric oncology is also presented according to the guidelines of SIOP.

European paediatric surgeons, experts in their fields, write this book not only for students and trainees but also for anybody who treats children surgically.

Contents

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PART I
INTRODUCTION

1 Introduction

1.1 What is Pediatric Surgery?

The easiest answer to this question is the semantic interpretation of the words pediatric and surgery, meaning surgery on a child. Is pediatric surgery, however, the transposition of surgical procedures to a smaller human being? The main difference between an adult and a child is the fact that the child is growing and throughout the time between its birth and entrance to adulthood – about 18 years – this child never stays the same. Considering this fact, the surgical procedure in pediatric surgery has to be adapted to the child's age, something with which an adult surgeon is not confronted. This philosophy was best expressed in the nineteenth century by the British novelist Charles Dickens who wrote the following statement:

"It is not enough, that a wise physician, who succeeds in curing an adult with a medicine, reduces this prescription in order to treat a child. Some diseases occur only in children and some others, that afflict adults also, develop in other forms in children that diverge from each other as a child and an adult. Children are not miniature adults."

This visionary statement reflects the quintessence of pediatric surgery. Malformations of the gastrointestinal tract for instance are only found in children. Then again, acute appendicitis takes a very different course in children compared to adults, with perforation occurring much earlier. Bone fracture treatment is the best example showing that any therapy applied to a growing organism has to consider the child's growth potential.

Pediatric surgery is not the mere application of surgical procedures to a child but requires special knowledge about embryology, pediatrics, growth

pathophysiology, and specific therapy principles. Pediatric surgery is a specialization on the growing organism.

Medical knowledge has grown enormously in recent decades. This fact has led to the subspecialization of medicine on the basis of organs. Pediatric surgeons are perhaps the only remaining physicians who treat the whole organism and not simply an individual organ. This task is very difficult, as it is impossible to be the best in all fields. The pediatric surgeon is, in my opinion, the "manager" of the surgically ill child. Indeed, pediatric surgery offers the infrastructure in which the medical specialist can come to the child, in a child- and family-friendly environment, to offer her/his great expertise. The pediatric surgeon puts the child in the center and cares for them with the best possible treatment. If this treatment is not offered by pediatric surgery, then, according to the local setting, adult surgeons from the institution, from elsewhere in the country or from other parts of the world may provide the solution. The pediatric surgical "bull's eye" (Fig. 1.1) illustrates the solution providing optimal treatment.

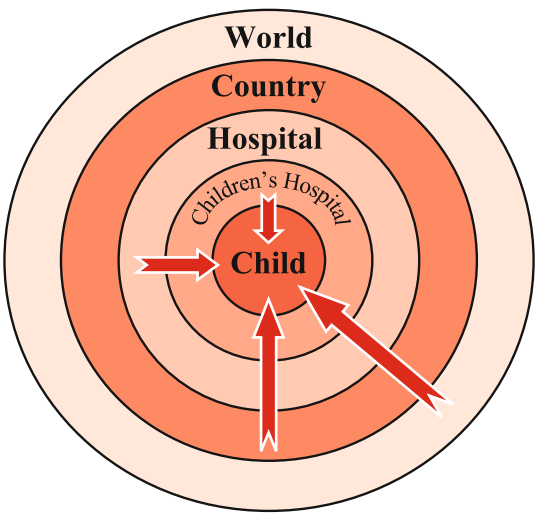


Fig. 1.1 Pediatric Surgical "Bulls eye"

1.2 Timing of Operations in Pediatric Surgical Patients

General

The following considerations are necessary before the indication for operation is set (see Table 1.1 for proposed ideal age for elective operations):

- Is the patient compromised due to the disease? (i.e., intrauterine bowel perforation)
- What risks for the patient could occur due to the operation? (i.e., premature newborn)
- Is the operation at this point technically possible? (i.e., hand malformations)
- Is a spontaneous healing possible? (i.e., umbilical hernia or hydrocele in the newborn)
- What are the psychosocial aspects of the therapy and the hospitalization? (i.e., intersex)

Table 1.1 Proposed ideal age for elective operations in pediatric surgery

Malformation	Age	Hospitalization
Craniosynostosis	1st to 3rd month	2 weeks
Meningocele	3rd to 6th month	2 weeks
Lip, cleft	3rd to 9th month	1 week
Palate, cleft	18th to 36th month	1 week
Prominent ears	5th to 6th year	Day surgery
Thyroglossal fistula, cyst	As from the 3rd month	5 days
Torticollis	6th to 12th month	4 days
Inguinal hernia	After diagnosis	3 days
Umbilical hernia	After the 12th month	Day surgery
Testicular position anomalies	18th to 24th month	4 days
Varicocele	Grade III and according to signs	2 days
Phimosis (medical indication)	3rd to 5th year	Day surgery
Hypospadias	6th to 12th month	4–14 days

Table 1.1 (*continued*) Proposed ideal age for elective operations in pediatric surgery

Malformation	Age	Hospitalization
Bladder exstrophy ▪ Turn-in ▪ Epispadia repair ▪ Continence repair	 Newborn period 3rd year 4th to 5th year	 2–3 weeks 2–3 weeks 2–3 weeks
Kidney and descending urinary tract ▪ Ureteropelvic- junction obstruction ▪ Vesicoureteral reflux	3rd month	2–3 weeks
Hirschsprung's disease (definitive)	3rd to 6th month	3 weeks
Anorectal anomalies	3rd to 12th month	1–3 weeks
Hexadactyly	3rd to 6th month	1–2 weeks
Syndactyly	6th to 24th month	1–2 weeks
Funnel chest	8th to 10th year	2 weeks
Exostosis	With hitting puberty	3–7 days
Bone cysts	As from the 5th year	3–7 days
Hemangioma	According to signs, 6th to 18th month	2 days/day surgery
Ambiguous genitalia	As soon as possible –18th month	3 weeks

1.3 Preoperative Management

Medical history

- Pre-, peri- and postnatal anamnesis
- History of the surgical disease
- Anamnesis of pediatric diseases
- Risk factors (diabetes, hemophilia, asthma, heart malformations, etc.)
- Time of last food ingestion

Clinical status

- General status
- Surgical local finding
- Inspection (scars), palpation, auscultation
- Anthropometric measurements (weight, height, head circumference)
- Rectal examination (especially in abdominal diseases)

Laboratory examinations

- Full blood analysis (including serum glucose)
- Differential blood count with thrombocytes
- Electrolytes
- Coagulation status
- Urine analysis
- Laboratory examinations can be omitted in cases where the child is over 1 year old, has never been seriously ill, and is undergoing a routine operation

Additional diagnostic measures

- Chest X-ray (as indicated by the history)
- Electrocardiogram (ECG; as indicated by the history)
- Ultrasonography (appendicitis, hypertrophic pylorus, blunt abdominal trauma, etc.)
- C-reactive protein (CRP) and blood culture (sepsis, infections)

Special diagnostic measures

- Liver enzymes
- Liquor puncture
- Head CT/MRI
- Electroencephalography (EEG)
- 24-h creatinine clearance

Contraindications for elective surgical procedures

- Infections of the upper respiratory system with fever and leukocytosis
- Children who have teething problems

- Those who have received vaccinations in the previous 4 weeks (especially live vaccines)
- Those who have chronic disease (diabetes, asthma, etc.) and whose treatment is not optimally adjusted

Emergency measures

- Respiratory distress (see Chap. 3): mechanical ventilation if
  - Breathing rate is 50% lower than age norm
  - The patient's general condition is getting worse in light of the $PaCO_2$ (normal range 4.7–6 kPa)
- Cardiac decompensation: catecholamines (see Sect. 3.5)
- Anemia: transfusion when Hb under $7\,g \cdot dl^{-1}$
- Dehydration: fluid and electrolyte substitution and treat the acidosis
- Convulsion attack: substitution in case of metabolic cause, lower the temperature if there is fever, lumbar puncture

Transportation of high-risk pediatric surgical patients

- Transport in an incubator accompanied by a neonatal specialist
- Vascular access
- Stomach drainage
- Sterile dressing of open wounds
- Respiratory support if necessary; intubation before transport
- Vitamin K prophylaxis in newborns

1.4 The Newborn as a Pediatric Surgical Patient

Definition of the newborn period

- The first 3 days (short sense), the first 28 days (broad sense)

Classification according to birth weight (Fig. 1.2)

- Normal: between the 10th and 90th percentiles for the gestational age
- Small for gestational age: under the 10th percentile for the gestational age
- Large for gestational age: over the 90th percentile for the gestational age
- Low birth weight infant: less than 2500 g independent of gestational age
- Very low birth weight infant: less than 1500 g
- Extremely low birth weight infant: less than 1000 g

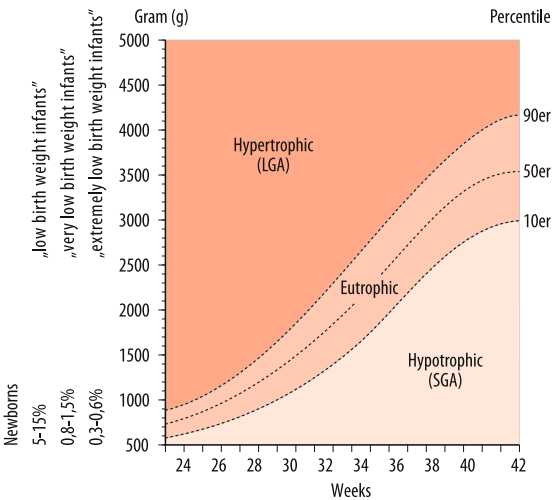


Fig. 1.2 Newborn classification according to gestational age and body weight

Organ function changes after birth

- Organ function changes and adaptation processes are listed in Table 1.2 and must be considered when deciding perioperative and operative therapy
- During the newborn period a steady state should be aimed for every operation (Table 1.3)

Table 1.2 Changes of the organ functions after birth and adaptation

Organ system	Normal values	Malfunctions
Respiratory ▪ Lung drainage during birth ▪ Surfactant activation ▪ Initial hypoxia as stimulus	▪ Breathing rate: 30–55 · min^{-1} ▪ $PaCO_2$ 40–45 mmHg ▪ PaO_2 75–85 mmHg	Respiratory distress syndrome ▪ Tachypnea ▪ Flaring of alae nasi ▪ Moan ▪ Inspiratory retraction ▪ Cyanosis
Cardiovascular ▪ Fetal circulation conversion ▪ Pulmonary resistance ↓ ▪ Oval foramen/Botalli (ductus arteriosus) duct obliteration	▪ Heart rate: 110–140 beats · min^{-1} ▪ Systolic blood pressure 45–65 mmHg ▪ Blood volume: 8%–10% of birth weight ▪ Hb: 16–22 g · dl^{-1} ▪ Hct: 48–66%	Heart malformations and re-establishment of the fetal circulation
Metabolic ▪ Hypoglycemia <40 mg% ▪ Tremor ▪ Apathy ▪ Convulsion ▪ Hypotension	▪ Hypocalcemia <2 mmol · l^{-1} ▪ Agitation	▪ Hyperbilirubinemia >18–20 mg% ▪ Rhesus incompatibility, with hemolysis ▪ Prematurity ▪ Biliary atresia
Water/electrolytes ▪ Immature kidneys ▪ Body weight water fraction: 80% ▪ Concentration ability ↓ ▪ Na^+ reabsorption ↓	▪ Glomerular filtration rate: 25 ml · min^{-1} · 1.73 m^{-2} ▪ Concentration ability: 600–700 mosmol · l^{-1}	▪ Kidney malformations ▪ (Potter syndrome) ▪ Obstruction/reflux urinary system
Thermoregulatory ▪ Deficient isolation ▪ Large body surface ▪ Poor cutaneous vasoconstriction		▪ Hypothermia <36°C ▪ Breathing distress ▪ Hypoglycemia ▪ Acidosis

Table 1.3 Establishing a steady state in newborns before operation

Preoperative	Intraoperative	Postoperative
▪ Correct respiratory insufficiency ▪ Balance fluid deficiency ▪ Treat anemia ▪ Normoglycemia ▪ Compensate acidosis	▪ Normothermia ▪ Normoglycemia ▪ Normoxia ▪ Adequate fluid application	▪ Same principles as intraoperatively ▪ Wound treatment ▪ Bowel movements ▪ Pain treatment

PART II
GENERAL CONSIDERATIONS

2 Enteral/Parenteral Nutrition in Infants and Children

2.1 Nutrition of the Newborn

Breastfeeding should be encouraged for at least the first 4 months of life; this is the optimal formula for the newborn, protecting it from infections and allergies as well as offering beneficial effects in terms of the child's psychological development. From the 6th to 7th month of life, complimentary food, providing iron, trace elements, and fibers, should be introduced. See Table 2.1 and Fig. 2.1 for a schedule of energy and essential nutritional elements for infants according to age and gender and Table 2.2 for energy data for carbohydrates, fats and proteins.

Table 2.1 Schedule of energy and essential nutritional elements for infants according to age and gender

Age	Energy ($kcal \cdot kg^{-1} \cdot day^{-1}$) m/f [a]	Protein ($g \cdot kg^{-1} \cdot day^{-1}$) m/f	Fat (% kcal)	Essential fatty acids (% kcal)	Essential fatty acids (% kcal)	Calcium ($mg \cdot day^{-1}$)	Magnesium ($mg \cdot day^{-1}$) m/f
0 to <4 months	91/94	1.5–2.7	45–50	4.5	0.5	220	24
4 to <12 months	90/91	1.1–1.3	35–45	3.5	0.5	400	60
1 to <4 years	91/88	1.0	30–40	3.0	0.5	600	80
4 to <7 years	82/78	0.9	30–35	2.5	0.5	700	120
7 to <10 years	75/68	0.9	30–35	2.5	0.5	900	170
10 to <13 years	64/55	0.9	30–35	2.5	0.5	1.100	230/250
13 to <15 years	56/47	0.9	30–35	2.5	0.5	1.200	310
15 to <19 years	46/43	0.9/0.8	30	2.5	0.5	1.200	400/350

Age	Iron ($mg \cdot day^{-1}$) m/f	Iodine ($\mu g \cdot day^{-1}$)	Zinc ($mg \cdot day^{-1}$) m/f	Vitamin A (mg retinal equiv. $\cdot day^{-1}$) m/f	Vitamin D ($\mu g \cdot day^{-1}$)	Vitamin K ($mg \cdot day^{-1}$) m/f
0 to <4 months	0.5	40	1	0.5	10	4
4 to <12 months	8	80	2	0.6	10	10
1 to <4 years	8	100	3	0.6	5	15
4 to <7 years	8	120	5	0.7	5	20
7 to <10 years	10	140	7	0.8	5	30

m/f = male/female.

Table 2.1 *(continued)* Schedule of energy and essential nutritional elements for infants according to age and gender

Age	Iron (mg · day^{-1}) m/f	Iodine (µg · day^{-1})	Zinc (mg · day^{-1}) m/f	Vitamin A (mg retinal equiv. · day^{-1}) m/f	Vitamin D (µg · day^{-1})	Vitamin K (mg · day^{-1}) m/f
10 to <13 years	12/15	180	9/7	0.9	5	40
13 to <15 years	12/15	200	9.5/7	1.1/1.0	5	50
15 to <19 years	12/15	200	10/7	1.1/0.9	5	70/60

Age	Thiamine (mg · day^{-1}) m/f	Riboflavin (mg · day^{-1}) m/f	Niacin (mg niacin equiv. · day^{-1}) m/f	Vitamin B6 (mg · day^{-1})	Folate (µg folate equiv. · day^{-1})	Vitamin B12 (µg · day^{-1})	Vitamin C (mg · day^{-1})
0 to <4 months	0.2	0.3	2	0.4	60	0.4	50
4 to <12 months	0.4	0.4	5	0.8	80	0.8	55
1 to <4 years	0.6	0.7	7	1.0	200	1.0	60
4 to <7 years	0.8	0.9	10	1.5	300	1.5	70
7 to <10 years	1.0	1.1	12	1.8	300	1.8	80
10 to <13 years	1.2/1.0	1.4/1.2	15/13	2.0	400	2.0	90
13 to <15 years	1.4/1.1	1.6/1.3	18/15	3.0	400	3.0	100
15 to <19 years	1.3/1.0	1.5/1.2	17/13	3.0	400	3.0	100

m/f = male/female.

Table 2.2 Energy data for carbohydrates, fats and proteins

Substance	Calorific value kcal (kJ)	Caloric equivalent $kJ \cdot l^{-1}$	Respiratory quotient
Carbohydrates	4.1 (17)	21.1	1.0
Fat	9.3 (38)	19.6	0.7
Protein	4.1 (17)	18.7	0.8

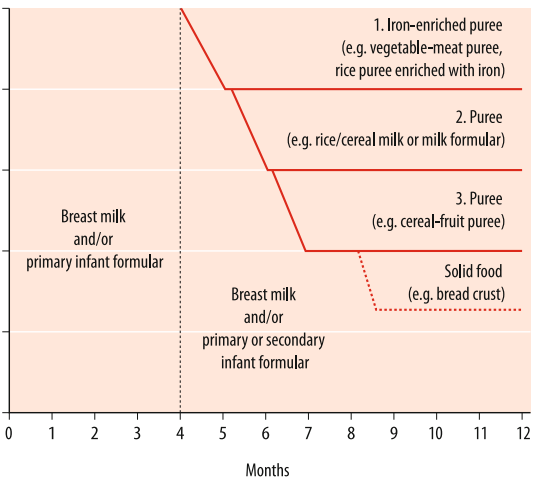


Fig. 2.1 Nutrition plan for the newborn in the first year

Definition

- Physiological calorific value = utilized energy per gram food
- Caloric equivalent = physiological caloric value per liter of used O_2
- Respiratory quotient = CO_2 release/O_2 consumption

Energy needed in the first year of life

- For the eutrophic newborn: 80–100 kcal · kg^{-1} · day^{-1}
- For the premature/hypotrophic newborn: 120–150 kcal · kg^{-1} · day^{-1}
- Calorific data for commonly available newborn formula milks are given in Table 2.3

Table 2.3 Caloric value of common newborn formulas per 100 ml

Initial milk	kcal	Antigen reduced milk	kcal	Newborn milk	kcal	Sequential milk	kcal
Pre Humana [a]	69	Humana HA	72	Humana 1 Humana 1 Baby-fit	72 72	Humana 2	75
Pre Aptamil [b]	67	Aptamil HA	67	Aptamil 1	72	Aptamil 2	75
Pre Milumil [b]	70			Milumil 1	74	Milumil 2	74
Hippon A [c]	71			Hippon 1	71	Hippon 2	77
Pre Aletemil [d]	71	Aletemil HA	67	Aletemil 1	72	Aletemil 2 plus	77
Aponti Pre [e]	71			Aponti 1	70	Aponti 2	77
Beba Pre [d]	71	Beba HA	67	Beba 1	72	Beba 2	78
Lactana A [f]	68			Lactana B	71		

Manufacturers: [a] Humana, Herford, [b] Milupa, Trowbridge, [c] HiPP, Newbury, [d] Nestlé, Vevey, [e] Aponti, Germany, [f] Töpfer, Dietmannsried.

Oral nutrition in cases of diarrhea and dysentery

- Humana HN (100 ml = 62 kcal, Herford)
- Alfaré (100 ml = 72 kcal; Nestlé, Vevey)

Vitamin D prophylaxis

- Newborns: 500 IU · day^{-1} as from the 10th day of life for 1 year (oral application)
- Premature newborns: 1000 IU · day^{-1} as from the 10th day of life for 1 year

Vitamin K prophylaxis

- Newborns: 2 mg vitamin K (oral application)
  - 1st day of life
  - 5th to 10th day of life
  - 4th to 6th week of life

Nasogastric or nasojejunal feeding

- Indication
  - In cases where feeding is required more than 10 times a day
  - To define the volume of milk remaining in the stomach
  - Swallowing difficulties (myopathies)
  - Malformations of the pharynx (clefts)
- Procedure
  - Measure distance: nose → earlap → neck → epigastrium = probe in stomach
  - Introduce the probe either through the nose (preferably) or the mouth
  - Insufflate air through the probe whilst auscultating the epigastrium, checking if the probe is in the stomach
- Use
  - Alimentation can be applied slowly through the probe according to individual demands
  - By hanging the probe below body level, the stomach is unburdened of fluids (ileus signs)
  - By hanging the probe above body level the stomach is disburdened, as elusion of air is possible leaving nourishment in the stomach

Enteral nutrition controlled by a pump system

- Indication
  - Newborns with intestinal stoma (jejunostomy/ileostomy) or colostomy
  - This enteral nutrition of the isolated intestinal section conditions it in order to reduce the effects of a non-used bowel

- Procedure (Fig. 2.2)
  - The prerequisite is the presence of a split stoma with openings that are close together
  - A special colostomy bag with an outlet is placed over both stomas
  - A probe (Charr 8) is placed in the distal branch and blocked (de-block twice a day for 2 h)
  - The stool can be continuously transferred from the colostomy bag to the distal bowel under the control of a pump system [Sondomat® (Fresenius, Bad Homburg, Germany) Kangaroo 2100]

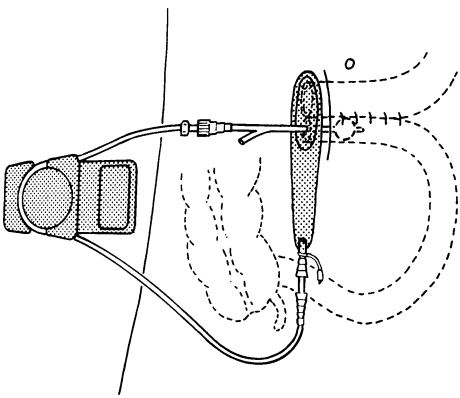


Fig. 2.2 Enteral nutrition via a pump

2.2 Vascular Access in Infants and Children

Peripheral venous access

In pediatric surgical patients a venous line is essential, as most such patients undergo a procedure under anesthesia. In comparison to adults, however, it is more difficult to achieve due to anatomical differences. In principle peripheral venous access (PVA; Fig. 2.3) is always preferable to a central line.

Central venous access

Central venous access (CVA) is essential for management of the majority of patients needing:

- Parenteral nutrition for more than 3 days
- Drug administration over a long period
- Regular blood sampling for diagnostic reasons

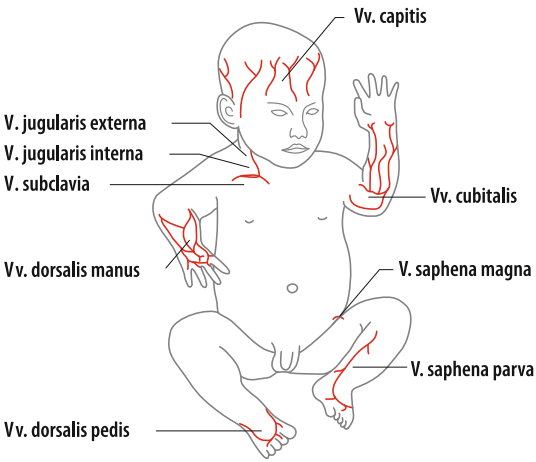


Fig. 2.3 Possible locations for peripheral and central lines in children

Oncologic patients in need of modern chemotherapy and intensive care patients frequently require massive fluid infusions as well as drugs that are cytotoxic to the vein wall. They are better utilized if diluted immediately in a large vessel with high blood flow. Central venous access allows continuous access to such large vessels, the most common being the superior cava vein (Fig. 2.4).

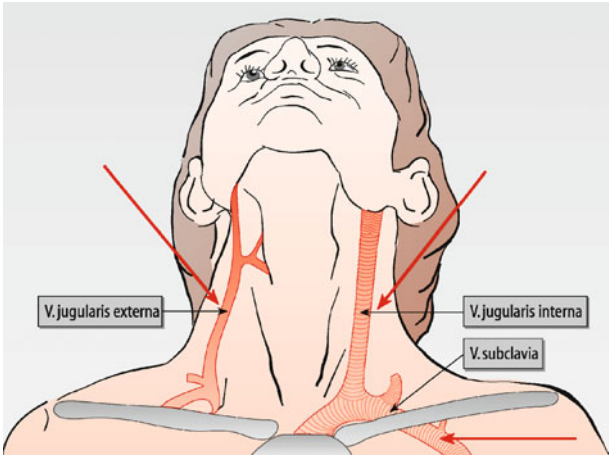


Fig. 2.4 Puncture sites of large veins for central lines

Venous access

- Table 2.4 details the indications, contraindications, siting, and complications of venous access

Table 2.4 The indications, contraindications, siting, complications and techniques of venous access

	Peripheral	Central
Indication	▪ Shock ▪ Hypovolemia ▪ Drug application	▪ Shock ▪ Total parenteral nutrition ▪ Infusion of hyperosmolar substances
Contraindications	None	Coagulation problems
Localization	See Fig. 2.3	See Fig. 2.4

Table 2.4 *(continued)* The indications for, and the contraindications, siting, complications and technique of venous access

	Peripheral	Central
Complications	▪ Infection ▪ Thrombophlebitis ▪ Tissue infiltration with necrosis	▪ Infection/sepsis ▪ Thrombosis ▪ Puncture of an artery ▪ Pneumothorax ▪ Infusiothorax (i.e., fluid entering the thoracic cavity after penetration of a large vessel in the thorax by the central line) ▪ Hemothorax ▪ Air embolism ▪ Pericardium tamponade
Technique	See text	See text

Insertion techniques for central vein lines

Catheters may be implanted either surgically (surgical technique) or by transcutaneous puncture (anesthetic technique). For both techniques either the vein, commonly the jugular internal or subclavian vein, is directly punctured, or the aforementioned large veins are reached by puncturing peripheral ones, e.g., cephalic, jugular external, facial vein, and occasionally the saphena magna vein. The distal tip of the catheter should be positioned in the superior cava vein just above the right atrium. Its location should be confirmed by X-ray or alternatively by ECG control (intra-atrial ECG; αv-Card®).

Surgical technique

- General anesthesia obligatory
- Coagulation status should be within normal range
- Skin incision and direct vein visualization
- Control of blood leakage by additional vascular sutures, e.g., "en pouch"
- Occlusion of major vessels should be avoided
- The proximal end of the catheter (vein entrance site) is tunneled to a more distant cutaneous (external) site. This can be performed either prior to accessing the vein or after, depending on the particular device used

- Technically easy when performed for the first time on the particular vein
- A reliable method for reaching the right position of the catheter tip
- A surgeon should be able to ensure CVA in every case and circumstance

Common veins used for the surgical technique

- Internal jugular vein in younger children, with bodyweight less than 15 kg
- Subclavian vein in older children
- The cephalic vein may be accessed surgically to reach the subclavian vein, and the external jugular to reach the superior cava vein

Anesthetic technique

- Veins are punctured transcutaneously
- Puncture with the patient under sedation is possible
- Location of the vein may be assured by ultrasonography
- Reduced bleeding risk (coagulation problems)
- Occlusion of the vessel possible
- Skin puncture site is close to the vein puncture site

Common veins used for the anesthetic technique

- Internal jugular vein in older children, with bodyweight more than 15 kg
  - Palpate the carotid artery; the internal jugular lies lateral
  - Puncture whilst palpating the carotid artery with the free hand
  - The puncture angle to the skin should be about 30°–45°
  - The needle tip should point in the direction of the sternoclavicular joint
  - Puncture of the right vein is preferable as its course is straighter
- External jugular vein
  - Localize the middle of the sternocleidomastoid muscle in the longitudinal direction
  - Turn the head to face the opposite direction whilst simultaneously performing a hyper flexion
  - By compressing the clavicula the vein becomes prominent
  - Puncture the vessel in its course

- Subclavian vein
  - Find the crossing point of the clavicula and the first rip
  - Puncture the skin under the clavicula and guide the needle behind the clavicula
  - The needle tip should point in the direction of the sternoclavicular joint
  - The subclavian vein runs ventrally to the subclavian artery
- Femoral vein
  - In the supine position the leg should be externally rotated and slightly abducted
  - Palpate the femoral artery; the femoral vein lies medial
  - Puncture at an angle of 45° in the direction of the inguinal ligament
- Umbilical vein
  - In the newborn the umbilical vein and arteries can be dissected
  - Anesthesia is not needed
  - Cut the umbilical cord with a scalpel preferably horizontally
  - The vessels have a typical arrangement (see Fig. 2.5)
  - Find the vein and clamp the arteries
  - Pass the catheter trough the vein and ligate

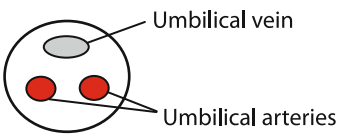


Fig. 2.5 Arrangement of umbilical vein and umbilical arteries in the umbilicus

Access in an emergency

In an emergency and when puncture of a vein is not possible, an intramedullary infusion via the tibia can be performed (Fig. 2.6):

- Use a thick cannula
- Puncture the ventral tibia into the medulla
- Fluids and pharmaceuticals can be applied
- **Note:** endotracheal application of pharmaceuticals is possible but not for volume substitution

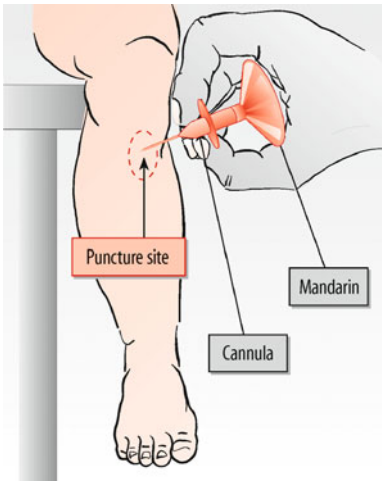


Fig. 2.6 Intramedullary puncture of the tibia as emergency access

Central line catheter types: advantages and disadvantages

There are many different types of catheters available. In general, they are classified as Broviac-type catheters and ports. Catheters may have one, two or three lumens.

- Ports are catheters consisting of a reservoir that is implanted totally under the skin. They can be accessed with a needle through the skin and through the port's perforable membrane. Only the needle provided by the manufacturer (Huber needle) should be used so as not to damage the membrane, resulting in a leak. The best location for the port seems to be the infraclavicular fossa. Ports are very comfortable, remain unexposed to infection when not in use, and allow activities such as swimming. The disadvantage is the fact that they do not allow massive fluid infusion due to the limited diameter of the puncture needle
- Broviac-type catheters, according to the author's experience, should be always tunneled subcutaneously along the thoracic wall, avoiding having the vein puncture site directly over the proximal exit site. It is recommended using catheters with cuffs, which stabilize the device and have anti-infection properties. The cuff should be located in the subcutaneous tissue 1–2 cm from the exit site. Broviacs may be more

susceptible to infections compared with ports, as there is a connection to the environment, and they are a little less comfortable because part of them hangs out of the body. Their big advantages, however, are that they allow massive infusion and are relatively cheap

Central line care

When using both catheter types maximal care must be taken to keep them uninfected and free of thrombosis. The parents and the patient need to be trained and guided in this. Disconnection of the catheter or puncturing of the port should be performed under sterile conditions regardless of whether blood is being sampled or drugs administered. Initially a small amount of saline must be injected to ensure that there is no thrombosis and that the device functions freely. The device must be flushed with 5 ml of heparin solution (20–100 IU · ml^{-1} of saline) directly after use and once a week. The amount of solution required increases with longer and thicker catheters.

Complications

- Both catheter types may occasionally have faults, but these are rare when they are gently handled
- Infections and thrombosis are well-known problems and when present indicate the need to remove the device and insert a new one. Some efforts may be undertaken to manage both problems, however, if not successful, they should not be continued. Treatment of infected catheters may only be carried out for a short time and only in cases of relatively non-pathogenic bacteria that are sensitive to antibiotics
- A particular problem of ports is that drugs might be administered subcutaneously when the needle is not sited properly in the port, resulting in extensive necrosis of the skin covering the port. Surgical intervention is then possible
- There are descriptions of a heart tamponade resulting from hard CVA placed too deeply, so keep in mind to locate the tip of the device in the superior vena cava or just above the atrium
- Ports should not be punctured within 5–7 days after implantation, to allow possible post-implantation hematomas, a potential source of infection, to be absorbed

2.3 Parenteral Nutrition

A prerequisite of parenteral nutrition is a central line. Parenteral nutrition is associated with increased morbidity and increased treatment costs.

Indication

- Reduced reserves and high demand
  - Premature newborns
  - Surgery for intestinal malformations
  - Necrotizing enterocolitis
  - Immature intestine
- Enteral nutrition is always preferable

Aim

- Energy and water supply
- Intestine relief

Basic demand

- Table 2.5 gives the nutritional requirements according to age for newborns, infants, and children

Table 2.5 Calculation of the composition for parenteral nutrition according to age for newborns, infants, and children

Age	Water ($ml \cdot g^{-1} \cdot day^{-1}$)	Glucose ($g \cdot day^{-1}$)	Fat ($g \cdot day^{-1}$)	Amino acids ($g \cdot day^{-1}$)	Energy ($kcal \cdot day^{-1}$)
1st day of life	50–70				80–100
2nd day of life	70–90				
3rd day of life	80–100				
4th day of life	100–120				
5th day of life	100–130				
1st year	100–140	8–15	2–3	1.5–2.5	60–100
2nd year	80–120	12–15	2–3	1.5	70–90
3rd to 5th year	80–100	12	1–2	1.5	60–70
6th to 10th year	60–80	10	1–2	1.0	50–60
11th to 14th year	50–70	8	1	1.0	50

- Table 2.6 gives the nutritional requirements according to weight for infants and children

Table 2.6 Calculation of the amount of parenteral nutrition according to weight for infants and children

Weight (kg)	Requirement ($ml \cdot kg^{-1} \cdot day^{-1}$)
<6	100
<16	80
<36	60
<56	50
>56	40

Correction of fluid balance

- Table 2.7 gives the fluid demand for premature and newborn children
- Correction of fluid balance is given in Table 2.8

Table 2.7 Fluid demand for premature and newborn children

Day post partum	Fluid required ($mg \cdot kg^{-1} \cdot day^{-1}$)			
	Premature newborn			Mature newborns
	<1000 g	1000–1500 g	>1500 g	
1	70	70	60	60
2	90	90	80	80
3	110	100	100	90
4	120	110	110	110
5–7	130	130	120	130
>7	150–180	140–170	130–160	130–160

Table 2.8 Correction of fluid balance

Increase amount of fluids		Decrease amount of fluids	
Fever	20%–30%	Mechanical ventilation	20%–30%
Phototherapy	20%	Incubator with high humidity >80%–100%	20%
Tachypnea	20%	Heart insufficiency	50–60 ml · kg^{-1} · day^{-1}
Loss of weight >15%	Variable, dependent on weight loss or urine output	Inadequate antidiuretic hormone secretion and kidney insufficiency (Na↓, K↓, edema, diuresis↓, Hct↓)	30 ml · kg^{-1} · day^{-1} + urine
Hypovolemia (urine production <0.5 ml · kg^{-1} · h^{-1})	Variable, dependent on weight loss or urine output		
Enhanced diuresis	Variable, dependent on weight loss or urine output		

Electrolyte basic demand

- The basic demand for electrolytes is detailed in Table 2.9

Table 2.9 Electrolyte basic demand

Electrolyte	Requirement (mmol · kg^{-1} · day^{-1})
Sodium	3–5
Potassium	1–3
Calcium	0.1–1–3[a]
Magnesium	0.1–0.7
Chloride	3–5
Phosphate	0.5–1–2.5[a]

[a]Growth demand for preterm newborns.

Selection of marketable infusion solutions for parenteral nutrition

Table 2.10 details the composition of a selection of commercial infusion solutions for parenteral nutrition, while Table 2.11 indicates the additions to the infusions required. The daily vitamin requirements are listed in Table 2.12.

Table 2.10 The composition of a selection of commercial infusion solutions for parenteral nutrition

Solution	Glucose ($g \cdot l^{-1}$)	Cations ($mmol \cdot l^{-1}$)				Anions ($mmol \cdot l^{-1}$)					Energy ($kcal \cdot l^{-1}$)	Osmolality ($mosmol \cdot kg^{-1}$)
		Na	K	Ca	Mg	Cl	Acetate	Lactate	Malate	Phosphate		
Glucoven ped 12.5% [a]	125	25	20			40					500	2100
Jonosteril ped I [a]	40	29.4	0.8	0.45		31.1					160	
Jonosteril ped II [a]	33.3	49.1	1.3	0.75		51.9					136	
Jonosteril ped III [a]	25	73.6	2.0	1.1		77.8					100	
Paediafusin I [b]	50	35	18	1	1.5	34	20		3	2	200	390
Thomaejonin ped I [c]	50	35	18	1	1.5	29	25		0.2	5	195	400
Paediafusin I [b]	50	70	18	1.5	2	64	26.5		3	2	200	390
Thomaejonin ped II [c]	50	70	18	1.5	2	60	32		0.2	5	195	460
Paediafusin op [b]	50	100		2	3	110					200	480
Thomaejonin ped op [c]	50	100		2	3	91	20				190	490
Tutofusin ped [b]	40	40	5		2.5	35	5		5		200	380

Manufacturers: [a] Fresenius, [b]Kabi Pharmacia, [c]Thomae.

Table 2.10 *(continued)* The composition of a selection of commercial infusion solutions for parenteral nutrition

Solution	Glucose ($g \cdot l^{-1}$)	Cations ($mmol \cdot l^{-1}$)				Anions ($mmol \cdot l^{-1}$)					Energy ($kcal \cdot l^{-1}$)	Osmolality ($mosmol \cdot kg^{-1}$)
		Na	K	Ca	Mg	Cl	Acetate	Lactate	Malate	Phosphate		
Ringer lactate [b]		130	5.5	1.8		112		27			7.5	
Ringer [b]		147	4	2.3		155					7	
Sterofundin		140	4	2.5	1	106		45			6.5	
Isot. NaCl solution		154				154						

Manufacturers: [a] Fresenius, [b]Kabi Pharmacia, [c]Thomae.

Table 2.11 Infusion additions (mmol · ml^{-1})

Solution	Na	K	Ca	Cl	PO_4
NaCl (10%)	1.71			2	
Na-glycero-PO_4	2				1
$NaPO_4$	1				0.6
KCl		1		1	
KPO_4		1			0.6
CaCl			0.5	1	
Ca-Glucon. (10%)			0.225		
Ca-Glucon. (20%)			0.45		
Human alb. (5%)	0.125	0.002	0.002	0.1	
Human alb. (20%)	0.125	0.002	0.002	0.1	
Biseko® (5%)			Like serum		

Parenteral nutrition plan

The aim of parenteral nutrition is to supply the child with the fluids and energy it needs. As a guideline 80–90 cal · kg^{-1} · day^{-1} is required consisting of:

- 40%–45% carbohydrates (glucose)
- 40% fat (Intralipid®, Fresenius)
- 10%–15% amino acids (Aminoped®, Fresenius)

Table 2.12 Vitamin demand per day

Vitamin	Age				
	Premature newborns	0–6 months	6–12 months	2–11 years	> 11 years
A (mg)	450	420	400	400–700	700
B_1 (mg)	0.7	0.3	0.5	0.7–1.2	1.1–1.4
B_2 (mg)	0.9	0.4	0.6	0.8–1.4	1.3–1.6
B_3 (mg)	11	6	8	9–16	15–18
B_6 (mg)	0.65	0.3	0.6	0.9–1.6	1.8
B_{12} (µg)	0.65	0.5	1.5	2–3	3
C (mg)	30	35	35	45	50
D (IU)	500	400	400	400	200
E (mg)	4.6	3–4	4–5	5–7	7
K ($\mu g \cdot kg^{-1}$)	15	15	15	15–30	10
Folic acid (µg)	90	30	45	100–300	400
Biotin (µg)	4	20	20	20–120	120
Panthenol (mg)	2	2	2	2–4	5
Iron ($\mu mol \cdot day^{-1}$)	1–2				
Zinc ($\mu mol \cdot day^{-1}$)	1–2–4.6 (premature newborns)				

Infants have a small blood volume, therefore it is important to ensure that the total infusion volume (e.g., blood, Intralipid®, albumin) is appropriate for the infant's weight and clinical condition.

Infusion plan

- A typical infusion plan is given in Table 2.13

Table 2.13 A typical infusion plan

Infusion (amount kg · day^{-1})	Initial volume ml (g)	On day	Enhancing volume ml · day^{-1}	Total volume ml (g) · day^{-1}
Glucose (10%) [a]	50 (5)	1	10	150 (15)
Intralipid® (I0%) [c]	5 (0.5) 10 (1) [b]	2 (1) 1 [b]	5 5 [b]	20 (2) 35 (3.5) [b]
+ Vitintra® (Fresenius) infant	1 ml	2	–	Maximum 4 ml
Aminoped® (Fresenius) (10%)	5 (0.5)	1	5 (<1500 g) 10 (>1500 g)	25–30 (2.5–3)
NaCl	2 mmol	2	Accordingly	
KCl	1 mmol	2	Accordingly	
Calcium gluconate	1–2 mmol	1	–	
Glucose-1-phosphate Na	1–2 mmol	3	–	
Soluvit N® (Fresenius)	1 ml	2		
Vitamin E	5 trps. p.o.	When oral feeding begins,		
Inzolen-Infantibus sine Na, K (Köhler, Alsbach, Germany)	0.5 ml	alternative 5–10 ml Biseko® per week		

[a] For blood glucose between 50 and 150 mmol · l^{-1}.

[b] For normal newborns.

[c] Intralipid® with Vitintra should be applied via separate lines.

Laboratory controls

Laboratory controls should not be carried out routinely but as indicated by the clinical findings and the disease. We propose the following plan for severely diseased children, newborns, and premature infants (Table 2.14).

Table 2.14 Proposed plan for laboratory controls for severely diseased children, newborns, and premature infants

Manifold (per day)	Once a day	Once a week
Blood gas Blood sugar Urine weight/density	Proteins Electrolytes Blood count Triglycerides Bilirubin	Creatinine Urea Transaminase Alkaline phosphatase Serum magnesium Serum phosphate

2.4 Dehydration/Rehydration

Indications

- Severe dehydration with circulatory insufficiency
- Continuing fluid loss
- Electrolyte imbalance (acidosis)
- Reduction in awareness
- Acute trauma
- Postoperatively

Aim

- Fluid sustainment (basic demand)/24 h
- Correction of the present deficit
- Cover abiding fluid losses

Signs and classification

- The signs and classification of dehydration are given in Tables 2.15 and 2.16

Table 2.15 The signs and classification of dehydration

	Degree of dehydration		
Sign	**Light**	**Moderate**	**Severe**
Weight loss			
▪ Infants	< 5%	5%–10%	10%–15%
▪ Children	< 3%	3%–6%	6%–9%
Skin turgor	Normal/↓	↓↓↓	↓↓↓ (vertical creases)
Skin color	Pallid	Pallid – gray	Grey, marmorated
Mucosa	Reduced moisture	Dry	Brittle
Fontanel	At level	Sunken	Prominent sunken
Pulse	Normal	↑	↑↑
Urine production	Normal	Oliguria	Anuria

Table 2.16 Type of dehydration (pathophysiological criteria)

	Dehydration type (loss relation: salt/H_2O)		
	Isotonic salt = water	**Hypotonic salt > water**	**Hypertonic salt < water**
Laboratory criteria			
▪ Osmolality (serum) mosmol · kg^{-1}	281–297	<281	>297
▪ Osmolality (urine)	↑	↑	↑
▪ Na (serum) mmol · l^{-1}	133–145	<133	>145
▪ Hematocrit	↑	↑	↑
▪ Urea (serum)	Normal – ↑	↑	↑
Signs			
▪ CNS	Apathy, lethargy	Seizure, unconsciousness	Seizure, irritability
▪ Skin turgor	↓	↓↓	Pasty
▪ Skin temperature	Cool	Cold, marmorated	Warm
▪ Pulse frequency/ quality	↑/Soft	↑↑↑/amplitude ↓↓	↓/normal
Causes	Diarrhea, vomiting	Salt deficit syndrome: sweating in mucoviscidosis (cystic fibrosis) Cholera	Diabetes insipidus (renal, central) Diarrhea Fluid deficit

Oral rehydration

- A scheme for oral rehydration is shown in Fig. 2.7

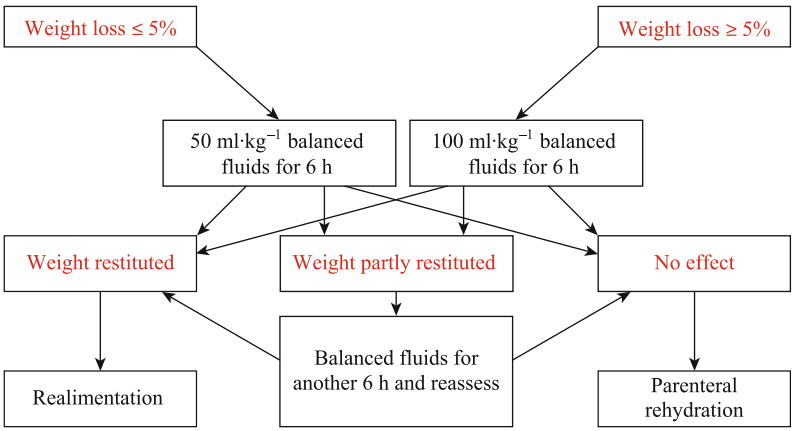


Fig. 2.7 Scheme for oral rehydration

Oral rehydration solutions

- Table 2.17 gives details on various commercial oral rehydration solutions

Table 2.17 Details of various commercial oral rehydration solutions

Composition	Carbohydrates: glucose					
	ESPGAN[a] Recommendation	Elotrans (Fresenius)	GES 60 (Milupa)	GES 45 (Milupa)	Humana electrol. (Humana)	Oralpaedon (Fresenius)
Osmolarity (mosmol·l^{-1})	200-250	311	270	298	215	377
Sodium (mmol·l^{-1})	60	90	60	45	46	30
Potassium (mmol·l^{-1})	20	20	20	25	35	20
Chloride (mmol·l^{-1})	≤25	80	50	25	45	30
Bicarbonate (mmol·l^{-1})	–	–	30	23	–	20
Citrate (mmol·l^{-1})	10	10		9	12	–
Glucose (g·l^{-1})	13.3–20.0	20	19.8	19.6	18	49.9

Composition	Carbohydrates: glucose + oligosaccharides + starch		
	ORS 200 Carrot-rice gruel (Hipp)	Reis gruel/electrolyte diet (Töpfer)	RES 55 (Milupa)
Osmolarity (mosmol·l^{-1})	265	220	210
Sodium (mmol·l^{-1})	57	55	55
Potassium (mmol·l^{-1})	22	30	35

[a] ESPGAN: European Society of Pediatric Gastroenterology and Nutrition.

Table 2.17 *(continued)* Details of various commercial oral rehydration solutions

Composition	Carbohydrates: glucose + oligosaccharides + starch		
	ORS 200 Carrot-rice gruel (Hipp)	Reis gruel/electrolyte diet (Töpfer)	RES 55 (Milupa)
Chloride ($mmol \cdot l^{-1}$)	45	60	55
Bicarbonate ($mmol \cdot l^{-1}$)	–	25	–
Citrate ($mmol \cdot l^{-1}$)	5	–	10
Carboxylases ($g \cdot l^{-1}$), incl. glucose ($g \cdot l^{-1}$)	42 14	46 5	51 10.8

[a] ESPGAN: European Society of Pediatric Gastroenterology and Nutrition.ww

Parenteral rehydration

- Fluid replacement (Table 2.18) and electrolyte replacement (Table 2.19) regimens are followed during parenteral rehydration

Electrolyte content in different body fluids is given in Table 2.20 and schedules to correct it during rehydration are outlined in Table 2.21. The appropriate buffering is calculated using Eq. 2.1.

(Eq. 2.1)

pH < 7.15 and/or bicarbonate <12 mmol•l^{-1}
[15 – actual bicarbonate] × mass (kg) × 0.6 = ml 8.4% bicarbonate solution or Base excess × 0.3 × mass (kg)

Table 2.18. Fluid replacement regimen (per 24 h)

Basic demand	Deficit	Estimated losses	
Age dependent	Weight loss of	i.e., Diarrhea	
		Stool consistency	Water fraction
see Tables 2.5, 2.7, 2.8	5% → 50 ml	▪ Formed	< 80%
	10% → 100 ml	▪ Mushy	Approx. 85%
	15% → 150 ml	▪ Liquid	Approx. 90%
		▪ Aqueous	> 90%

Table 2.19 Electrolyte replacement regimen (per 24 h)

Electrolytes	Sustainment demand	Deficit + actual losses Substitute according to the formula
Na (mmol·l^{-1})	2–3: ≤2 years 1–2: >2 years	140 (Serum-Na) × body weight (in kg) × 1.2 = mmol·l^{-1}
K (mmol·l^{-1})	1: days 2–7 2–3: ≤ 2 years 1–2: > 2 years	Serum-K and pH (see Fig. 2.8) Maximal daily dose 8 mmol; begin with 0.1 mmol·h^{-1} when urine production starts

Table 2.20 Electrolyte content of various body fluids

	Na^+	K^+	Cl^-	HCO_3^-
	($mmol \cdot l^{-1}$)			
Sweat	30	10	30	
Saliva	15	30	30	
Stomach fluids	50	10	120	
Pancreatic secretions	140	7	120	30
Small bowel secretions	100	5	100	
Stool	70	40	70	30

Table 2.21 Rehydration schedule to correct fluid and electrolyte imbalances

Independent of the dehydration type apply an initial volume of isotonic solution at 20 $ml \cdot kg^{-1}$ in 1 h (i.e., Ringer lactate)			
Hour	**Isotonic dehydration**	**Hypotonic dehydration**	**Hypertonic dehydration**
2–24	$^1/_2$ isotonic solution (i.e., Jonosteril ped III)	0.9% NaCl solution 50% glucose (5 $ml \cdot kg^{-1}$)	$^1/_2$ isotonic solution
24 onwards	$^1/_3$ isotonic solution (i.e., Jonosteril ped II)		$^1/_3$ isotonic solution
Correction after laboratory examination of the blood			

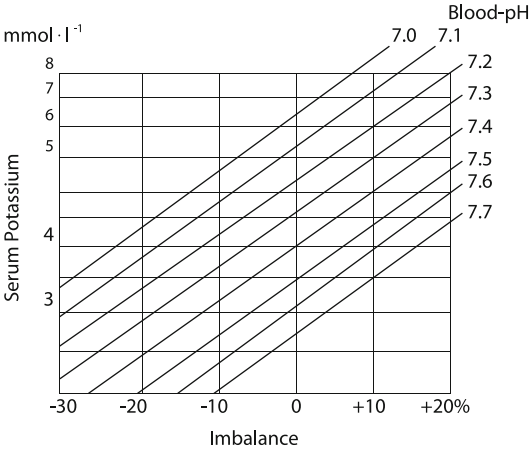


Fig. 2.8 Nomogram for calculating potassium imbalance

2.5 Antibiotics

- Table 2.22 lists the most common oral antibiotics used in pediatric surgery. The measuring units are the smallest that are available on the market
- Antibiotics should not be used at random but after an antibiogram, which should be acquired as soon as possible after the infection has been verified
- Preferably oral antibiotics should be used, as morbidity is lower than with parenteral antibiotics (Table 2.23) and the price is as low as 15-fold compared with parenteral treatment
- The dosage regimens are given in Table 2.24

Table 2.22 Oral antibiotics (groups in alphabetical order). (*btl* Bottle, *ms* measuring spoon, *tab* tablet, *vl* vial)

Generic name	Measuring unit	Daily dose ($mg \cdot kg^{-1}$)	Application per day
Amino benzyl penicillin			
Amoxicillin	1 ml = 100 mg	50–100	3–4 × p.o.
Ampicillin	1 ml = 70 mg	100–150	3 × p.o.
Bacampicillin	1 tab = 400 mg	25–50	2–3 × p.o.
Anthelminthics			
Pirantel	1 ml = 50 mg	2.5–10 ml	1 × p.o.
Mebendazol	1 tab = 100 mg	2 × 1 tab	On 3 days
Niclosamide	1 tab = 500 mg	2–4 tab 1–2 tab	On 1st day as from 2nd day
Albendazole	1 tab = 400 mg	1 tab > 2 years ½ tab < 2 years	1 × p.o. for 3 days
Antimycotics			
Amphotericin B	1 bag = 50 mg	10	2–4 × inhalation
	1 bag = 100 mg		2–4 × p.o. local
Clotrimazole	1 ml = 100 mg		Local
Fluconazole	1 tab = 50 mg	Superficial mycosis: 1–2 Systemic mycosis: 3–6	1 × p.o.
Miconazole	1 tab = 250 mg	20	4 × p.o.
Ketoconazole	1 tab = 200 mg	2.5–5	1 × p.o.
Nystatin	1 ml = 100000 E	4 × 0.5 ml	4 × p.o.
Voriconazole	1 tab = 50 mg 1 tab = 200 mg	6 initially 2 ×; 4 afterwards	2 × p.o.
Cephalosporins			
Cefaclor	1 ml = 25 mg	20–40	3–4 × p.o.
Cefalexin	1 ml = 50 mg	25–100	3–4 × p.o.
Cefixim	1 ml = 20 mg	8–12	2 × p.o.
Cephadroxil	1 ml = 50 mg 1 tab = 500 mg	50–100	2 × p.o.

Table 2.22 (*continued*) Oral antibiotics (groups in alphabetical order). (*btl* Bottle, *ms* measuring spoon, *tab* tablet, *vl* vial)

Generic name	Measuring unit	Daily dose ($mg \cdot kg^{-1}$)	Application per day
Combinations			
Amoxicillin + Clavulanic acid	1 tab = 500 mg	37.5 20	3 × p.o.
Co-trimoxazole Trimethoprim (T) Sulfamethoxazole (S)	1 tab = T 20 mg S 100 mg 1 ml = T 8 mg S 40 mg	5(–20) T 25(–100) S	3–4 × p.o.
Macrolides			
Clarithromycin	1 ml = 25 mg	15	2–3 × p.o.
Clindamycin	1 ms = 75 mg	10–20	3–4 × p.o.
Erythromycin	1 ml = 250 mg	30–50	2–4 × p.o.
Josamycin	1 ms = 150 mg	30–50	3–4 × p.o.
Roxithromycin	1 btl = 50 mg	15–20	2 × p.o.
Spiramycin	1 tab = 0.75×10^6	80,000 IU	4× p.o.
Oxazolidinones			
Linezolid	1 tab = 600 mg 1 ml = 20 mg	10 35 (>12 years)	3 × i.v. 2 × i.v.
Penicillin			
Azidocillin	1 tab = 750 mg	60–120	3 × p.o.
Penicillin V	Variable	2×10^5– 4×10^6 IU	4–6 × p.o.
Propicillin	1 ml = 1 mg	2×10^5–1.6×10^6 IU	4 × p.o.
Penicillinase-resistant penicillins			
Flucloxacillin	1 bag = 5g	20–80	2–3 × p.o.
Sulfonamides			
Sulfadiazine	1 tab = 500 mg	100	1 × p.o.
Sulfasalazine	1 tab = 500 mg	30–40–60	3 × p.o.

Table 2.22 (*continued*) Oral antibiotics (groups in alphabetical order). (*btl* Bottle, *ms* measuring spoon, *tab* tablet, *vl* vial)

Generic name	Measuring unit	Daily dose ($mg \cdot kg^{-1}$)	Application per day
Tetracyclines (only for children >8 years)			
Doxycycline	1 tab = 100 mg	First day: 4 as from second day: 2	2× p.o.
Minocycline	1 tab = 50 mg	First day: 4 as from second day: 2	2× p.o.
Tuberculostatics			
Ethambutol	1 tab = 300 mg	20–25	1× p.o.
Isoniazid	1 tab = 100 mg	5–10	1× p.o.
Pyrazinamide	1 tab = 500 mg	30–40	1× p.o.
Rifampicin	1 tab = 150 mg	10–15	1–2× p.o.
Virostatics			
Brivudin		5	3× p.o.
Zidovudine	1 ml = 10 mg	3.5	6× p.o.
Aciclovir	1 tab = 200 mg	Single dose 1 tab Single dose ½ tab < 2 years	2–5× p.o.
Other antibiotics			
Metronidazole	1 tab = 250 mg	20–30	2–3× p.o.
Nitrofurantoin	1 ml = 58 mg	Days 1–6: 5 as from day 7: 2.5	2× p.o.

Table 2.23 Parenteral antibiotics (groups in alphabetical order). (*btl* Bottle)

Generic name	Measuring unit	Daily dose ($mg \cdot kg^{-1}$)	Application
Acylamino penicillins			
Apalcillin	1 btl = 1 g	200	3–4× i.v.
Azlocillin	1 ml = 50 mg	200–300	4× i.v.
Mezlocillin	1 ml = 100 mg	200–300	4× i.v.
Piperacillin	1 btl = 1 g	100–300	4× i.v.
Aminobenzyl penicillins			
Ampicillin	Variable	100–300	3–4× i.v.
Aminoglycosides			
Amikacin	1 btl = 100 mg	10–15	2× i.v.
Aztreonam	1 btl = 500 mg	45–120	3–4× i.v.
Gentamicin	1 ml = 5 mg	4–7.5	2–3× i.v.
Netilmicin	1 Amp = 15 mg	4–5	2–3× i.v.
Tobramycin	1 ml = 10 mg	4–7.5	2–3× i.v.
Antimicotics			
Voriconazole	1 ml = 6.66 mg	6 initially 2×; 4 afterwards	2× i.v.
Carbapenem			
Imipenem	1 btl = 500 mg	50–80–100	4× i.v.
Meropenem	1 btl = 500 mg 1 btl = 1000 mg	10–40	2× i.v.
Cephalosporins			
Cefamandol	1 ml = 50 mg	100–150	3–4× i.v.
Cefazedone	1 btl = 1 g	50–100	2–3× i.v.
Cefazolin	1 btl = 1 g	50–100	2–3× i.v.
Cefmenoxime	1 btl = 500 mg	50–100	2–3× i.v.
Cefoperazone	1 btl = 5 g	50–80	2–3× i.v.
Cefotaxime	1 ml = 250 mg	50–100–150	3× i.v.
Cefotetan	1 btl = 1 g	50–80	1–2× i.v.
Cefotiam	1 btl = 500 mg	50–100	2–3× i.v.

Table 2.23 (*continued*) Parenteral antibiotics (groups in alphabetical order). (*btl* Bottle)

Generic name	Measuring unit	Daily dose ($mg \cdot kg^{-1}$)	Application
Cefoxitin	1 ml = 66 mg	50–100	3× i.v.
Cefsulodin	1 btl = 1 g	50–80	3× i.v.
Ceftazidime	1 btl = 0.5 g	30–100	2–3× i.v
Ceftizoxime	1 btl = 0.5 mg	50–100	2–3× i.v.
Ceftriaxone	1 btl = 500 mg	50–80	1–2× i.v.
Cefuroxime	1 ml = 50 mg	30–100	3–4× i.v.
Combinations			
Amoxicillin + Clavulanic acid	1 btl = 275 mg	27.5 20	4× i.v.
Ampicillin + Sulbactam	1 btl = 750 mg	150	3–4× i.v.
Ticarcillin + Clavulanic acid	1 btl = 1.6 g	80–260	2–3× i.v.
Co-trimoxazole	1 ml = 16 mg T 1 ml = 80 mg S	5(–20) T 25(–100) S	3–4× i.v.
Glycopeptides			
Aciclovir	1 btl = 250 mg	250–500 $mg \cdot m^{-2}$	3× i.v.
Amphotericin B	1 btl = 50 mg	0.1–0.3–1.0	1× i.v.
Ethambutol	1 btl = 400 mg	20–25	1× i.v.
Fluconazole	1 btl = 100 mg	Superficial mycosis: 1–2 Systemic mycosis: 3–6	1× i.v.
Flucytosine	1 btl = 500 mg	150	1× i.v.
Ganciclovir	1 btl = 500 mg	5–10	2× i.v.
Miconazole	1 ml = 10 mg	15–20	1× i.v.
Streptomycin	1 btl = 1 g	10–20–30	3× i.v.
Teicoplanin	1 btl = 100 mg	First dose 6, 3–6	1× i.v.
Vancomycin	1 btl = 500 mg	20–30–40	2–3× i.v.
Zidovudine	1 btl = 200 mg	2.5	6× i.v.

Table 2.23 (*continued*) Parenteral antibiotics (groups in alphabetical order). (*btl* Bottle)

Generic name	Measuring unit	Daily dose ($mg \cdot kg^{-1}$)	Application
Macrolides			
Clindamycin	1 ml = 150 mg	10–20	3–4 × i.v.
Erythromycin	1 btl = 500 mg	30–50	2–4 × i.v.
Metronidazole	1 btl = 500 mg	20–30	2–3 × i.v.
Oxazolidinones			
Linezolid	1 bag = 600 mg	10 35 (>12 years)	3 × i.v. 2 × i.v.
Penicillins			
Benzylpenicillin	1 btl = 0.5×10^6 IU	(25×10^3)–(25×10^6) IU	4–6 × i.v.
	1 btl = 1.2×10^6 IU	$(1–1.5) \times 10^6$ IU	1 × i.v.
Penicillinase-resistant penicillins			
Flucloxacillin	1 btl = 250 mg	50–100	4–5 × i.v. Maximal dose: 33 $mg \cdot kg^{-1}$ application
Oxacillin	1 btl = 500 mg	20	3–4 × i.v.
Tetracyclines (only for children > 8 yrs)			
Minocycline	1 btl = 200 mg	First day: 4 as from second day: 2	2 × i.v.
Other antibiotics			
Fosfomycin	1 btl = 2 g	50–80	2–3 × i.v.
Fusidin acid	1 btl = 500 mg	20	3 × i.v.

Table 2.24 Antibiotic dosage in neonatal period (groups in alphabetical order)

Generic name	Newborn <7 days		Newborn >7 days	
	Dose ($mg \cdot kg^{-1}$) (per day)	Number of dose (per day)	Dose ($mg \cdot kg^{-1}$)	Dosages (per day)
Aminoglycoside				
Amikacin [a]	15	2	20	3
Gentamicin [a]	5	2	7.5	3
Netilmicin [a]	5	2	6	3
Tobramycin [a]	4	2	6	3
Cephalosporins				
Cefotaxime	100	2	150	3
Ceftazidime	25–50	2	25–60	2
Cefuroxime	30–100	2	30–100	2–3
Penicillins				
Ampicillin	50 –100	2	100–200	3–4
Flucloxacillin	40–80	2	40–100	3–4
Mezlocillin	150	2	225	3
Penicillin G	50,000–100,000 IU	2	100,000 IU	3
Piperacillin	150–200		200–300	3
Other antibiotics				
Imipenem	50	2	50–75	2–3
Isoniazid	5–10	1	5–10	1
Metronidazole	15	2	15	2
Vancomycin	2–30	2	30–45	3

[a] Premature ≤1500 g ½ day dose, 1500–2000 g ¾ day dose as initial doses.

Implicitly determine the serum level after 2 days.

3 Emergencies/Resuscitation

3.1 Systemic Infections in Newborns

Routes of infection

- Intrauterine ascending or perinatal infections
  - Listeriosis
  - Lues (syphilis)
  - Cytomegaly
  - Toxoplasmosis
  - Human immunodeficiency virus (HIV)
  - B-streptococcus
  - Hepatitis B, C
  - *Chlamydia*
- Catheters, wounds, airway tubes
  - *Staphylococcus epidermidis* and *S. aureus*

Clinical signs

- Clinical signs are given in Table 3.1

Table 3.1 Clinical signs of systemic infection in a newborn

	Signs
General signs	Feeding problems, lethargy, hypothermia or fever
Heart/circulation	Tachycardia >160 beats/min, hypotension, centralization, cold extremities
Breathing/respiration	Dyspnea, apnea attacks, intercostal retractions, acidosis
Skin	Pale, icteric, cyanotic, marbled skin, petechiae
Gastrointestinal	Abdominal distension, gastric stasis, diarrhea, dysphagia
Nervous system	Apathy, irritability, convulsion
Local	Pustules, omphalitis, mastitis, paronychia

Investigations

- Blood counts
  - Leukocytosis or leukocytopenia
  - Thrombocytopenia
  - C-reactive protein (CRP) can be high due to birth stress
- Infection detection in the mother
  - Vaginal swab
  - Placenta swab
- Infection detection in the child
  - Blood culture
  - Respiration tube swab
  - Urine culture
  - Gastric fluid culture
  - Stool culture
  - Ear and eye swab
  - Liquor culture (in special cases)
- Imaging procedures
  - Chest X-ray
  - Skull ultrasonography
  - Abdominal ultrasonography
  - Abdominal X-ray (suspicion of necrotizing enterocolitis)

3.2 Shock

Definition

- Clinical syndrome with cellular metabolic deficiency as a result of tissue hypoperfusion

3.2.1 Hypovolemic Shock

Definition

- Shock from loss of circulating volume

Common etiology

- External or internal hemorrhage
- Burns
- Dehydration (gastroenteritis, diabetes, ileostomy)

Signs of hemorrhage

- Signs of hemorrhage are listed in Table 3.2

Table 3.2 Signs of hemorrhage

	Blood loss <25%	Blood loss 25%–40%	Blood loss >40%
Heart rate	Tachycardia	Tachycardia	Tachycardia/bradycardia
Blood pressure	Normal	Normal or hypotensive	Hypotensive
Pulse pressure	Normal or reduced	Reduced	Reduced
Capillary refill time	Prolonged	Prolonged	Prolonged
Extremities	Cold and pale	Cold and pale	Cold and pale
Breathing	Tachypnea	Tachypnea	Gasping
Diuresis	Oliguric	Oliguric or anuric	Anuric
Consciousness level	Irritable	Lethargic	Coma, responds only to pain

Therapy

- Volume substitution fast and pre-warmed (colloid or crystalloid, $20\,ml \cdot kg^{-1}$)
- If no improvement, second bolus of $20\,ml \cdot kg^{-1}$
- If no improvement (hemorrhagic shock), blood $20\,ml \cdot kg^{-1}$
- Intubation and mechanical ventilation
- Resuscitation

3.2.2 Septic Shock

Definition

- Shock from maldistribution caused by microorganisms or their products

Signs

- Table 3.3 lists signs of septic shock

Table 3.3 Signs of septic shock

	Compensated shock	Uncompensated shock
Heart rate	Tachycardia	Tachycardia
Blood pressure	Normal	Hypotensive
Pulse pressure	Wide	Narrow
Capillary refill time	Prolonged	Prolonged
Extremities	Warm	Warm or cool
Breathing	Tachypnea	Respiratory insufficiency
Mental state	Anxious, confused	Lethargic

Investigations

- Blood analysis
  - White blood cell count with differentiation
  - Thrombocytes
  - CRP
- Lumbar puncture (cells, bacteria, glucose, protein)
- Cultures
  - Ear
  - Wounds
  - Umbilicus
  - Blood (temperature >38.5°C)
  - Stomach
  - Tracheal aspirate
  - Urine

- Feces
- Cerebrospinal fluid
- Arterial blood in suspected *Candida* infection

- Chest X-ray and abdominal X-ray (if abdominal signs)
- Abdominal ultrasonography or computer tomography in suspected abdominal sepsis

Hematological data

- Normal hematological values (mean values) are given in Table 3.4

Table 3.4 Normal hematological values (means)

	Age				
	1 week	6 months	1 year	6 years	14 years
White cells (total) (10^6/ml)	12.2	11.9	11.4	8.5	7.8
Neutrophils (%)	45	32	31	51	57
Lymphocytes (%)	41	61	61	42	35
Monocytes (%)	9	5	5	5	3
Eosinophils (%)	4	3	3	3	3
Thrombocytes (10^6/ml)	300	300	300	300	300

Therapy

- Oxygen supply (mechanical ventilation may be needed)
- Volume substitution (mainly colloid, 20 ml · kg^{-1}; up to four boluses may be needed)
- Correct hypoglycemia
- Antibiotic treatment
  - Initiated as soon as possible in suspected sepsis
  - The recommended antibiotic therapy (and doses) for sepsis of unknown etiology varies among different age groups and institutions
  - It depends on the most likely causative organisms and antibiotic resistance
  - The antibiotic regimen is changed on the basis of culture results or after 2–3 days if it does not have the desired effect

- Inotropic drugs (dopamine, dobutamine)
  - Due to sepsis-associated depression of myocardial function, inotropic support is started together with the second fluid bolus
- Correct acidosis (bicarbonate)
- Suggested antibiotics to prescribe in suspected sepsis are given in Table 3.5

Table 3.5 Suggested antibiotics to prescribe in suspected sepsis

Patients	Antibiotics
Neonates 0–28 days old	Amoxicillin and aminoglycoside
Older infants and children	Amoxicillin/clavulanate and aminoglycoside or third generation cephalosporin

3.2.3 Cardiogenic Shock

Definition

- Shock from decreased cardiac function

Common etiology

- Myocarditis
- Cardiomyopathy
- Congenital malformations
- Arrhythmias

Signs

- Tachycardia
- Normal or reduced blood pressure
- Reduced pulse pressure
- Prolonged capillary refill time
- Cold and pale extremities
- Tachypnea, dyspnea (pulmonary edema)
- Oliguria
- Irritable to coma

Therapy

- Oxygen supply (may need mechanical ventilation)
- Decrease oxygen consumption (treatment of pain and hyperthermia, sedation)
- Optimize (reduce) circulatory volume (e.g., furosemide)
- Vasodilatation (nitroprusside, nitroglycerin, enoximone)
- Improve cardiac contractility (inotropic medication, reduce acidosis)

3.2.4 Anaphylactic Shock

Definition

- Shock from an acute hypersensitivity reaction caused by an allergen

Common etiology

- Medication (penicillin, radiographic contrast)
- Proteins
- Polysaccharide

Signs

- Signs of anaphylactic shock are shown in Table 3.6

Table 3.6 Signs of anaphylactic shock

Stage I	Stage II	Stage III	Stage IV
▪ Itching ▪ Nausea ▪ Light-headedness ▪ Abdominal pain ▪ Flushing ▪ Edema (facial swelling) ▪ Wheezing	▪ Dyspnea ▪ Tachycardia ▪ Sweating ▪ Pallor ▪ Vomiting ▪ Diarrhea	▪ Bronchospasm ▪ Difficulty breathing ▪ Hypotension ▪ Collapse ▪ Fecal incontinence	▪ Respiratory arrest ▪ Cardiac arrest

Therapy

- Remove allergen
- Oxygen supply
- Intubation or tracheostomy
- Volume replacement (colloid, 20 ml · kg^{-1})
- Medication
  - Antihistamine (diphenhydramine i.m. or slow i.v.)
  - Adrenaline (10 µg · kg^{-1} i.m., 5 mg 1:1000 nebulized)
  - Hydrocortisone 4 mg · kg^{-1} i.v.
  - Aminophylline, salbutamol

3.3 Hypoglycemia

Definition

- Blood glucose level below
  - 40 mg · dl^{-1} in children
  - 30 mg · dl^{-1} in neonates during the first week of life
  - 20 mg · dl^{-1} in prematures

Common etiology

- Decreased glycogen stores (preterm neonate)
- Increased glucose demand (hypothermia, infection, respiratory distress)
- Diabetic mother
- Islet cell adenoma

Signs

- Bradycardia and tachycardia
- Apnea periods
- Hypothermia
- Irritability, lethargy
- Sweating
- Vomiting
- Convulsion

Therapy

- If unconscious, intubate; resuscitate if necessary
- Serum glucose measurement
- Glucose 500 mg · kg^{-1} i.v. (5 ml · kg^{-1} glucose 10%) bolus
- Glucose infusion (glucose 5 mg · kg^{-1} · min^{-1})
- Tests to assess the cause of hypoglycemia (neonates and particularly prematures often present with hypoglycemia in other disorders such as infections)

3.4 Acid–Base Balance

Definition

- Metabolic acidosis: primary decrease in HCO_3^- concentration
- Metabolic alkalosis: primary increase in HCO_3^- concentration
- Respiratory acidosis: primary increase in arterial PCO_2
- Respiratory alkalosis: primary decrease in arterial PCO_2
- Estimated normal values are given in Table 3.7

Table 3.7 Estimated normal values in arterial blood

Age	PO_2 (mmHg)	pH	PCO_2 (mmHg)	Standard bicarbonate (mmol · l^{-1})
Birth (umbilical artery)	12.1–19.7	7.18–7.30	43.3–54.9	16.9–20.5
5–10 min	39.7–59.5	7.15–7.25	39.1–53.1	15.1–18.3
30 min	42.6–65.6	7.25–7.33	32.0–43.4	16.7–19.7
60 min	52.0–74.6	7.30–7.36	31.9–40.3	18.0–20.4
24 h	63.2–82.2	7.33–7.39	33.3–36.5	18.9–21.5
7 days	63.4–82.8	7.35–7.40	32.8–39.0	20.5–23.1
2 years	80–100	7.35–7.40	32–39	20–22
>2 years	80–100	7.35–7.45	35–45	22–24

Normal values of base excess (age independent) = –2.5 ± 2.5.

Diagnosis

- Diagnosis based on acid–base balance data is given in Table 3.8

Table 3.8 Interpretation of acid–base balance data

	pH	PCO_2	Bicarbonate
Metabolic acidosis	↓	= ↓	↓[a]
Metabolic alkalosis	↑	= ↑	↑[a]
Respiratory acidosis	↓	↑[a]	= ↑
Respiratory alkalosis	↑	↓[a]	= ↑

[a] Primary changes.

Etiology

- **Metabolic acidosis**
  - Increased endogenous acid production
    - Lactic acidosis (e.g., hypoxia, hypothermia, shock)
    - Diabetic ketoacidosis
    - Alcoholic ketoacidosis
    - Starvation
  - Ingestion of toxins
    - Salicylates
    - Paraldehyde
    - Methanol, ethylene glycol
  - Acute and chronic renal failure
  - Loss of HCO_3^-
    - Diarrhea
    - Renal failure
    - Treatment with acetazolamide
    - Ileus
    - Pancreatic fistula
- **Metabolic alkalosis**
  - Loss of acid
    - Gastrointestinal (vomiting)
    - Renal (hypocalcemia, loop diuretics, Cushing syndrome, primary hyperaldosteronism)

- Excessive HCO_3^- administration
  - Milk-alkali syndrome
  - Parenteral HCO_3^-
- Contraction alkalosis
  - Sweating (cystic fibrosis)
  - Massive diuretics

- **Respiratory acidosis**
  - Neuromuscular disease
    - Central nervous system disorders
    - Peripheral nervous system disorders
    - Muscular disorders (myopathies)
  - Cardiovascular disorders (patent ductus arteriosus, ventricular septal defect)
  - Respiratory disease
    - Upper airway obstruction (e.g., laryngitis, epiglottitis)
    - Impaired lung motion (pneumothorax)
    - Lower airway diseases (asthma, aspiration, pneumonia, acute respiratory distress syndrome (ARDS), cystic fibrosis)
- **Respiratory alkalosis**
  - Hyperventilation (psychogenic, mechanical, central nervous system disorders)

Therapy

- Treatment of acid-base imbalance is given in Table 3.9

Table 3.9 Treatment of acid–base imbalance

pH	Treatment
<7.15	Immediate infusion of 2.5 ml · kg^{-1} of 8.4% bicarbonate (add to glucose solution 1:4 to reduce osmolarity) Treatment of underlying cause
7.15–7.25	Correction of base deficit Treatment underlying cause
7.25–7.55	Treatment underlying cause
pH >7.65	L-Arginine-HCl (slow infusion over hours) HCl (100 mEq $H^+ \cdot l^{-1}$); infuse half of calculated dose through central line

Calculation of acid–base correction

- mmol bicarbonate = base deficit × weight (kg) × 0.6
  (only 50% of the calculated base deficit should be corrected at a time)
- mmol HCl = base excess × weight (kg) × 0.6
  (only 50% of the calculated base excess should be corrected at a time)

3.5 Resuscitation

Clinical assessment of children and infants

A, B, C, D, E assessment

(for normal values see Table 3.10; for neonatal assessment see Table 3.11)

Airway and Breathing

- Effort of breathing (recessions, grunting, accessory muscle use)
- Rate, thoracic excursions
- Stridor/wheeze
- Chest auscultation
- Skin color (pink, pale, cyanosis)

Circulation

- Heart rate (infants: brachial or inguinal artery, children: carotid artery)
- Pulse volume
- Capillary refill (normally less than 2 s)
- Peripheral skin temperature
- Urinary output

  Start cardiopulmonary resuscitation if necessary!

Disability

- Assess consciousness (alert, verbal reaction, reaction to pain, unconscious)
- Pupils (size and reaction)
- Posture

Exposure/Environmental control

- Body check
- Body temperature (avoid hypothermia)

Table 3.10 Normal parameters for ABC assessment

Age	Breathing rate (breaths per min)	Heart rate (beats per min)	Systolic blood pressure (mmHg)
Neonate	30–60	100–160	50–70
6 months	25–40	100–140	70–90
12 months	20–30	90–130	80–100
2 years	20–30	90–120	80–100
5 years	18–25	80–120	80–110
9 years	15–20	70–100	90–110
12 years	15–20	60–100	90–120
15 years	15–20	60–100	90–120

Table 3.11 Neonatal assessment

	Apgar score		
	0	1	2
Breathing	Absent	Slow, irregular	Good, crying
Heart rate	Absent	<100 beats · min^{-1}	>100 beats · min^{-1}
Color	Blue or pale	Blue extremities	Completely pink
Reflex irritability	No response	Grimace	Cough, sneeze, cry
Muscle tone	Limp	Some flexion	Active motion

Neonatal resuscitation treatment

- **Call for help and keep the infant warm!**
- A typical resuscitation plan is outlined in Table 3.12 and the guidelines are shown in Table 3.13

Table 3.12 Resuscitation plan

Time (min)	Breathing	Heart rate	Treatment	
0 –1	Inadequate	> 100	Stimulate by suction (mouth, nose, throat)	
	Inadequate	< 100	Suction, face mask ventilation	
2	Inadequate	> 100	Face mask ventilation	
	None	< 100	Intubation and ventilation	
	None	< 60	Intubation Ventilation	Neonates 3:1
				Children < 12 15:2
			Chest compression	Adolescents 30:2
5	None	< 60	Ventilation, chest compression Medication ▪ Adrenaline 10 µg · kg^{-1} i.v. or i.o. ▪ Sodium bicarbonate 1–2 mmol i.v. or i.o.	
8–10	None	< 60	Ventilation, chest compression Medication ▪ Adrenaline 100 µg · kg^{-1} i.v.	
			Repeat every 5 min	
20–30	None	None	Stop resuscitation, except if ▪ Poisoning with cerebral depressant drugs ▪ Core temperature below 32°C	

Table 3.13 Resuscitation guidelines. Sodium bicarbonate inactivates epinephrine and dopamine! The lines should be flushed between medications

Age	Weight (kg)	Mask size	Endotracheal tube inner diameter (mm)	Adrenaline (mg)	Atropine (mg)	Bicarbonate 8.4% (ml) 1 ml = 1 mmol	Defibrillation (J)
Premature neonate	< 2	0	2.5	0.02–0.2	–	2.0	5/10/10
Neonate	3	0	3.0–3.5	0.03–0.3	–	3.0	5/10/10
6 months	7	0–1	3.5–4.0	0.07–0.7	0.14	7.0	15/30/30
1 year	10	1–2	4.0	0.1–1.0	0.2	10	20/40/40
2 years	12	2–3	4.5	0.12–1.2	0.24	12	25/50/50
5 years	19	4.0	5.0–5.5	0.19–1.9	0.38	19	40/75/75
10 years	30	4–5	6.0	0.3–3.0	0.6	30	60/120/120
12 years	40	5	6.5	0.4–4.0	0.8	40	80/150/150
15 years	50	5–6	7.0–7.5	0.5–5.0	1.0	50	110/200/200
				0.01–0.1 mg · kg^{-1}	0.02[a] mg · kg^{-1}	1 ml · kg^{-1}	ca. 2/4/4 J/kg

[a]Minimum dose 0.1 mg.

Airway management

Airway obstruction can be caused by the tongue, blocking the pharynx. Chin lift and jaw thrust are the first maneuvers performed in order to establish an open airway.

- Chin lift (Fig. 3.1)
  - Tilt the head back with one hand
  - The chin is lifted forwards with the fingers of the other hand
- Jaw thrust (Fig. 3.2)
  - Keep the head in a neutral position with two hands
  - The fingers of both hands push the jaw forwards.

Fig. 3.1 Chin lift in infants

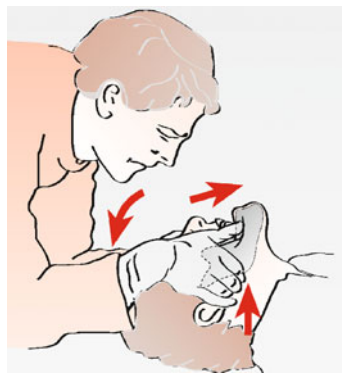


Fig. 3.2 Jaw thrust

Orotracheal intubation

- **General information**
- Emergency intubation is usually performed orotracheally (Fig. 3.3)
- In infants nasotracheal intubation may be preferred
- Uncuffed tubes are used for children less than 8 years old

- **Estimated internal diameter (mm)**
- (Age in years ÷ 4) + 4
- 3–3.5 mm (for neonates)
- 2.5 mm for prematures
- Same diameter as little finger
- Size just fitting into the nostril

- **Estimated tube length (cm)**
- (Age in years ÷ 2) + 12 (for oral intubation)
- (Age in years ÷ 2) + 15 (for nasal intubation)

- **Indication for intubation**
- Airway protection and maintenance
- Facial injury
- Airway obstruction
- Absent reflexes (coma)
- Need for mechanical ventilation

- **Intubation technique**
- Immobilize neck in trauma patients
- Clear pharynx and stomach by suction
- Ventilate by face mask
- Introduce laryngoscope with tip in the vallecula
- Insert endotracheal tube with tip 2–4 cm below vocal cords
- Check chest movement, auscultate chest and epigastrium
- Fasten tube
- Check tube position with X-ray
- If attempt is not successful within 30 s, discontinue and ventilate with face mask

- **Pit falls**
- Too much hyperextension of neck (in smaller children)
- Do not lean on the teeth
- Lift blade of laryngoscope towards ceiling, do not lever

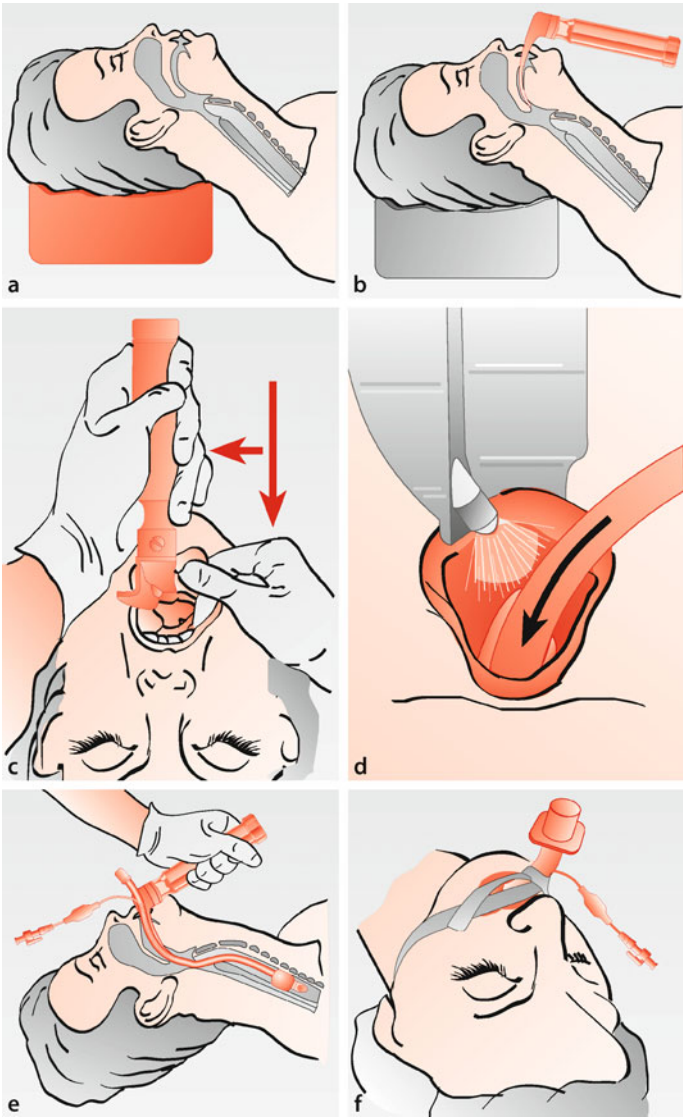


Fig. 3.3a–f Orotracheal intubation

- **Complications of endotracheal intubation**
- Acute
  - Traumatic
    - Cervical spine injury in trauma patient
    - Dental trauma
    - Vocal cord damage
    - Bleeding
    - Laryngeal edema
  - Mechanical
    - Tube malposition (bronchus, esophagus)
    - Tube obstruction
    - Accidental extubation
    - Tube kinking
  - Other
    - Pneumothorax (e.g., if endobronchial intubation)
    - Apnea
    - Hypoxia from prolonged attempt to intubate
    - Aspiration
- Chronic
  - Otitis media
  - Choanal stenosis
  - Subglottic stenosis
  - Granuloma
  - Necrotizing tracheobronchitis
  - Tracheal stenosis
  - Tracheomalacia

- **Complications of endotracheal suction**
- Bleeding
- Accidental extubation
- Pulmonary or bronchial perforation
- Bronchopulmonary fistula
- Hypoxia
- Bradycardia

- Increased cerebral perfusion and intracranial pressure
- Increased blood pressure

- **Checklist for intubation**
- Materials and instruments
  - Face mask with ventilation bag
  - Gloves
  - Stethoscope
  - Respiratory mask
    - Right mask size
    - Oxygen supply on
  - Suction devices
    - Suction tube on device
  - Prepared ventilation system
  - Oxygen supply
  - ECG monitor
  - Intubation set
  - Laryngoscopes
  - Endotracheal tubes and connectors
  - Silicon spray
  - Tube introducers
  - Magill's forceps
- Medication
  - Adrenaline, 1 ml = 1 mg dilute 1:9 NaCl
  - Atropine, 1 ml = 0.5 mg dilute 1:4 NaCl
  - Xylocaine 2%, 5 ml = 100 mg
  - Bicarbonate 8.4%, 1 ml = 1 mmol
  - Glucose 5%, 10 ml
  - Glucose 10%, 10 ml

Cricothyroidotomy

- Access to the airway through the cricothyroid membrane (Fig. 3.4)
- In infants the trachea is punctured below the cricoid cartilage instead of puncturing through the cricothyroid membrane
- In infants and small children needle cricothyroidotomy is advocated

- In older children and adolescents the airway can be established percutaneously or with an open surgical procedure
- Needle cricothyroidotomy is a temporary relatively small airway access, requiring more definite (surgical) airway access as soon as possible
- If prolonged airway access is required in a child, who cannot be tracheally intubated, tracheostomy is preferred over cricothyroidotomy

Indication

- Urgent need for an open airway if tracheal intubation is impossible

Needle cricothyroidotomy technique

- Hyperextension of neck (immobilize neck in trauma patients)
- Stabilize the cricoid and thyroid cartilage with one hand
- Puncture the trachea between the cricoid and thyroid cartilage with a cannulated needle on a syringe, penetrating the trachea at a slight caudal angle
- Withdraw needle when air is aspirated
- Introduce cannula further, recheck intraluminal position
- Attach Y-connector with oxygen flow at one leg
- Ventilate by opening (4 s) and closing (1 s) the other leg of the connector
- The required oxygen flow in liters is the child's age in years
- Establish a more definitive airway

Percutaneous cricothyroidotomy

- Hyperextension of neck (immobilize neck in trauma patients)
- Stabilize the cricoid and thyroid cartilage with one hand
- Puncture the trachea between the cricoid and thyroid cartilage with a cannulated needle on a syringe, penetrating the trachea at a slight caudal angle
- Withdraw needle when air is aspirated
- Introduce cannula further, recheck intraluminal position
- Introduce guide wire through the cannula
- Insert appropriately sized cannula on a dilator over the guide wire
- Remove dilator and guide wire
- Fasten cannula to the neck

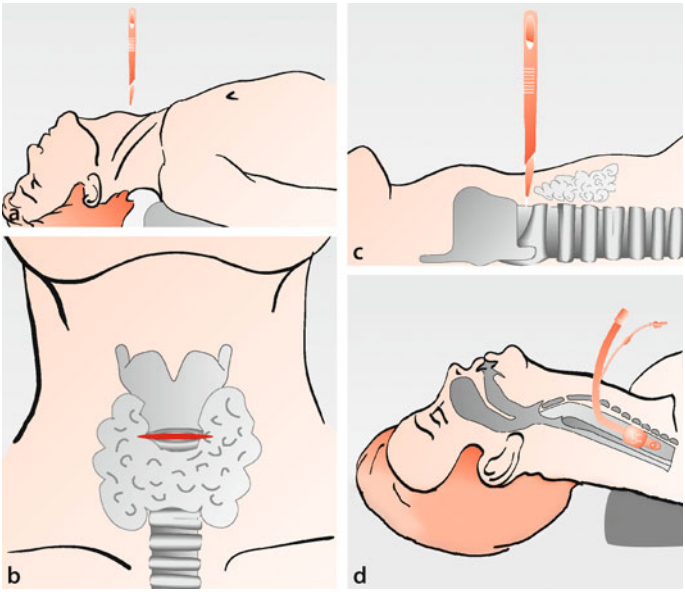


Fig. 3.4a–d Cricothyroiditomy

Surgical cricothyroidotomy

- Hyperextension of neck (immobilize neck in trauma patients)
- Infiltrate with local anesthetic
- Small midline incision over cricothyroid membrane
- Transverse incision through cricothyroid membrane
- Insert spreader into wound
- Insert appropriately sized cannula
- Fasten cannula

Complications of cricothyroidotomy

- Hemorrhage
- Tracheo-esophageal fistula
- Dislodgement of cannula
- Wound infection

- Pneumothorax, subcutaneous emphysema
- Subglottic stenosis
- Voice changes

Checklist for cricothyroidotomy

- Surgical blade
- Cannulated needle
- Guide wire
- Different size cannulas and dilators
- Tracheal spreader
- Y-connector
- Oxygen with flow meter

3.6 Ventilation

3.6.1 Ventilation Without Equipment

Mouth-to-mouth ventilation technique

- Chin lift or jaw thrust
- Mouth-to-mouth and -nose ventilation in smaller children, mouth-to-mouth ventilation (nose closed) in older children
- Administer slow breaths of 1 s duration at low pressure
- Check chest movements
- Rate: 30–60 per minute in neonates; 13 per minute in children <12; 7 per minute in adolescents
- Beware of too large a volume of inflation

Ventilation by mask (Fig. 3.5)

- Choose appropriately sized mask
  - Infant mask (<7 kg)
  - Children mask (7–30 kg)
  - Adult mask (>30 kg)
- In infants the mask may be used upside down

- If available, insert oro- or nasopharyngeal airway (Guedel tube) keeping airways patent
  - Length of the oropharyngeal airway: from center of the incisors to mandible angle
  - Length of the nasopharyngeal airway: from the tip of the nose to tragus of the ear)
- Slight hyperextension of neck
- Apply mask to the face with one hand (with the little, ring and middle finger the jaw is held forward to prevent air leak; with the thumb and index finger, the mask is pushed on the face)
- The bag is attached to the mask and squeezed with the other hand

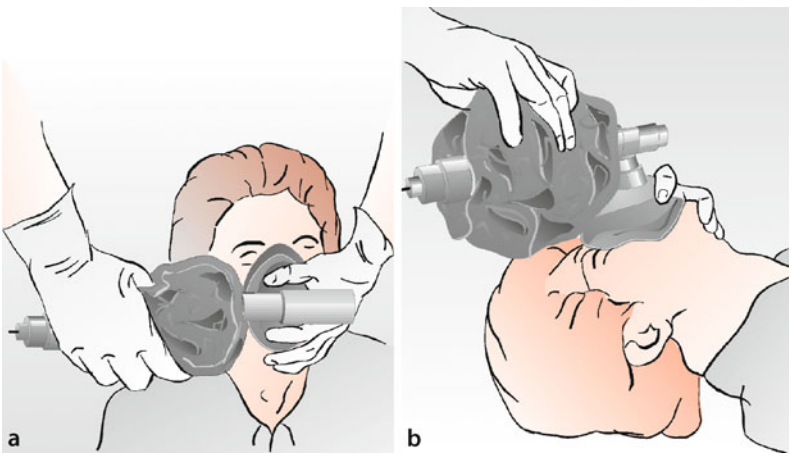


Fig. 3.5a,b Mask ventilation with ventilation bag

Advantages

- Can be applied in most circumstances
- No mouth-to-mouth contact
- Pressure-limiting valves in smaller self-inflating bags may prevent barotrauma
- Special valves for positive end-expiratory pressure (PEEP) ventilation

Disadvantages

- More difficult than generally considered (beware of insufficient ventilation)
- Danger of inadequate ventilatory volume

Checklist for mask ventilation

- Face masks
- Self-inflating bags
- T-piece and open-ended bag
- Oxygen supply

3.6.2 Mechanical Ventilation

Definitions

- Controlled ventilation: complete ventilatory effort is done by the ventilator
- Assisted ventilation: breaths are initiated by the patient; the ventilator delivers a preset volume or pressure
- Commonly encountered abbreviations are listed in Table 3.14

Table 3.14 Commonly encountered abbreviations

CMV	Controlled mechanical ventilation
CPAP	Continuous positive airway pressure
CPPV	Continuous positive pressure ventilation
HFPPV	High-frequency positive pressure ventilation
HFJV	High-frequency jet ventilation
HFOV	High-frequency oscillatory ventilation
IMV	Intermittent mandatory ventilation
IPPV	Intermittent positive pressure ventilation
PEEP	Positive end-expiratory pressure
PIP	Positive inspiratory pressure
SIMV	Synchronized intermittent mandatory ventilation
SIPPV	Synchronized intermittent positive pressure ventilation

Ventilation forms

- IPPV: during inspiration positive inspiratory pressure is generated, expiration is caused by elasticity of the lung and thorax (usually it is combined with PEEP)
- IMV: between mandatory breaths from the machine, the child can breathe spontaneously. This can be synchronized with the patient's own breaths
- PEEP: at the end of expiration the ventilator pressure remains positive
- CPAP: positive respiratory pressure in a spontaneously breathing patient

Ventilatory modes to be controlled

- Fraction of inspiratory oxygen (FiO_2)
- PEEP/CPAP level (cmH_2O)
- PIP (cmH_2O)
- Ventilatory frequency
- Inspiratory time to expiratory time ratio (*I*:*E* ratio)
- Respiratory minute volume ($l \cdot min^{-1}$), tidal volume (ml)
- Mean respiratory pressure
- Breaths may be volume controlled or pressure controlled (Table 3.15)

Table 3.15 Advantages and disadvantages of the various types of ventilation

	Advantages	Disadvantages
Volume-controlled ventilation	▪ Fixed tidal volume	▪ Risk of barotrauma
Pressure-controlled ventilation	▪ Small risk of barotrauma	▪ Variable tidal volume
PEEP/CPAP	▪ Prevents alveolar collapse ▪ Increases functional residual capacity ▪ Decreases intrapulmonary right–left shunt	▪ Decreases cardiac output ▪ Increases intracranial pressure ▪ Barotrauma ▪ Decreases pulmonary compliance
Increased ventilatory frequency	▪ Lower PIP necessary ▪ Lower mean respiratory pressure if hyperventilation required	▪ Air trapping
Decreased ventilatory frequency	▪ Improves oxygen exchange	▪ Relatively high PIP necessary
High PIP	▪ Improved oxygen exchange ▪ Prevents atelectasis	▪ May hinder venous return ▪ Decreases cardiac output ▪ Side-effects, such as pulmonary air leak

Indications for mechanical ventilation

- Failure of breathing (pulmonary, non-pulmonary such as apnea)
- Ventilatory failure (hypercapnia)
- Hypoxemia
- Chest wall instability
- To decrease respiratory effort (e.g., sepsis, cardiac failure)
- To control respiratory function (e.g., in high intracranial pressure)

Clinical criteria for mechanical ventilation

- Tachypnea
- Apnea
- Grunting
- Stridor

- Flaring of alae nasi
- Recessions
- Tachycardia
- Bradycardia
- Cyanosis
- Agitation
- Lethargy

Laboratory criteria for mechanical ventilation

- $PaO_2 < 50$ mmHg with $FiO_2 > 0.5$
- $PaCO_2 > 55$ mmHg with acidosis
- $PaCO_2 > 40$ mmHg with severe breathlessness
- Vital capacity $< 15\ ml \cdot kg^{-1}$

Guideline pediatric respiratory setting

- FiO_2 up to 1.0 to achieve required PaO_2, preferably below 0.6
- PEEP generally between 3 and 5 cmH_2O. Higher in reduced functional capacity with hypoxia if hemodynamically tolerated
- Aim for the physiological frequency corresponding to the age. Increase ventilatory frequency if $PaCO_2$ is high
- *I*:*E* ratio approximately 1:2; increase expiration time in obstructive pulmonary disease
- Tidal volume between 12 and 15 $ml \cdot kg^{-1}$
- Neonates, infants, and small children are generally ventilated with pressure control, in older children volume control may be preferred
- Estimated normal values of breathing are given in Table 3.16

Table 3.16 Estimated normal values of breathing

Age	Breathing rate (breaths per min)	Tidal volume (ml)
Neonate	30–60	20–35
12 months	20–30	40–100
5 years	18–25	150–200
9 years	15–20	300–400
15 years	15–20	300–500

Control of mechanical ventilation

- Auscultate bilateral ventilation
- Check hemodynamic side-effects such as heart rate, blood pressure, and peripheral perfusion
- Check transcutaneous oxygen saturation and the expiratory CO_2 concentration
- Control blood gases

Complications of mechanical ventilation

- Acute and chronic interstitial emphysema
- Pneumothorax, pneumomediastinum, pneumopericardium
- Pneumoperitoneum
- Subcutaneous emphysema
- Respiratory air trapping
- Air embolus

3.7 Circulatory Resuscitation

Indications for cardiac compression

- Pulseless patient
- Pulse less than 60 beats per minute in infants

Technique of cardiac compression

- Child flat on its back
  - Neonates (Fig. 3.6)
    - Hands encircle chest, compression with thumbs
    - Compress sternum 1.5 cm below nipples
    - Frequency 120 per minute
    - Press down to a depth of about 1.5 cm
    - Compression:breath ratio 3:1

- Infants (Fig. 3.7)
  - Use two fingers
  - Compress the lower third of the sternum
  - Frequency 100 per minute
  - Press down to a depth of about one-third of the depth of the chest
  - Compression:breath ratio 15:2
- Small children (Fig. 3.8)
  - Use one or two hands
  - Compress the lower third of the sternum
  - Frequency 100 per minute
  - Press down to a depth of about one-third of the depth of the chest
  - Compression:breath ratio 15:2

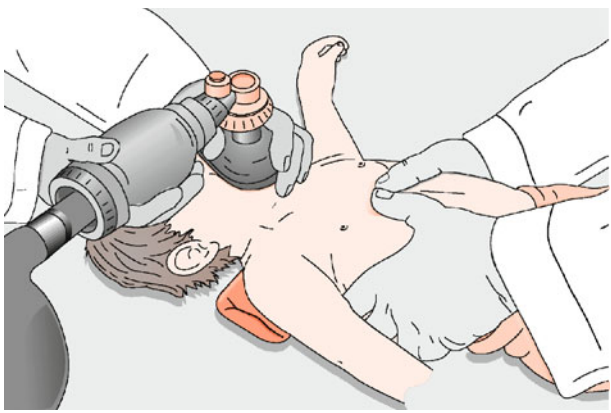


Fig. 3.6 Cardiac compression in neonates

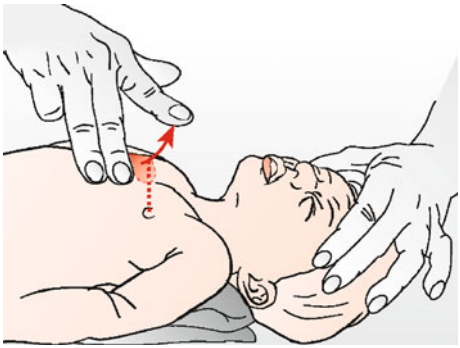


Fig. 3.7 Cardiac compression in infants

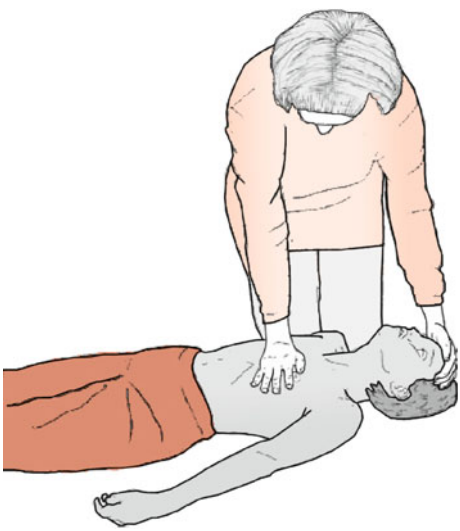


Fig. 3.8 Cardiac compression in small children

Complications of cardiac compression

- Rib or sternum fractures
- Pulmonary contusion
- Hematothorax, pneumothorax
- Liver, spleen rupture

3.8 Defibrillation

Indication

- ECG has confirmed ventricular fibrillation
- Pulseless ventricular tachycardia

Technique of defibrillation (Fig. 3.9)

- Cardiopulmonary resuscitation (CPR)
- Consider intubation
- Apply self-adhesive defibrillation pads below right clavicle and lateral to left nipple
- Apply paddles with firm pressure (infants <10 kg: 4.5-cm paddle; children, 8-cm paddle)
- Shock three times initially with 2, 4, and 4 $J \cdot kg^{-1}$ respectively with pauses to assess the rhythm
- Continue CPR for 2 min, adrenaline 10 $\mu g \cdot kg^{-1}$ i.v. or i.o.
- Assess rhythm
- If required shock three times with 4 $J \cdot kg^{-1}$
- Continue CPR for 3 min, adrenaline 100 $\mu g \cdot kg^{-1}$ i.v. or i.o., correct acidosis and so on
- Correct reversible causes
  - Hypoxia
  - Hypothermia
  - Hypovolemia
  - Electrolyte imbalance
  - Tension pneumothorax
  - Tamponade
  - Drugs

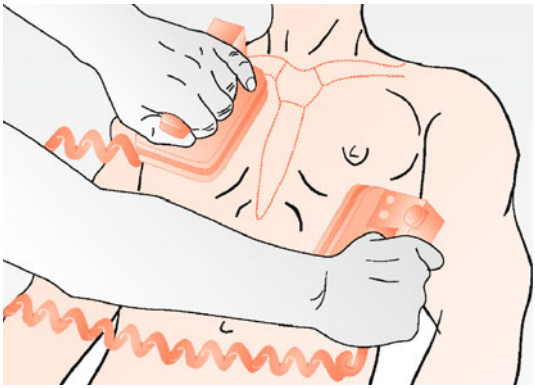


Fig. 3.9 Electrode position for defibrillation

Checklist

- Defibrillation pads
- Defibrillator
- ECG monitor
- Venous or intraosseous access
- Medication, e.g., adrenaline, amiodarone

4 Blood/Blood Products Transfusion

4.1 Blood Transfusion

Prerequisites

- Blood is usually cross-matched (which may take up to 1h)
- In emergency cases type-specific (ABO) or O-Rh-negative blood may be given
- In infants aged up to 3 months blood is also cross-matched with the mother's blood
- Whole blood is only given in children to replace an acute blood loss or during exchange transfusions
- Blood for children should be stored and ordered in small portions, e.g., 50 ml, if possible from the same donor

Physiologic values

Physiologic values are given in Table 4.1.

Table 4.1 Physiologic values

	Weight (kg)	Estimated circulating blood volume ($ml \cdot kg^{-1}$)	Hemoglobin ($g \cdot dl^{-1}$)	Hematocrit (%)
Preterm	1–3	90–100		
1 day	3–4	80–90	14–22	44–64
7 days	3–4	80–90	13–21	40–59
1 month	3–5	80–90	11–17	35–50
3 months to 1 year	5–10	80	10–15	31–41
5 years	20	75	12–15	33–42
10 years	32	70	12–16	34–40

Indication

- The indication for blood transfusion varies with age, underlying disease, and the operation
- Unnecessary transfusions should be avoided
- In children with acute hemorrhage transfusion is given to treat shock after two boluses of crystalloid or colloid
- In chronic anemia transfusion is indicated if the anemia is profound ($6–9\ g \cdot dl^{-1}$) or if complications such as heart failure are present
- Recommendations according to age are given in Table 4.2

Table 4.2 Recommendations for blood transfusion according to age

	Normal Hb($g \cdot dl^{-1}$)	Transfusion threshold ($g \cdot dl^{-1}$)
Premature newborns	16–20	< 12
Newborns	16–18	< 11
Children	10–12	< 6

Calculation of transfusion dose

- To increase the hemoglobin concentration by $1\ g \cdot dl^{-1}$ infuse $3\ ml \cdot kg^{-1}$ erythrocyte concentrate or $6\ ml \cdot kg^{-1}$ whole blood
- Use the formula
  $[Hb_{target} – Hb_{actual}] \times kg \times 3 = ml$ transfusion

Laboratory changes

Laboratory test changes in acute and in chronic blood loss, in blood group incompatibility and in hemolytic anemia are given in Table 4.3.

Table 4.3 Blood test results indicative of blood loss (acute and chronic), blood group incompatibility and of hemolytic anemia

	Hemoglobin	CVD	Coombs test	Jaundice	Reticulocytes
Acute blood loss	=, ↓	↓	–	–	=
Chronic blood loss	↓	=	–	–	↑
Blood group incompatibility	=, ↓	=, ↓	+	+	↑
Hemolytic anemia	↓	=	–	+	↑

Complications

Complications of blood transfusion are shown in Table 4.4.

Table 4.4 Complications of blood transfusion

Sign	Reason	Signs	Therapy	Prophylaxis
Hemolytic reaction	Transfusion of mismatched blood	▪ Hemoglobulinemia ▪ Hemoglobinuria ▪ Fever ▪ Shock	▪ Termination of transfusion ▪ Fluid therapy ▪ Mannitol	Control of blood compatibility
Febrile reaction	Antibodies against donor cells, release of pyrogens from recipient leukocytes	▪ Fever	▪ Antipyretics	Use washed erythrocytes
Circulatory overload	High volume	▪ Cardiac insufficiency ▪ Lung edema	▪ Diuretics	
Pulmonary emboli	Micro clots in old blood		▪ Corticosteroids	Use filter
Allergic reaction		▪ Urticaria ▪ Itching ▪ Wheezing ▪ Arthralgia	▪ Antihistamines ▪ Corticosteroids	
Graft-versus-host disease	Immunocompromised recipient		▪ Corticosteroids	Blood irradiation
Late hemolytic reaction		▪ Anemia ▪ Jaundice ▪ Fever		
Hemosiderosis	Iron overload		▪ Deferoxamine	
Infections		▪ Hepatitis ▪ HIV ▪ CMV		

General treatment of complications

In all transfusion reactions general signs should be treated first, starting with problems with the airway, breathing and circulation (ABC).

4.2 Platelet Transfusion

Indication

- Major hemorrhage with thrombocytopenia or platelet dysfunction
- Major operation and platelet count $< 50 \times 10^6 \cdot ml^{-1}$
- Minor operation and platelet count $< 20 \times 10^6 \cdot ml^{-1}$
- Sepsis and platelet count $< 20 \times 10^6 \cdot ml^{-1}$
- Abnormal bone marrow and platelet count $< 5 \times 10^6 \cdot ml^{-1}$
- Do not use platelet transfusion in conditions with platelet destruction

Dosage

- 12×10^9 thrombocytes in 10 ml plasma per kilogram, elevate the platelet count by $50 \times 10^6 \cdot ml^{-1}$

4.3 Fresh Frozen Plasma Transfusion

Definition

- FFP: fresh blood plasma separated from anticoagulated whole blood

Indication

- Depleted clotting factors, e.g., in liver failure or disseminated intravascular coagulation

Do not use FFP as volume substitution.

Dosage

- $10–15 \text{ ml} \cdot \text{kg}^{-1}$ twice a day

5 Pain Management

5.1 WHO Grades for Analgesia

WHO grades for analgesia are shown in Table 5.1, and their application to children is given in Table 5.2.

Table 5.1 WHO grades for analgesia

Grade 1	Non-opiate analgesics
Grade 2	Grade 1 + weak opiates
Grade 3	Grade 1 + strong opiates

Table 5.2 Application of WHO grades for analgesia to children

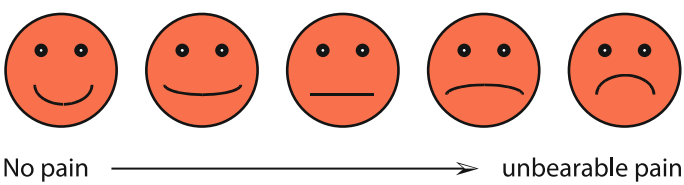
Grade	Drug	Single dose	Application	Duration of effect	Contraindication/ recommendations
1	Paracetamol	15–20 mg · kg^{-1}	p.o., supp.	4–6 h	Impaired liver, asthma
	Diclofenac	1 mg · kg^{-1}	p.o., supp.	8 h	
	Ibuprofen	5 mg · kg^{-1}	p.o.	4–6 h	
2	Tramadol	1–2 mg · kg^{-1}	p.o., supp., i.v.	6–8 h	Infants
	Codeine	1 mg · kg^{-1}	p.o.	4–6 h	Constipation

Table 5.2 *(continued)* Application of WHO grades for analgesia to children

Grade	Drug	Single dose	Application	Duration of effect	Contraindication/ recommendations
3	Morphine	0.025 mg·kg^{-1} (0–3 months) 0.05–0.1 mg·kg^{-1} (3–12 months) 0.1–0.2 mg·kg^{-1} (>1 year)	i.v.	6 h	Respiratory distress Respiration and ECG monitoring Slow injection
	Pethidine	10–20 µg·kg^{-1}·h^{-1} (continuous i.v.) 0.1–0.2 mg·kg^{-1} (<1 year) 0.2–0.4 mg·kg^{-1} (>1 year) 1 mg·kg^{-1} (max 6 mg·kg^{-1}·day^{-1})	p.o., s.c., i.m.	Continuous 6 h; 4–6 h	Respiration monitoring Asthma
	Naloxone = morphine antagonist	10 µg·kg^{-1}	i.v.		May be repeated after 2–3 min, acts for a shorter time than opiate

- The first drug listed per stage is usually the first choice
- In patients with acute pain long-lasting analgesics are preferred, which are provided on demand
- In patients with chronic pain long-lasting analgesics are preferred, which are taken prophylactically
- Analgesics are liberally used in every age group, including prematures and neonates

Smiley Scale



6 Wounds/Surgical Techniques

6.1 General Considerations

Defects of skin and/or mucosa

- Different types of skin defects are shown in Fig. 6.1 and are determined by the type of trauma experienced (Table 6.1)

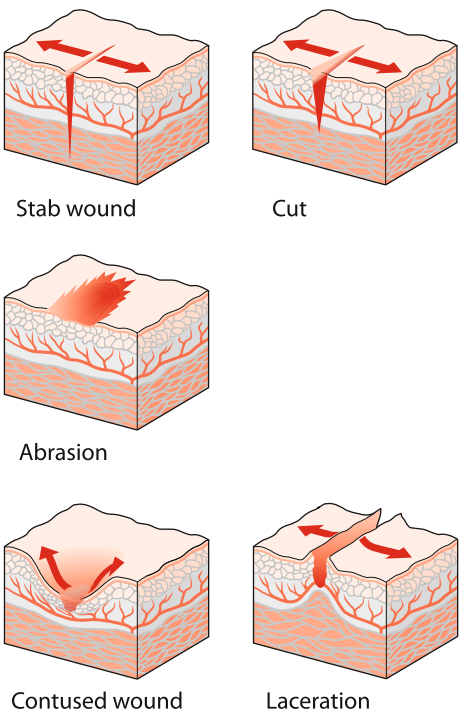


Fig. 6.1 Types of skin injury

Table 6.1 Form of skin defect that occurs following trauma

Trauma	Form
Sharp/peaking	Sharply cut, stabbed
Blunt	Contused, burst
Shearing	Abrasion
Combined	

Skin structure

- The structure of skin is shown in Fig. 6.2

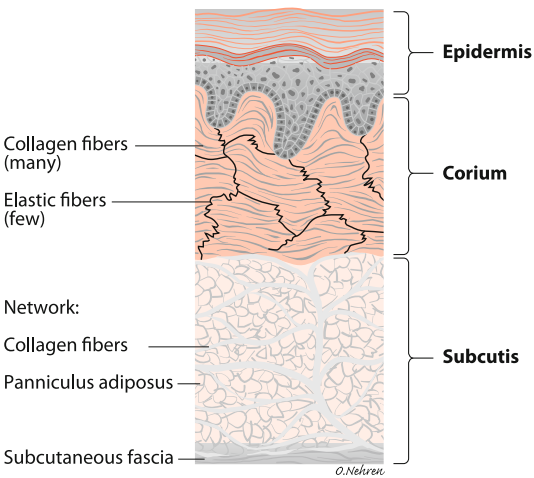


Fig. 6.2 Skin structure

6.2 Wound Healing

Phases of wound healing

- Exudation
- Resorption
- Proliferation
- Reparation

Primary wound healing

- Operative wound
- Primary closure after debridement
- Minimal scarring

Secondary wound healing

- Closure of the wound after granulation
- Scarring

Fundamentals of wound care

- Adequate wound cleaning and debridement (Fig. 6.3a,b)
- Secondary healing when necessary with air dressing
- The more caudal the wound, the more problematic the healing

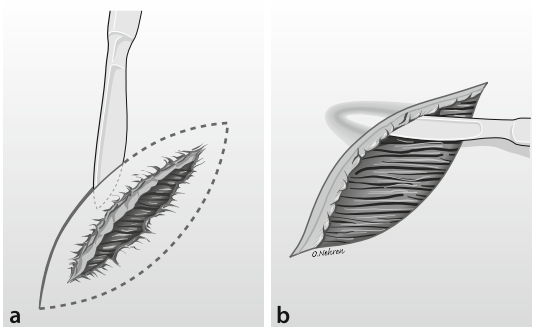


Fig. 6.3a,b Debriding the crushed edges of a wound. **a** Incision, **b** dissection

Signs indicating wound healing disorders

- Calor (warming)
- Dolor (pain)
- Rubor (redness)
- Tumor (swelling)
- Functio laesa (functionality loss)

Factors negatively influencing wound healing

- Preoperative, intraoperative, and postoperative factors that negatively influence wound healing are shown in Table 6.2

Table 6.2 Factors negatively influencing wound healing

Preoperative factors	Intraoperative factors	Postoperative factors
▪ Obesity ▪ Agranulocytosis ▪ Alcoholism ▪ Age (reduced fibroblasts and collagen) ▪ Anemia ▪ Diabetes ▪ Hypogammaglobulinemia ▪ Hypovolemia ▪ Infection, sepsis ▪ Long-standing disease ▪ Malignancy ▪ Malnutrition ▪ Drugs (corticosteroids) ▪ Renal failure ▪ Cirrhosis ▪ Coagulopathy ▪ Neuropathy ▪ Factor XIII deficit	▪ Desiccated tissue ▪ Hematoma ▪ Excessive cautery use ▪ Foreign bodies ▪ Foreign body reaction ▪ Wound contamination ▪ Hemorrhage ▪ Sutures under tension ▪ Seroma ▪ Tissue mishandling	▪ Long-standing catheters (venous, arterial, urinary) ▪ Contamination during wound dressing ▪ Keloids ▪ Malnutrition ▪ Massive transfusion

6.3 Common Types of Surgical Incisions

- The incision should follow, whenever possible, the tension lines of the skin (Fig. 6.4)
- Thoracic and abdominal incisions are illustrated in Figs. 6.5 and 6.6, respectively

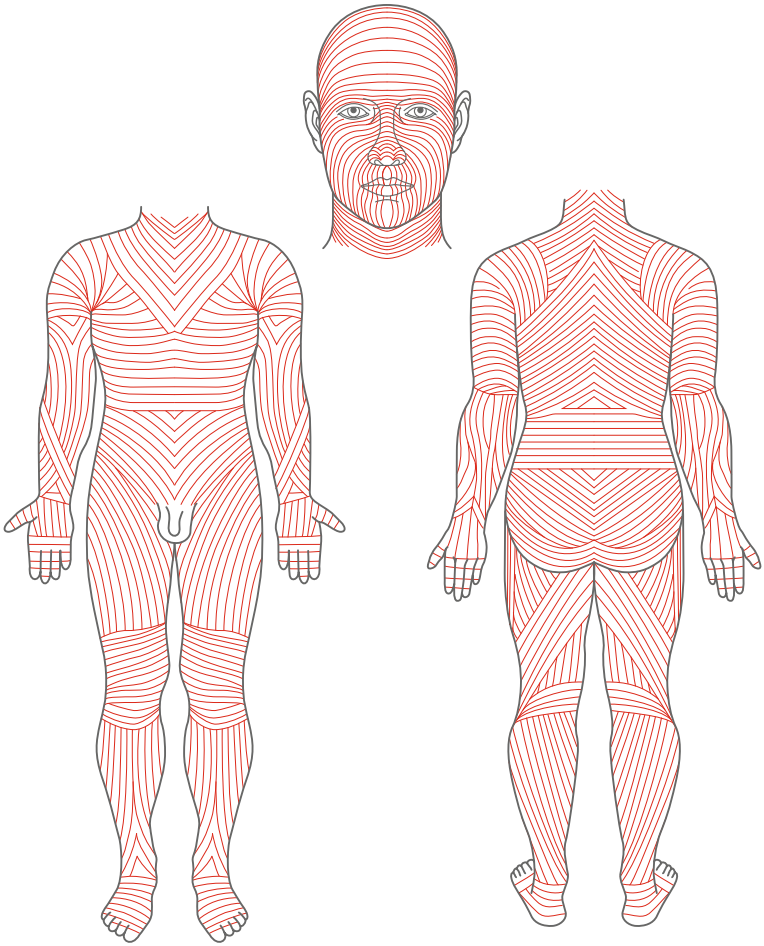


Fig. 6.4 Tension lines in the skin

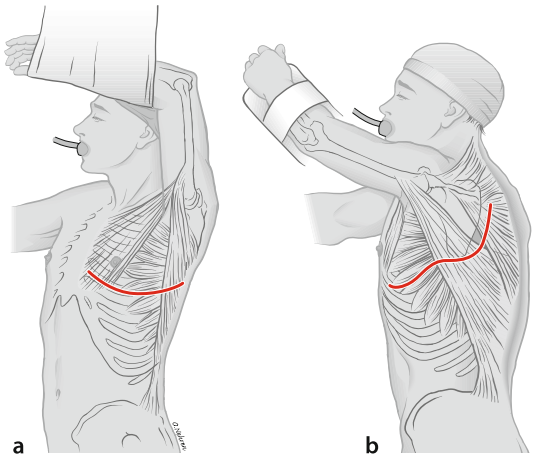


Fig. 6.5a,b Anterolateral **(a)** and posterolateral **(b)** thoracotomy

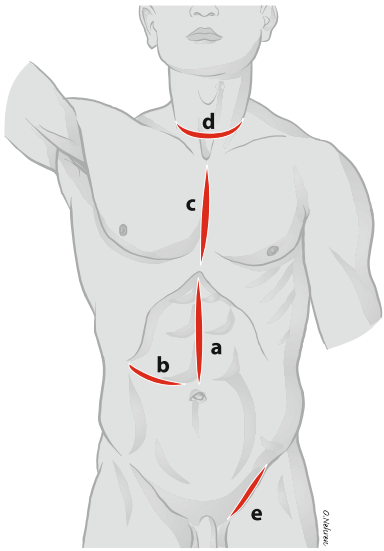


Fig. 6.6a–e Abdominal incisions. **a** Median laparotomy, **b** subcostal incision, **c** median sternotomy, **d** transverse collar incision, **e** inguinal incision

Thoracic incisions

- Median sternotomy (Fig. 6.6c)
- Axillary thoracotomy
- Anterolateral and posterolateral thoracotomy (Fig. 6.5)

Abdominal incisions

- Subcostal incision (Fig. 6.6b)
- Transverse laparotomy (supraumbilical)
- Median laparotomy (supraumbilical, infraumbilical) (Fig. 6.6a)
- Oblique inguinal laparotomy
- Pararectal incision
- McBurney incision
- Inguinal incision (Fig. 6.6e)
- Pfannenstiel incision

Other incisions

- Cervicotomy of Kocher (Fig. 6.6d)
- Suprasternal incision

6.4 Skin Flaps

Skin flaps are tissues that contain all the skin layers including vascular supply that can be mobilized and used to cover large skin defects or to increase the length of some scars or skin lines.

Skin flaps are tissues that the surgeon tries to keep alive in contrast to skin grafts that are dead tissues that the surgeon tries to bring alive.

The following plasties are recommended:

- Z-plasty (Fig. 6.7)
- Y-V plasty (Fig. 6.8)
- Transposition flap (Fig. 6.9)

Skin flap classification (after Berger–Kunert) is given in Table 6.3.

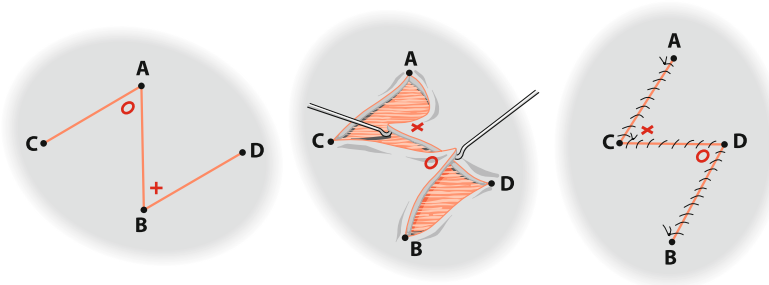


Fig. 6.7 Z-plasty

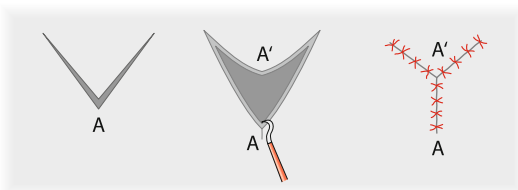


Fig. 6.8 Y-V plasty

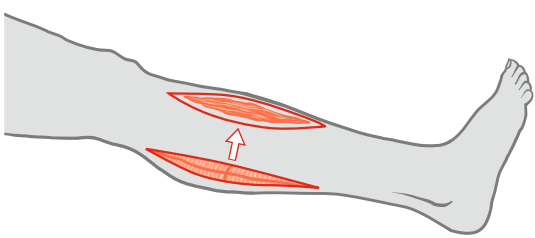


Fig. 6.9 Transposition flap

Table 6.3 Classification of skin flaps (after Berger–Kunert)

	Intact pedicle	Partially detached pedicle	Detached pedicle: (1) one-step or (2) two-step approach
Coincidentally vascularized	▪ Transposition flap ▪ V-Y plasty	▪ Subcutaneous island flap	▪ Split or full thickness grafts (1) ▪ Composite graft (1) ▪ Cross-leg graft (2)
Axial vascularization	▪ Temporal flap ▪ Dorsal foot flap ▪ Omental flap	▪ Dorsal foot island flap	▪ Dorsal foot free graft (1) ▪ Inguinal flap (2)
Fasciocutaneal vascularization	▪ Thigh fascia flap	▪ Thigh fascia island flaps	▪ Fasciocutaneous cross-leg graft (1, 2)
Myocutaneal vascularization	▪ Latissimus dorsi flap	▪ Latissimus dorsi island flap	▪ Free latissiums dorsi graft (1) ▪ Myocutaneous cross-leg graft (2)

6.5 Skin Grafts

Skin grafts (Fig. 6.10) are tissues that are transplanted from one region of the body to another without the vascular supply. They include all or only parts of the skin and can be full-thickness or split-thickness.

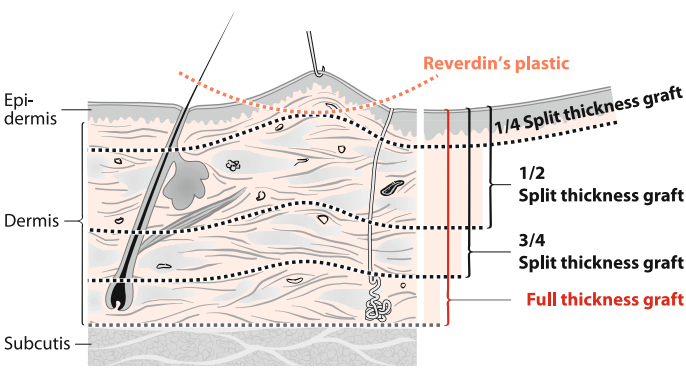


Fig. 6.10 Skin grafts

6.6 Surgical Sutures

Definitions

- Surgical sutures are described in Tables 6.4, 6.5, and Fig. 6.11

Indications

- Indications for suturing are given in Table 6.6

Table 6.4 Characteristics of surgical sutures

Name	Characteristics
Traumatic	Separate needle
Atraumatic	Assorted needle
Absorbable	Absorbed due to enzymatic or hydrolytic processes
Non-absorbable	Permanent in situ material
Monofilament (Fig. 6.11a)	Homogeneous structure, non-porous, minimal foreign body reaction
Polyfilament (Fig. 6.11b)	Knit suture, more pliable; porous, more reaction
Pseudomonofilament	Polyfilament with impermeable coating (Fig. 6.11c)

Table 6.5 Types of sutures

Non-absorbable	Structure	Features	Indications
Metallic	Iron, chrome, nickel	High resistance	Fascia, Bone
Polypropylene	Monofilament	Easy handling Little reaction	Viscera, skin, vessels
Polyamide	Monofilament	High resistance	Viscera, vessels, skin
Polyester	Polyfilament (knit)	Good resistance Good handling	Skin, vessels
Silk	Polyfilament (knit)	Good resistance Good handling Good knots Intense reaction	Vessels

Table 6.5 *(continued)* Types of sutures

Absorbable	Structure	Features	Indications
Polydioxanone (5 weeks)	Monofilament	Good handling Good resistance	Septic surgery, urology
Polyglyconate (4 weeks)	Monofilament	Good handling Good resistance	Septic surgery, urology
Polyglycolic acid, Polyglactin (2–3 weeks)	Polyfilament (knit)	Good handling Good resistance Medium reaction	Fascia, stomach, intestine, biliary tract, gynecology

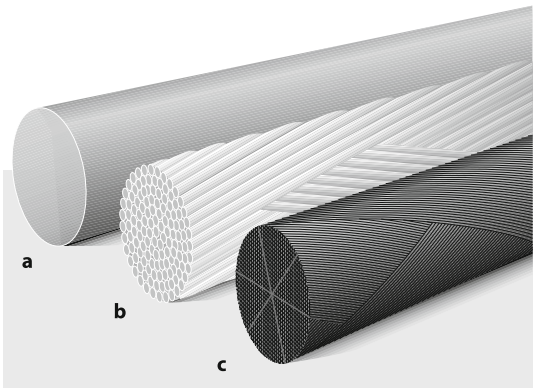


Fig. 6.11a–c Surgical suture materials. **a** Monofilament, **b** uncoated polyfilament, **c** coated polyfilament

Table 6.6 Indications for suturing

Situations	Effect
Spleen lesions/resection	Hemostasis and tissue preservation
Liver lesions/resections	Hemostasis and prophylaxis of bile leaks
Pancreas	Seals leaks
Intestinal anastomosis	Seals leaks
Plications	Makes sutures that may confine vascularization unnecessarily
Lung	Seals air and blood leaks
Fixation of skin grafts	Maintains vascularization
Hemangioma	Stops bleeding
Fixation of osteochondral fragments	Fixes small fragments

Principles of surgical suturing

- Careful approximation of wound edges without crushing the edges with the forceps
- Accurate alignment of the edges and their suture at points positioned appropriately to avoid the formation of a crease
- The edges should have good vascularization
- Avoid "caves" or "ears"
- Place knots on the side that has the better vascular supply
- Do not close primarily unsafe wounds

How to knot with instruments

- Take one end of the thread with the left hand and the needle holder with the right one
- With the left hand pass the thread above the needle holder
- Take the free end of the thread with the needle holder and pass it through the other rolled end
- Secure the knot
- To perform a square knot, both ends of the thread should be tractioned as shown in Fig. 6.12. This is crucial for the first knot
- The second knot should be performed in the same way
  - Take the other end of the thread
  - Secure the knot also as a square knot
- Both knots are now secure and unlikely to loosen

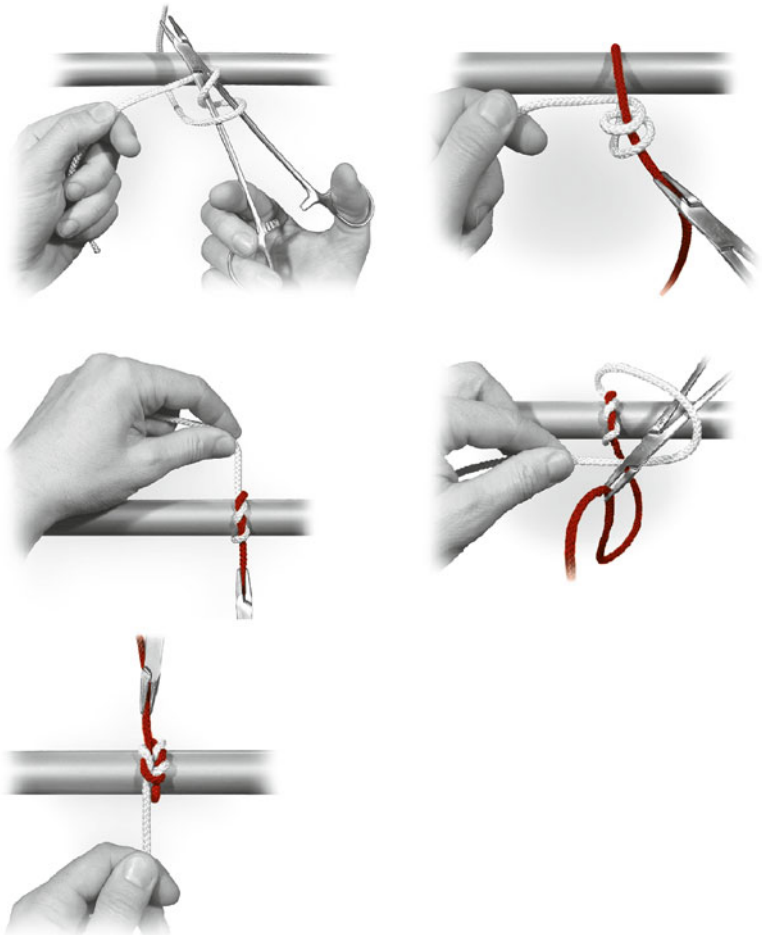


Fig. 6.12 Knotting with instruments

How to knot with the hands

- Knotting with the hands is demonstrated in Fig. 6.13

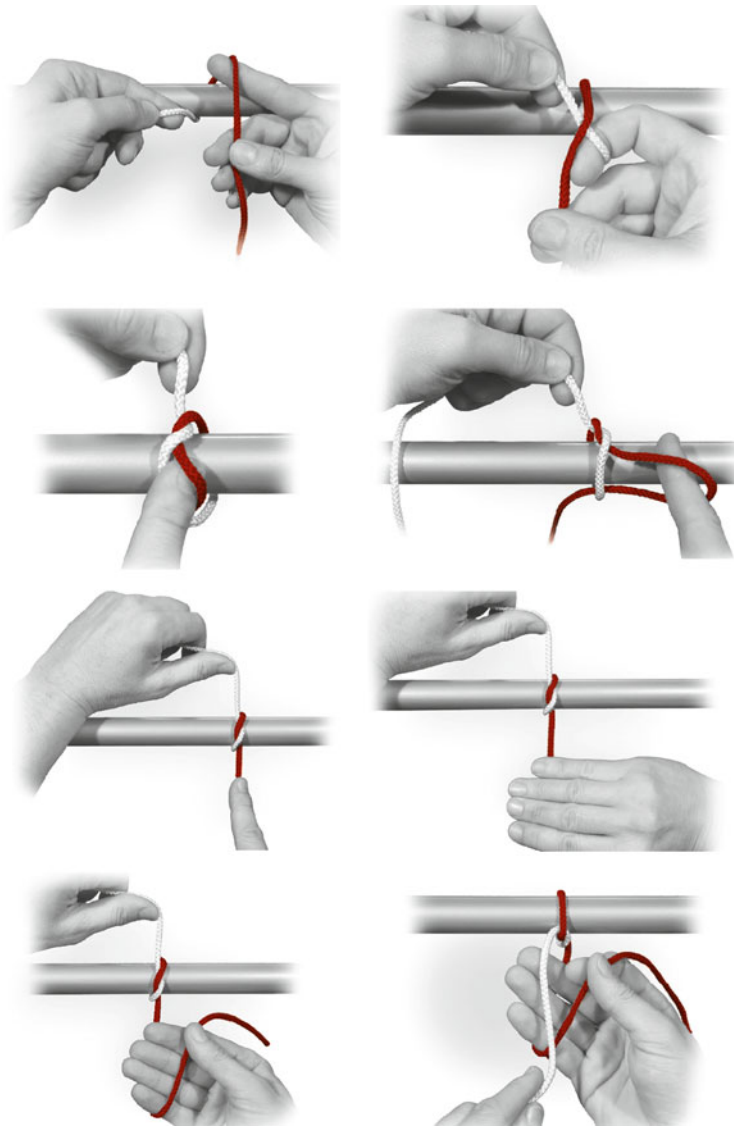


Fig. 6.13 Knotting with the hands

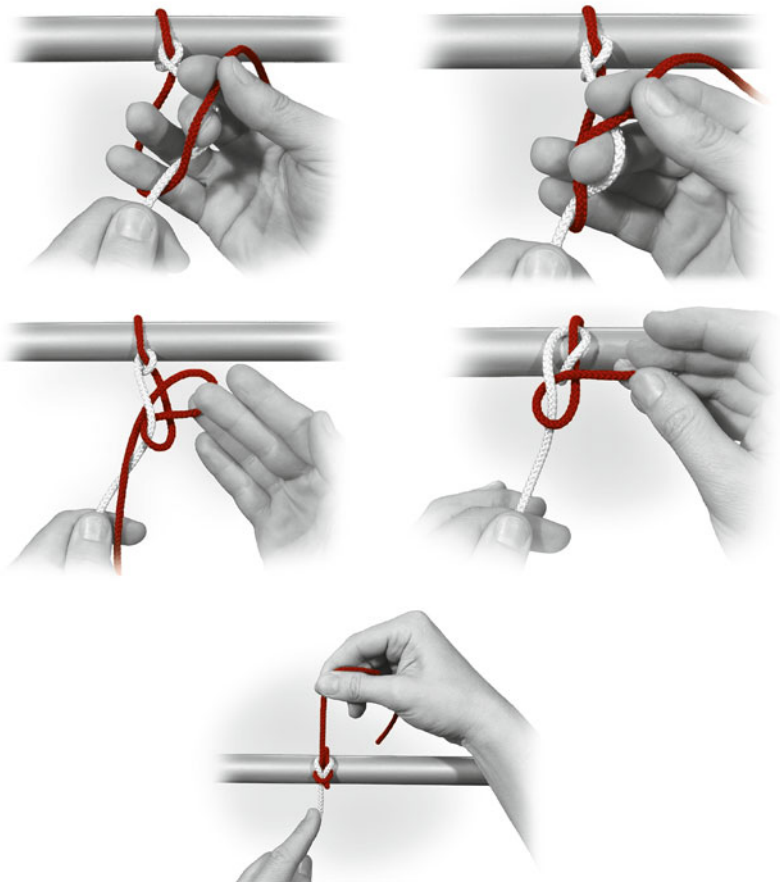


Fig. 6.13 *(continued)* Knotting with the hands

Sutures for skin closure

- Suturing the skin is illustrated in Fig. 6.14

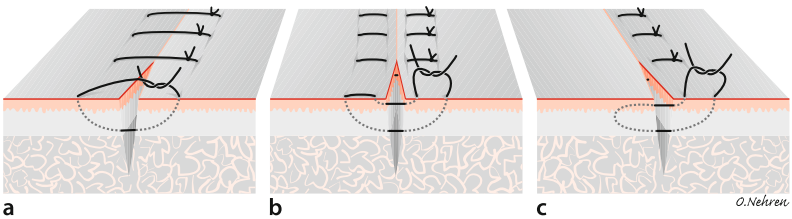


Fig. 6.14a–c Suturing the skin. **a** Simple knot, **b** Donati knot, **c** Allgöwer knot

Sutures for vessels

- Suturing techniques for blood vessels are shown in Fig. 6.15

Fig. 6.15 Suturing vessels

Sutures for gastrointestinal tract

- Suturing of the gastrointestinal tract can be in one or two layers, and is illustrated in Figs. 6.16 and 6.17

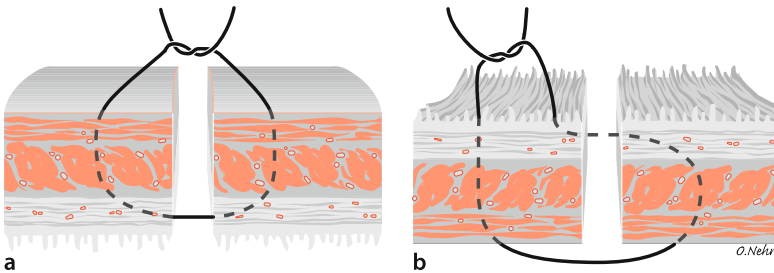


Fig. 6.16a,b Suturing the gastrointestinal tract. **a** Seromuscular suture, **b** backstitch mucosal suture

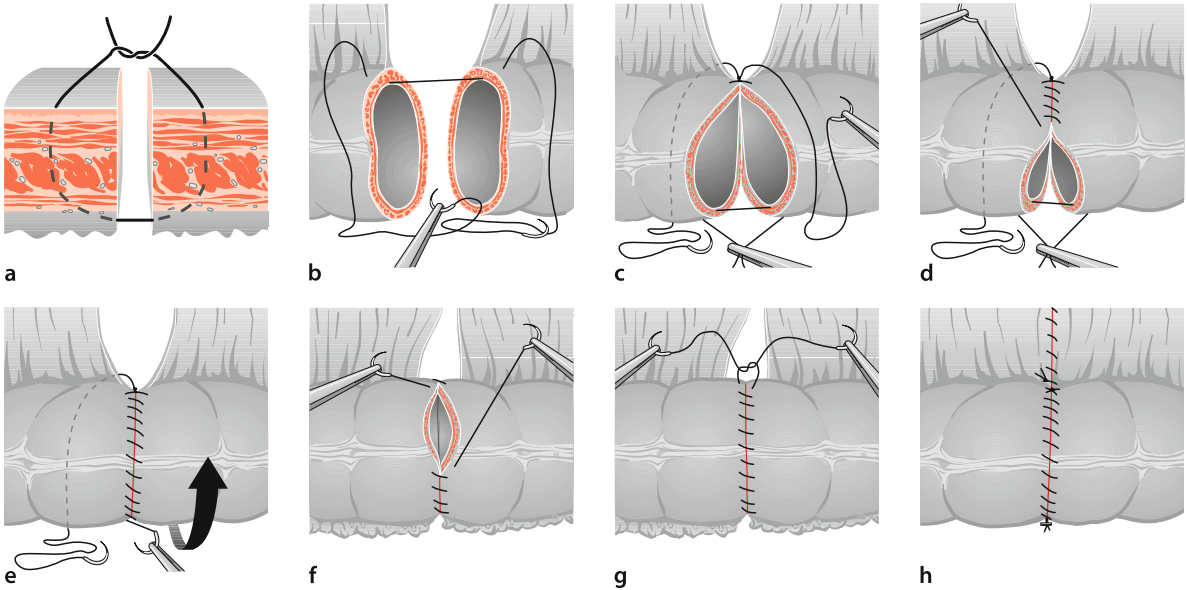


Fig. 6.17a–h Suturing the intestine

Sutures for nerves

- Suturing techniques for nerves are demonstrated in Fig. 6.18

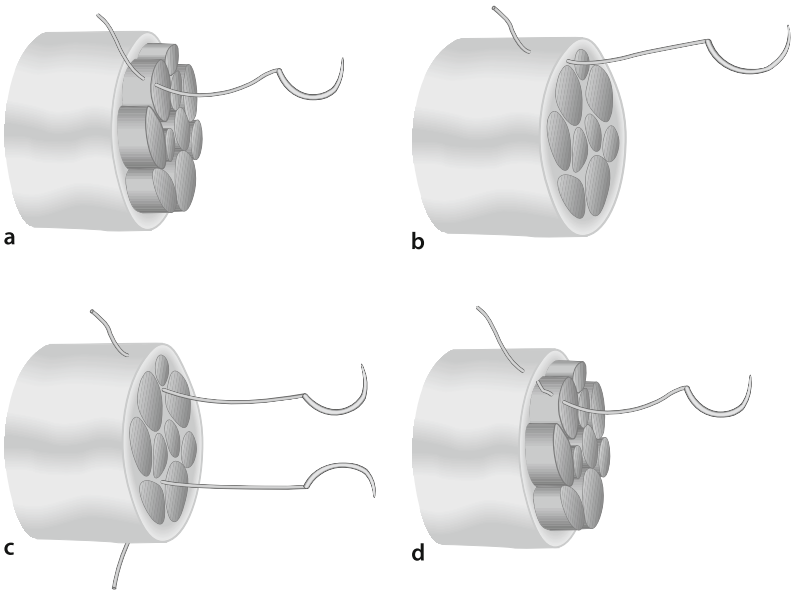


Fig. 6.18a–d Suturing nerves. **a** Perineural, **b** epineural, **c** interfascicular, **d** epi-perineural

Suture removal

- For children it is rarely necessary to maintain the sutures more than 1 week
- For some operations they can be removed on the 5th day
- The scars are usually relatively inconspicuous

Mechanical sutures

- The principles are the same as used for manual sutures, but using adequate mechanical devices:
  - Lateral suture section [with a mechanical transverse anastomosis (TA) device] (Fig. 6.19)
  - Functional end-to-end anastomosis [using a gastrointestinal anastomosis (GIA) device] (Fig. 6.20)
  - Anterior resection of the rectum with mechanic anastomosis (Fig. 6.21)

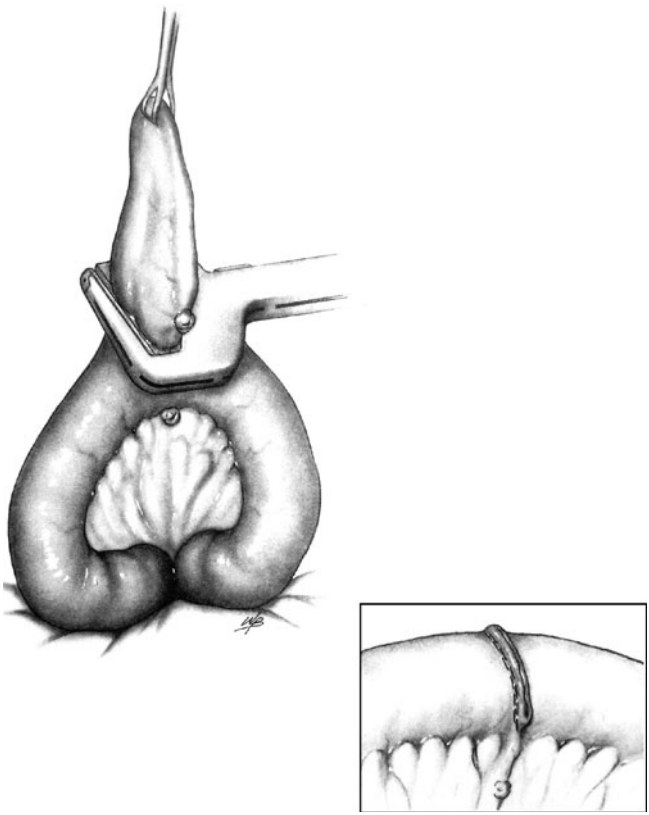


Fig. 6.19 Transverse mechanical anastomosis (TA device)

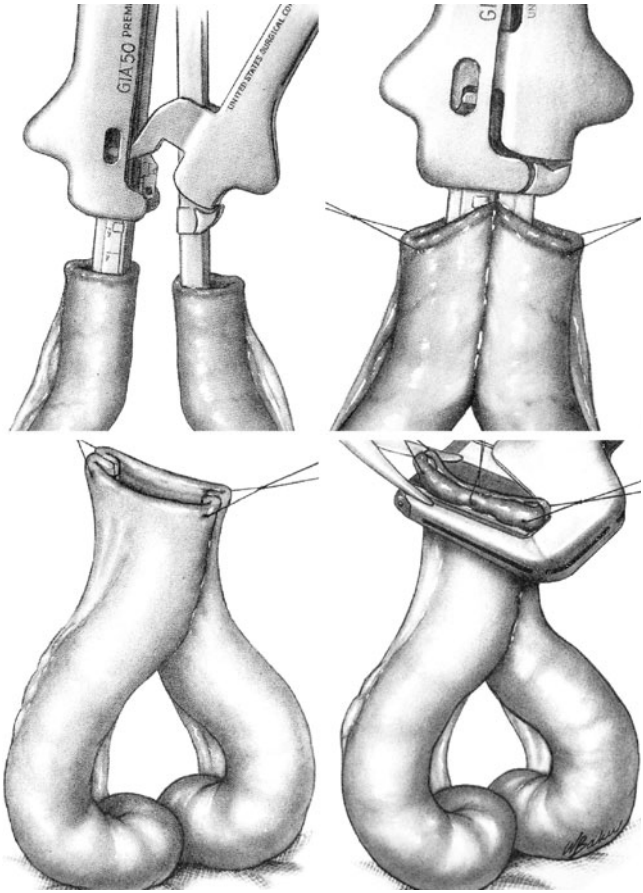


Fig. 6.20 Gastrointestinal anastomosis (GIA device)

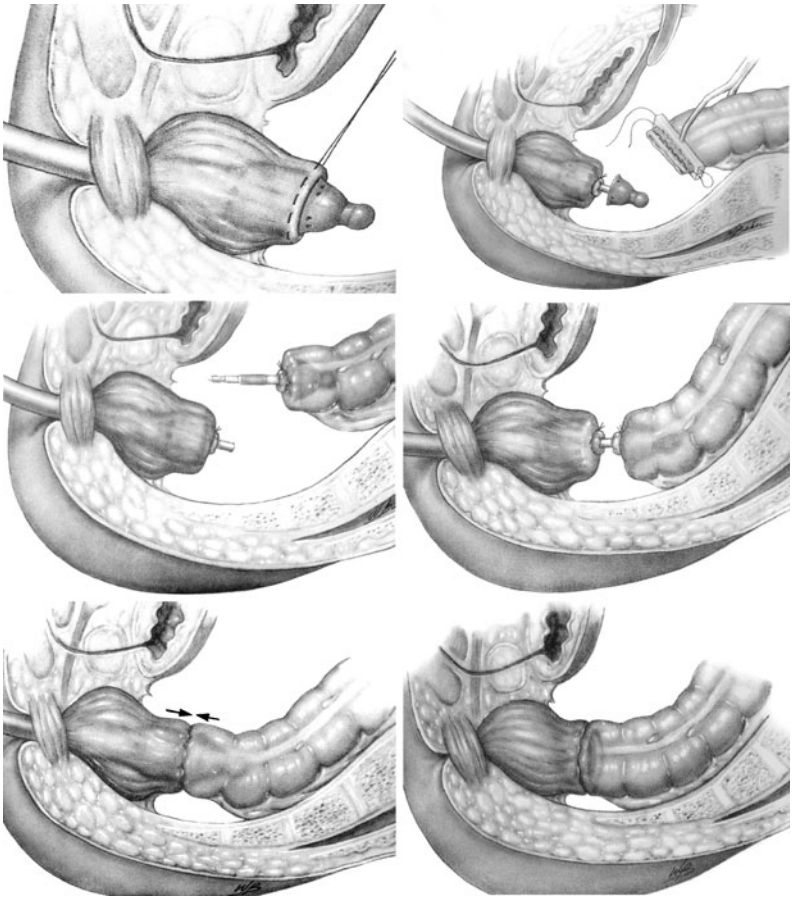


Fig. 6.21 Circular entero-enteral anastomosis (CEEA)

Surgical sealing

- The application of collagen and fibrin is now possible and this may help the surgeon to seal some surgical surfaces (Fig. 6.22)

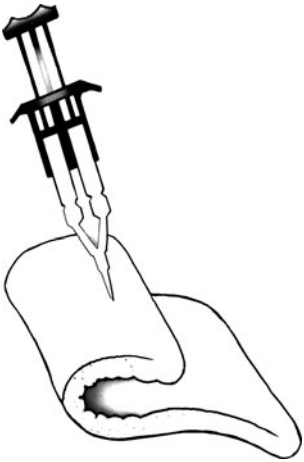


Fig. 6.22 Surgical sealing

7 Minimal Invasive Surgery Principles

7.1 General Considerations

Definition

Minimally invasive surgery may be defined as a method for performing established surgical procedures by remote manipulation in a confined anatomical space. This leads to a reduction of access trauma, thereby reducing surgical complications, accelerating recovery, and improving cosmesis. In order to succeed in this task, it is necessary to gain visual access to the operating site via scopes and operating access for the instruments via working ports.

Philosophy

- Laparoscopy and thoracoscopy have been incorporated into pediatric surgery
- Start with a minimally invasive technique
- Access the diseased site
- Set the diagnosis
- Proceed with therapeutic measures if possible
- Convert if laparoscopic approach is not possible or only after making a compromise

Differences to open surgery

- The surgeon views the monitor rather than the working hands
- The three-dimensional view is reduced to a two-dimensional one

- Size alteration according to distance from the scope
- Limited degree of freedom (DoF)

Advantages of minimal invasive surgery

- Minimal trauma
- Excellent view
- Magnification
- Possibility to explore the entire cavity (abdomen/thorax)
- Wound pain reduction
- Minimal intestinal atony (laparoscopy)
- Rapid postoperative mobilization
- Shorter hospital stay

Problems of minimal invasive surgery

- In spite of its obvious benefits, endoscopic surgery involves new problems and risks that should be acknowledged
- The main one is the learning curve that is experienced by every member of the team for every technique

Prerequisites

- Minimally invasive surgery requires teamwork between the patient, the surgeon, and the team:
  - The patient has to be positioned in such a way that the cavity in which the operation is to take place is sufficiently exposed
  - The surgeon has to be in line with the target organ and the monitor for maximal comfort
  - The team can be on either side of the patient, according to the procedure, and should keep the straight line principle (eyes–target–monitor)
  - The surgeon has to be trained in open surgery and be able to complete the operation

7.2 Instrumentation

- Several firms manufacture instruments for pediatric endoscopic surgery, most of which are shorter and thinner versions of the adult-size instruments in use (Fig. 7.1)

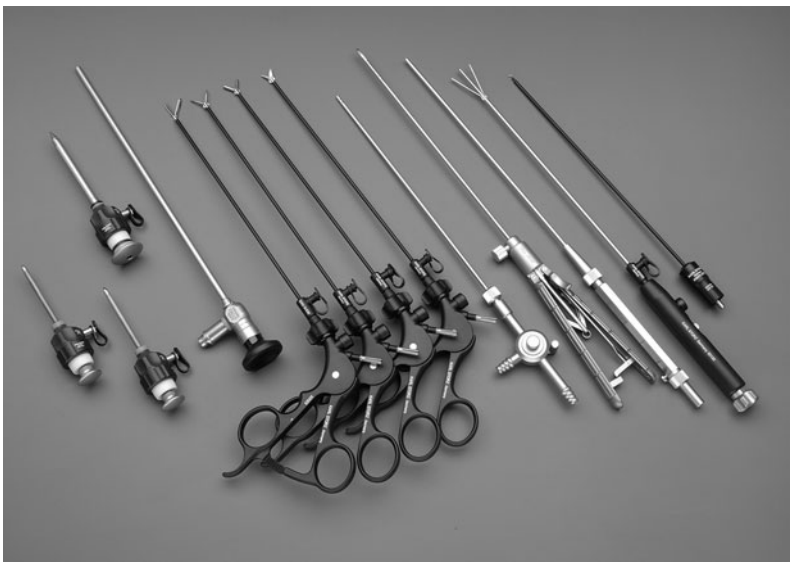


Fig. 7.1 Common laparoscopic instruments (KARL STORZ GmbH)

Endoscopic tower (Fig. 7.2)

- TV monitor connected to a camera
- Halogen light source
- Cable for the telescope
- CO_2 insufflators with control of the filling pressure
- Diathermy generator for bipolar and monopolar use

Fig. 7.2 Endoscopic tower (KARL STORZ GmbH)

7.3 Indications

Diagnostic

- Detection of intraabdominal testes
- Detection of contralateral hernia

- Trauma evaluation
- Intersex evaluation
- Recurrent abdominal pain
- Tumor staing

Therapeutic

- Abdominal
  - Cholecystectomy
  - Appendectomy
  - Meckel's diverticulum
  - Fundoplication
  - Pyloromyotomy
  - Laparoscopically assisted percutaneous endoscopic gastrostomy (PEG)
  - Gastrostomy
  - Jejunostomy
  - Malrotation
  - Colectomy
  - Varicocele
  - Orchidopexy
  - Splenectomy
  - Ovarian surgery
  - Rectosigmoidectomy
  - Nephrectomy
  - Heminephrectomy
  - Adrenalectomy
  - Pyelo-ureteral anastomosis
- Thoracic
  - Pleural empyema
  - Thoracic duplications and cysts
  - Palmar hyperhidrosis
  - Lung biopsy for tumor staging

7.4 Ergonomics

- For optimal performance the surgeon has to select appropriate places for the endoscope and working ports, and position the monitor correctly

Port and instrument sites

- Keep the angle between the instruments at 45°–125°
- Keep the instruments in front of the scope
- The correct positions for instruments in the abdomen are shown in Fig. 7.3
- The correct positions for instruments in the thorax are shown in Fig. 7.4
- Port sites for appendectomy are shown in Fig. 7.5 and those for cholecystectomy in Fig. 7.6

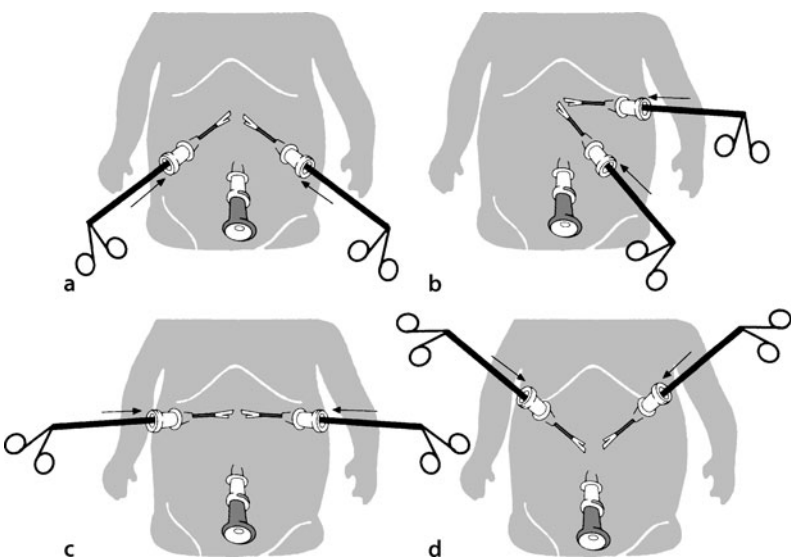


Fig. 7.3a–d Position of instruments in the abdomen

Fig. 7.4 Position of instruments in the thorax

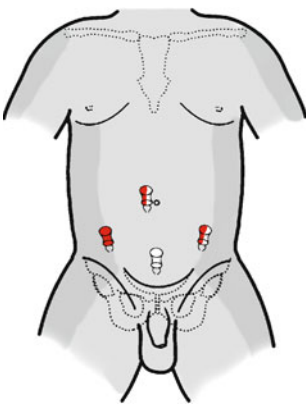


Fig. 7.5 Port sites for appendectomy

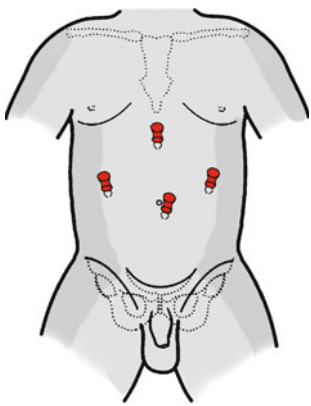


Fig. 7.6 Port sites for cholecystectomy

8 Imaging in Pediatric Surgery

8.1 General Considerations

The famous scholar Wolfgang von Goethe reported, "One only sees what one knows." This quotation has to be complemented with the sentence, "One has to know with which method the findings are best visualized." These methods have to be applied in a stepwise manner and only if they have implications for therapy or the prognosis.

Imaging methods

- Conventional X-ray
- Ultrasonography
- Computer tomography
- **M**agnetic **r**esonance **t**omography = **MR i**maging = **n**uclear **m**agnetic **r**esonance
- Scintigraphy/nuclear imaging

Imaging requirements

- Significance
- Risk
- Comfort for patient
- Costs

Terminology

- Antero-posterior (a.p.): the back of the body is on the film
- Posterio-anterior (p.a.): the front of the body is on the film
- Lateral views right or left: the right or left side of the body is on the film

- Seated, mostly for chest X-rays
- Standing, mostly for abdominal views
- Hanging, in newborns and children who do not stand; the child is held by two helpers under the arm pits on the film
- In bed, lying a.p. or lateral views
- Special positions for defined diseases (i.e., upside down for anal atresia)

8.2 Common Radiographic Images

Plain chest X-ray

- Figure 8.1a–e illustrates lung anatomy
- It is important to be aware of the common sources of error in chest radiographs (Fig. 8.2)

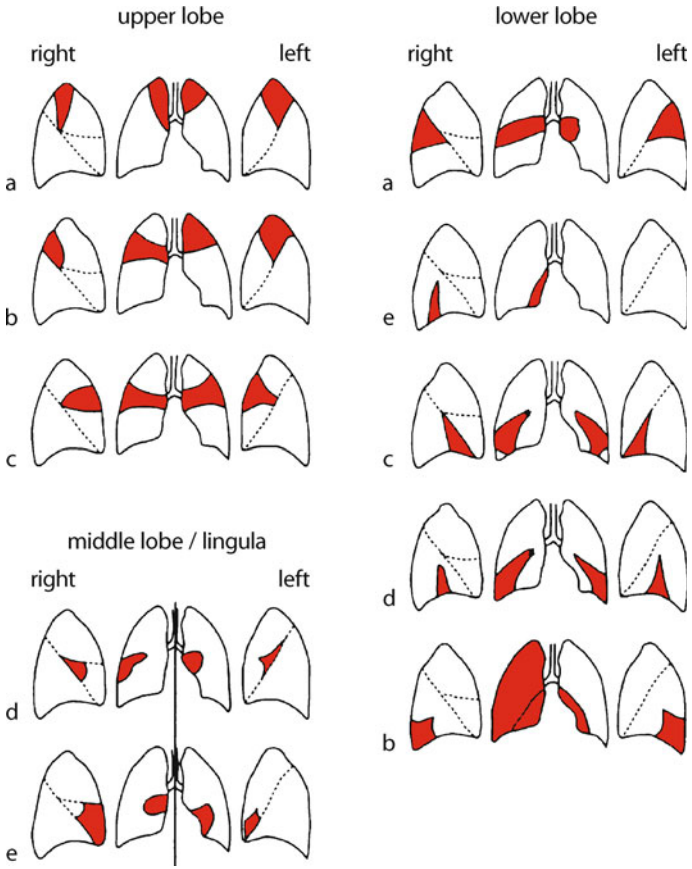


Fig. 8.1a–e Radiologic lung anatomy. **a** Superior, **b** posterior, **c** anterior, **d** lateral, **e** medial

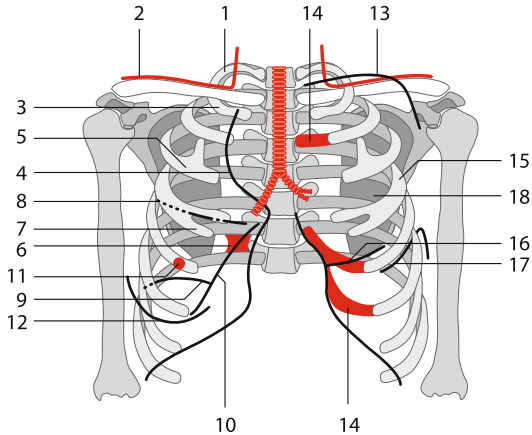


Fig. 8.2 Sources of error in chest X-rays. [*1* Neck rib, *2* sternocleidomastoid muscle, *3* shadow of the first and second rib, *4* azygos vein, *5* incomplete intercostal bridge, *6* complete intercostal bridge, *7* furcation of a rib, *8* interlobar line (upper and middle lobe), *9* accessory interlobar line, *10* cardiac lobe, *11* mammilla, *12* breast shadow, *13* subclavian artery, *14* ossified cartilage, *15* costal sulcus, *16* accessory interlobar line, *17* pectoral shadow, *18* scapula outline]

Age determination

- Age can be determined on the basis of ossification of the hand skeleton (Fig. 8.3)

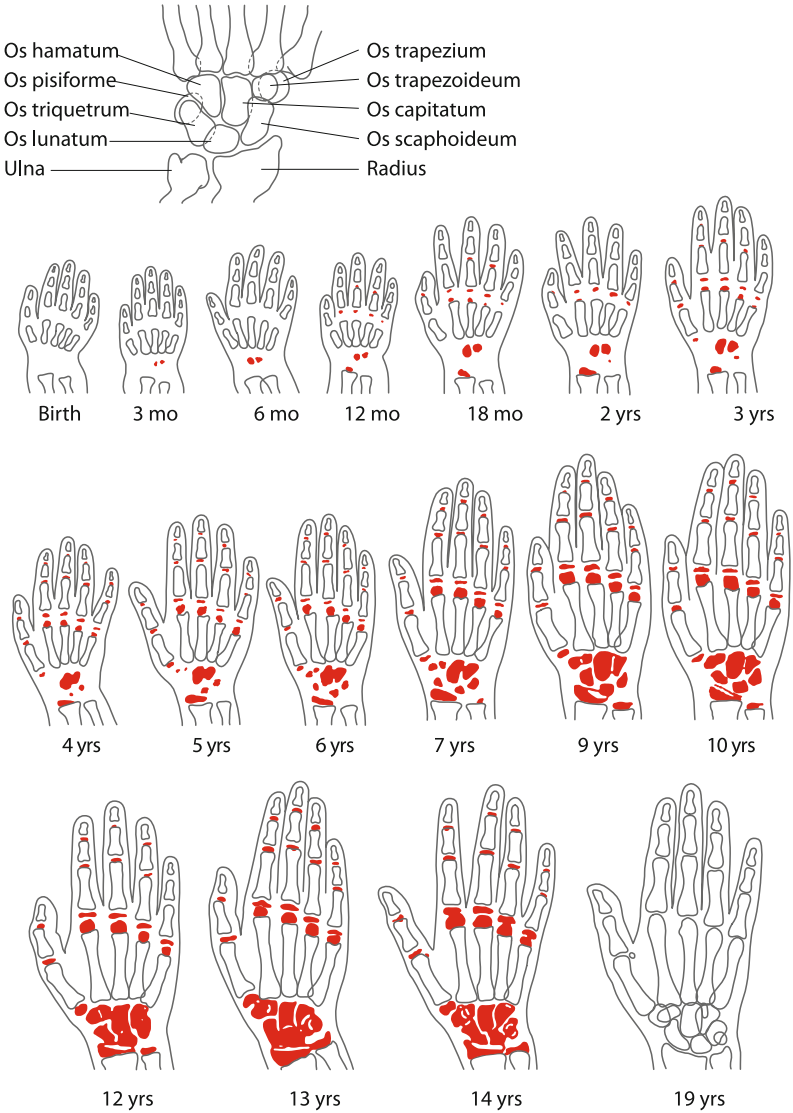


Fig. 8.3 Using radiologic images of the hand to determine age

Organ topography in a CT

- Computed tomography (CT) is an excellent method for imaging organ topography, as the diagrams in Fig. 8.4 demonstrate

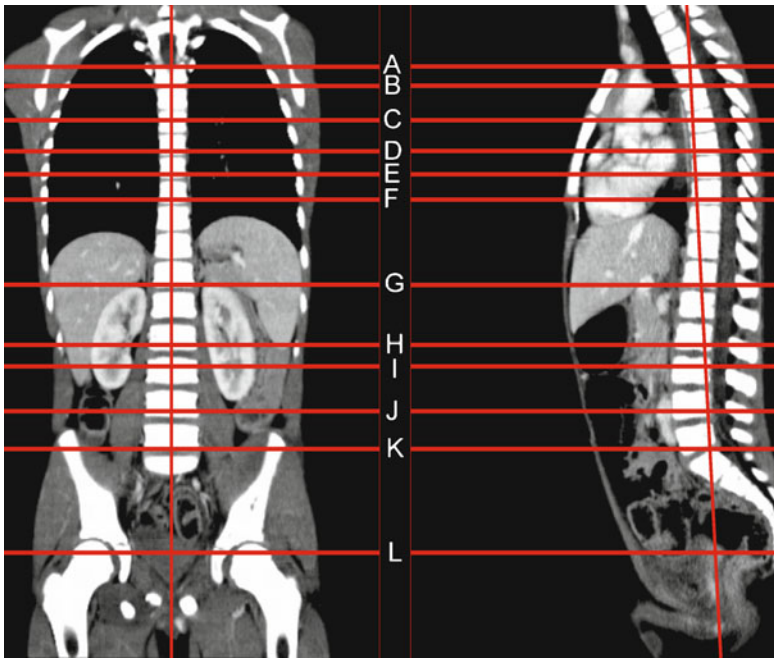


Fig. 8.4 Post-contrast whole-body CT of a 9-, 10-, 12-year-old boy. **Arteries** (*1* aorta, *2* pulmonary trunk, *3* pulmonary artery, *4* brachiocephalic trunk, *5* subclavian artery, *6* common carotid artery, *7* celiac trunk, *8* liac artery, *9* accessory renal artery, *10* superior mesenteric artery, *11* femoral artery). **Veins** (*12* superior vena cava, *13* brachiocephalic vein, *14* azygos vein, *15* inferior vena cava, *16* superior mesenteric vein, *17* portal vein, *18* femoral vein, *19* renal vein). **Organs** (*20* heart, left ventricle, *21* heart, right ventricle, *22* heart, left atrium, *23* heart, right atrium, *24* trachea, *25* main bronchi, *26* lung, *27* thymus, *28* esophagus, *29* diaphragm, *30* liver, *31* stomach, *32* spleen, *33* pancreas, *34* duodenum, *35* jejunum, *36* colon, *37* rectum, *38* adrenal gland, *39* kidney, *40* urinary bladder). **Musculosceletal** (*41* humeral bone, *42* rib, *43* sternum, *44* vertebra, *45* scapula, *46* sacrum, *47* ilium, *48* acetabulum, *49* femoral head, *50* psoatic muscle). Copyright Dr. R. Wolf, Dr. T. Strautz, Dept. of Pediatric Radiology, University Hospital Inselspital, Bern

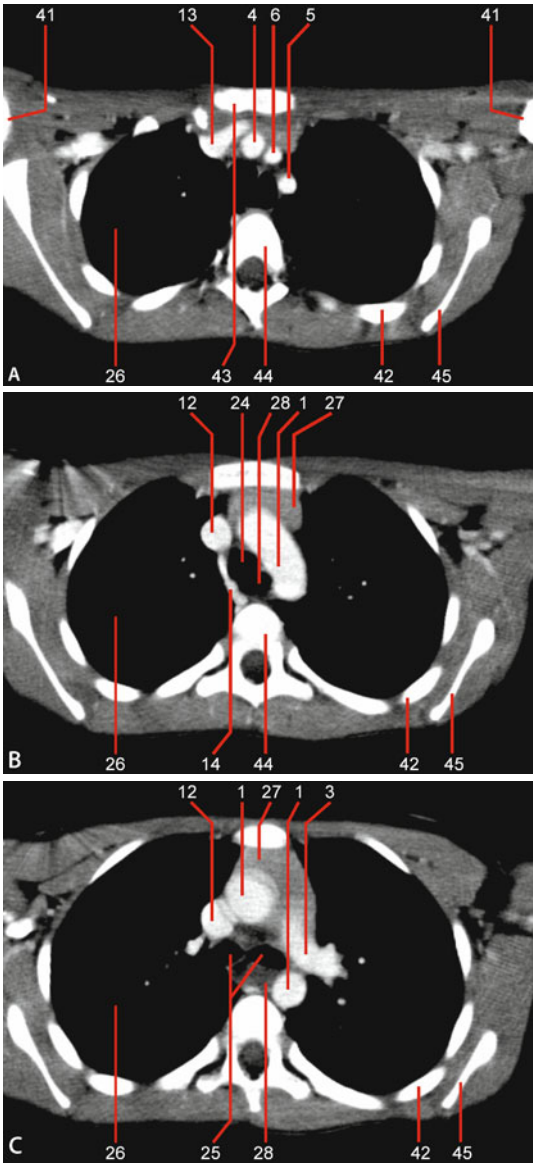


Fig. 8.4 (*continued*) Post-contrast whole-body CT of a 9-, 10-, 12-year-old boy. **Arteries** (*1* aorta, *2* pulmonary trunk, *3* pulmonary artery, *4* brachiocephalic trunk, *5* subclavian artery, *6* common carotid artery, *7* celiac trunk, *8* liac artery, *9* accessory renal artery, *10* superior mesenteric artery, *11* femoral artery). **Veins** (*12* superior vena cava, *13* brachiocephalic vein, *14* azygos vein, *15* inferior vena cava, *16* superior mesenteric vein, *17* portal vein, *18* femoral vein, *19* renal vein). **Organs** (*20* heart, left ventricle, *21* heart, right ventricle, *22* heart, left atrium, *23* heart, right atrium, *24* trachea, *25* main bronchi, *26* lung, *27* thymus, *28* esophagus, *29* diaphragm, *30* liver, *31* stomach, *32* spleen, *33* pancreas, *34* duodenum, *35* jejunum, *36* colon, *37* rectum, *38* adrenal gland, *39* kidney, *40* urinary bladder). **Musculosceletal** (*41* humeral bone, *42* rib, *43* sternum, *44* vertebra, *45* scapula, *46* sacrum, *47* ilium, *48* acetabulum, *49* femoral head, *50* psoatic muscle). Copyright Dr. R. Wolf, Dr. T. Strautz, Dept. of Pediatric Radiology, University Hospital Inselspital, Bern

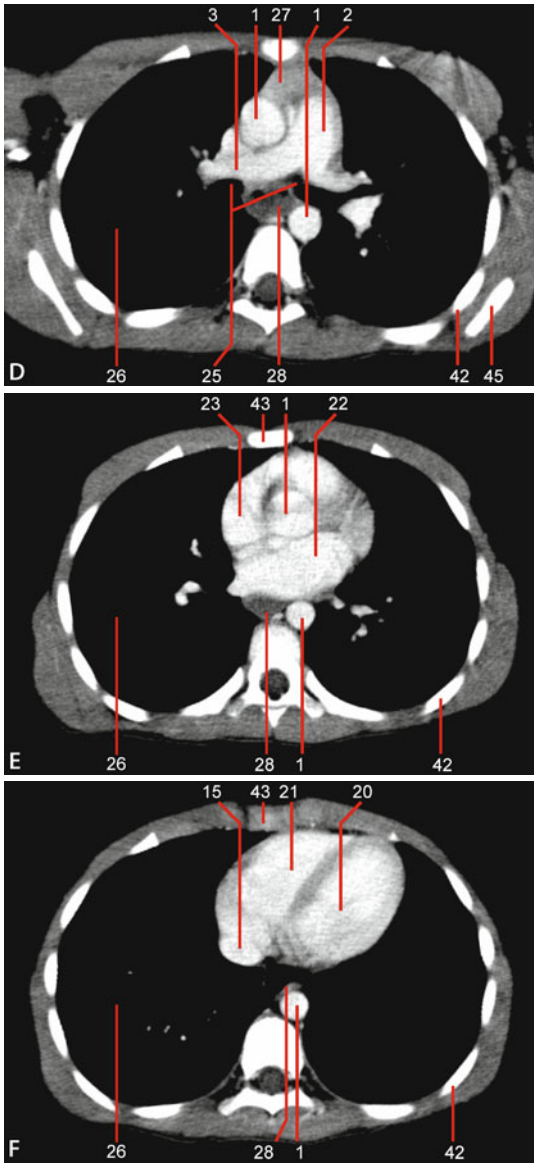


Fig. 8.4 (*continued*) Post-contrast whole-body CT of a 9-, 10-, 12-year-old boy. **Arteries** (*1* aorta, *2* pulmonary trunk, *3* pulmonary artery, *4* brachiocephalic trunk, *5* subclavian artery, *6* common carotid artery, *7* celiac trunk, *8* liac artery, *9* accessory renal artery, *10* superior mesenteric artery, *11* femoral artery). **Veins** (*12* superior vena cava, *13* brachiocephalic vein, *14* azygos vein, *15* inferior vena cava, *16* superior mesenteric vein, *17* portal vein, *18* femoral vein, *19* renal vein). **Organs** (*20* heart, left ventricle, *21* heart, right ventricle, *22* heart, left atrium, *23* heart, right atrium, *24* trachea, *25* main bronchi, *26* lung, *27* thymus, *28* esophagus, *29* diaphragm, *30* liver, *31* stomach, *32* spleen, *33* pancreas, *34* duodenum, *35* jejunum, *36* colon, *37* rectum, *38* adrenal gland, *39* kidney, *40* urinary bladder). **Musculosceletal** (*41* humeral bone, *42* rib, *43* sternum, *44* vertebra, *45* scapula, *46* sacrum, *47* ilium, *48* acetabulum, *49* femoral head, *50* psoatic muscle). Copyright Dr. R. Wolf, Dr. T. Strautz, Dept. of Pediatric Radiology, University Hospital Inselspital, Bern

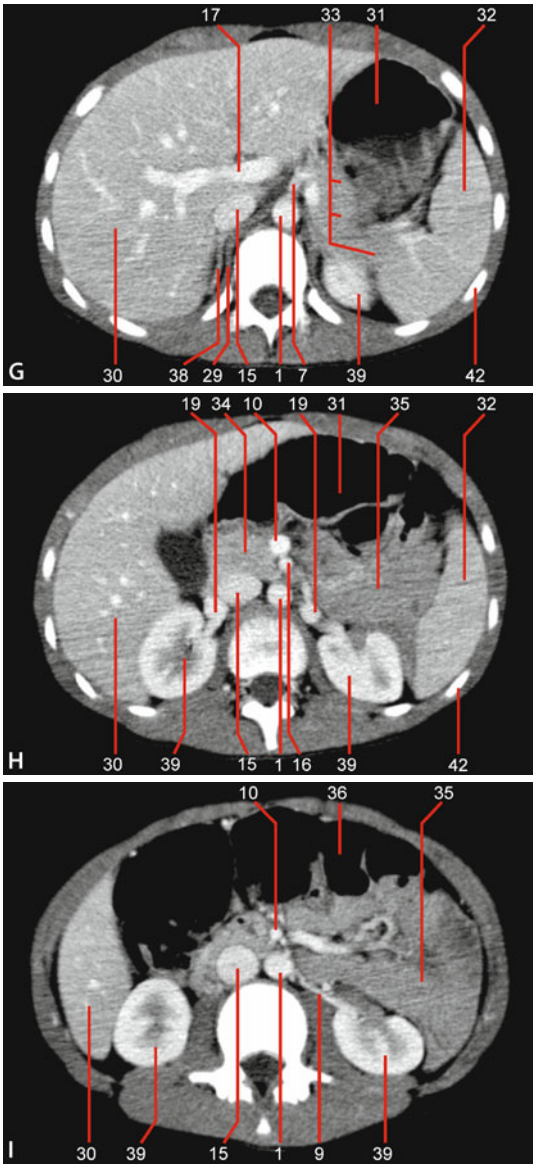


Fig. 8.4 (*continued*) Post-contrast whole-body CT of a 9-, 10-, 12-year-old boy. **Arteries** (*1* aorta, *2* pulmonary trunk, *3* pulmonary artery, *4* brachiocephalic trunk, *5* subclavian artery, *6* common carotid artery, *7* celiac trunk, *8* liac artery, *9* accessory renal artery, *10* superior mesenteric artery, *11* femoral artery). **Veins** (*12* superior vena cava, *13* brachiocephalic vein, *14* azygos vein, *15* inferior vena cava, *16* superior mesenteric vein, *17* portal vein, *18* femoral vein, *19* renal vein). **Organs** (*20* heart, left ventricle, *21* heart, right ventricle, *22* heart, left atrium, *23* heart, right atrium, *24* trachea, *25* main bronchi, *26* lung, *27* thymus, *28* esophagus, *29* diaphragm, *30* liver, *31* stomach, *32* spleen, *33* pancreas, *34* duodenum, *35* jejunum, *36* colon, *37* rectum, *38* adrenal gland, *39* kidney, *40* urinary bladder). **Musculosceletal** (*41* humeral bone, *42* rib, *43* sternum, *44* vertebra, *45* scapula, *46* sacrum, *47* ilium, *48* acetabulum, *49* femoral head, *50* psoatic muscle). Copyright Dr. R. Wolf, Dr. T. Strautz, Dept. of Pediatric Radiology, University Hospital Inselspital, Bern

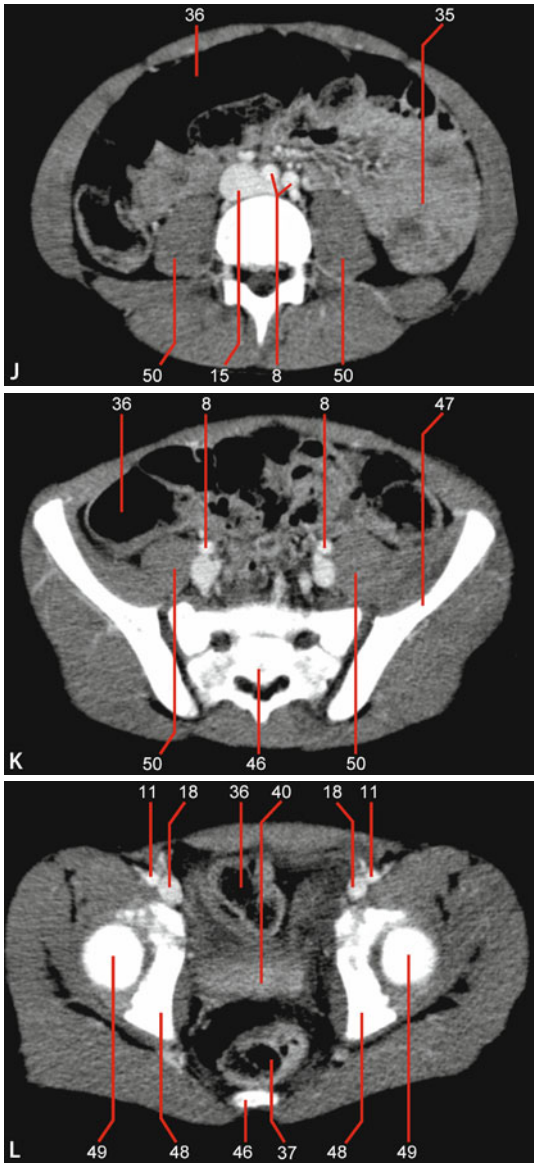


Fig. 8.4 (*continued*) Post-contrast whole-body CT of a 9-, 10-, 12-year-old boy. **Arteries** (*1* aorta, *2* pulmonary trunk, *3* pulmonary artery, *4* brachiocephalic trunk, *5* subclavian artery, *6* common carotid artery, *7* celiac trunk, *8* liac artery, *9* accessory renal artery, *10* superior mesenteric artery, *11* femoral artery). **Veins** (*12* superior vena cava, *13* brachiocephalic vein, *14* azygos vein, *15* inferior vena cava, *16* superior mesenteric vein, *17* portal vein, *18* femoral vein, *19* renal vein). **Organs** (*20* heart, left ventricle, *21* heart, right ventricle, *22* heart, left atrium, *23* heart, right atrium, *24* trachea, *25* main bronchi, *26* lung, *27* thymus, *28* esophagus, *29* diaphragm, *30* liver, *31* stomach, *32* spleen, *33* pancreas, *34* duodenum, *35* jejunum, *36* colon, *37* rectum, *38* adrenal gland, *39* kidney, *40* urinary bladder). **Musculosceletal** (*41* humeral bone, *42* rib, *43* sternum, *44* vertebra, *45* scapula, *46* sacrum, *47* ilium, *48* acetabulum, *49* femoral head, *50* psoatic muscle). Copyright Dr. R. Wolf, Dr. T. Strautz, Dept. of Pediatric Radiology, University Hospital Inselspital, Bern

8.3 Nomograms for Organ Size

- The nomogram for spleen volume is given in Fig. 8.5
- The nomogram for kidney volume in infants is given in Fig. 8.6; that for children is given in Fig. 8.7
- The hip joint is shown in Fig. 8.8, and classification of standard hip joint measurements and required treatments are given in Table 8.1

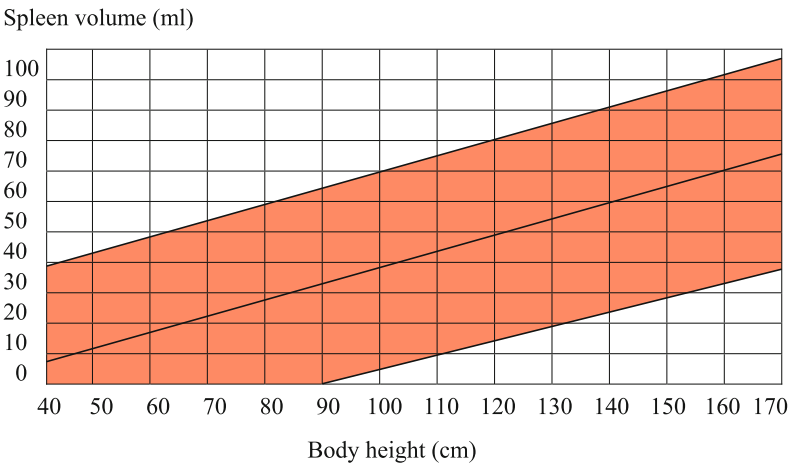


Fig. 8.5 Nomogram for spleen volume

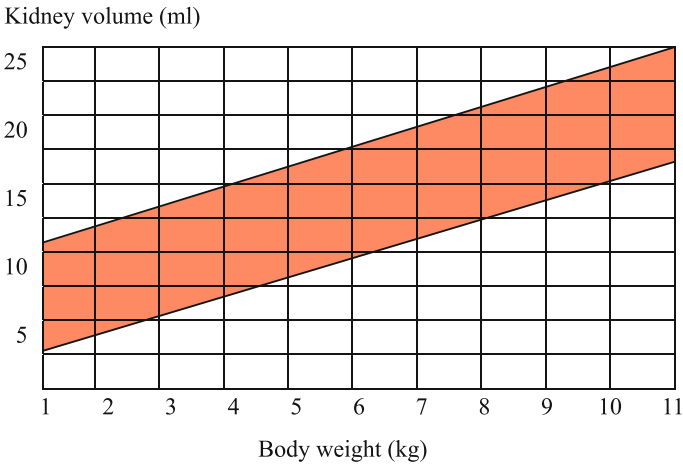


Fig. 8.6 Nomogram for kidney volume in infants

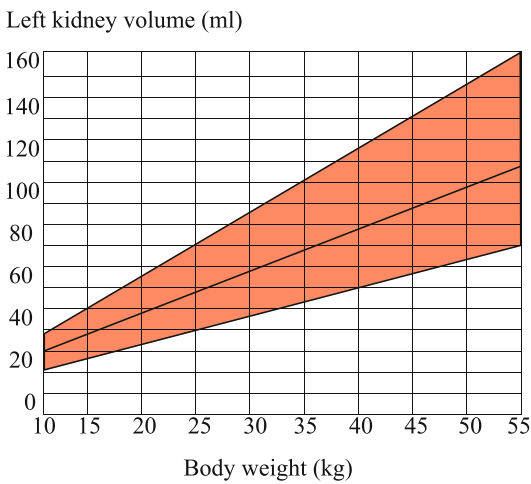


Fig. 8.7 Nomogram for kidney volume in children

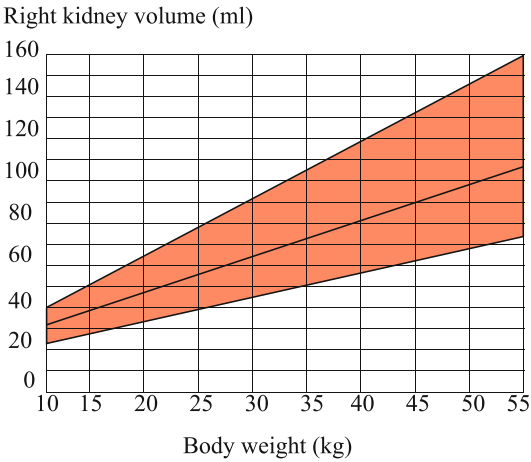


Fig. 8.7 *(continued)* Nomogram for kidney volume in children

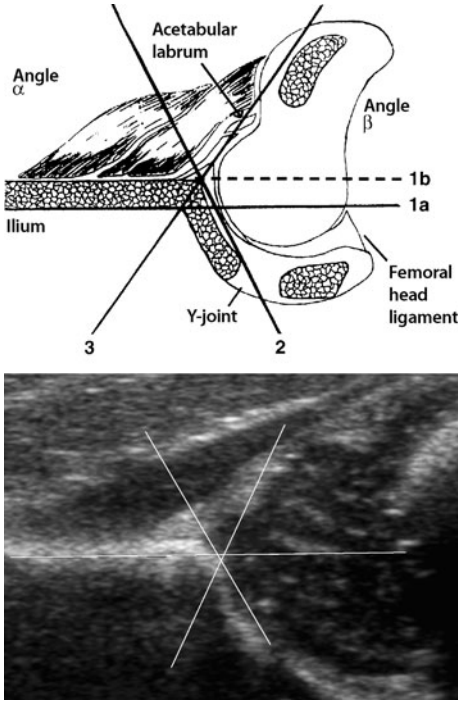


Fig. 8.8 Hip joint diagram and ultrasonographic image. Guidelines indicating the ilium thickness (*1a, 1b*) and defining the acetabulum (*2, 3*) are shown

Table 8.1 Classification of hip joint measurements and required treatment

Type	Classification by angle		Consequence
	α	β	
Ia	> 60°	< 55°	None
Ib	> 60°	> 55°	None
IIa	50°–59°	> 55°	Control Abduction therapy
IIb	50°–59°	> 55°	Abduction therapy
II "g" or "c"	43°–49°	70°–77°	Abduction therapy
Descended joint	43°–49°	> 77°	Immediate therapy
IIIa	< 43°	> 77°	Reposition
IIIb	< 43°	> 77°	Reposition
IV	< 43°	> 77°	Reposition

8.4 Conventional X-rays

- Chest radiograph findings in neonates are detailed in Table 8.2
- Gastrointestinal radiograph findings in neonates are detailed in Table 8.3

Table 8.2 Chest radiograph findings in neonates. (*CCAM* congenital cystic adenomatoid malformation, *CDH* congenital diaphragmatic hernia, *GI* gastrointestinal, *TEF* tracheo-esophageal fistula)

Disease	Patient's position	X-ray's characteristic signs
Esophageal atresia	Prone a.p. position with nasoesophageal tube	Air bubble at the upper thorax aperture
▪ With TEF		Air in stomach
▪ Without TEF		Complete absence of air in GI tract
▪ Isolated TEF	Prone a.p. position with nasogastric tube that is slowly withdrawn whilst contrast medium is injected	Fistula between esophagus and trachea
Esophageal stenosis	Decubitus position – oral contrast	Stenosis of the esophagus with prestenotic dilatation
Congenital lobar emphysema	Upright p.a. and lateral view	Hyperlucent and over-expanded lung with contralateral mediastinal shifting (common in left upper lobe)
Bronchogenic cysts	Upright p.a. and lateral views	Cystic lesion within the lung Air-fluid level indicates communication with airways
CDH	Upright a.p. position	Intestinal loops and stomach in the left hemithorax as well as mediastinal shifting to the right side with compression of the contralateral lung
CCAM	Upright a.p. position	Diffuse air-filled cavities within one or two lobes Normal-shaped diaphragm
Pneumothorax	Upright a.p. position	Partial or complete lung collapse Mediastinal shifting Depression of diaphragm

Table 8.3 Gastrointestinal radiograph findings in neonates

Disease	Patient's position	X-ray's characteristic signs
Pyloric stenosis	Upright a.p. position	Distended stomach and scanty air in the GI tract
Duodenal atresia	Upright hanging a.p. position	Double-bubble sign indicating the distended stomach and first portion of duodenum
High jejunal atresia	Upright hanging a.p. position	Typical double-bubble sign and a few air-fluid levels in the upper abdomen, no distal air
Low small bowel atresia	Upright hanging a.p. position	Significant air-fluid levels with intestinal and abdominal distension
High bowel stenosis	Upright a.p. position Contrast radiography	Air-fluid levels in the upper abdomen and scanty air within the colon A large proximal bowel and small portions of contrast percolating through the stenosis into the distal bowel
Contrast enema	Upright a.p. position and lateral view	Unused (non-used) microcolon
Meconium ileus	a.p. view in supine position	Moderate to significant loop distension with air-fluid levels Pearl-chain sign (viscid meconium at distal ileum) Soap bubble sign (air mixed with thick meconium)
NEC ▪ Stage I ▪ Stage II ▪ Stage III	Upright a.p. position	Nonspecific bowel distension with mild ileus Bowel distension, intestinal pneumatosis Free intraperitoneal gas
Hirschsprung's disease	Upright a.p. position Contrast radiography a.p. and lateral views	Several distended intestinal loops in the abdomen Dilated proximal colon segment Spastic distal colon segment with typical transition zone
Anorectal malformations	Upright a.p. position and lateral film of the pelvis	Sacral ratio (normal value 0.74–0.77)

8.5 Ultrasonography

Nowadays ultrasonography is the most important imaging technique for pediatric patients, as it has no side-effects and enables quick orientation (Tables 8.4, 8.5). Ultrasonography is called the physician's stethoscope of the twenty-first century. The main problem of ultrasonographic examinations is that many physicians are not adequately trained, leading to a lot of false-positive and false-negative results. The false-positive findings lead to further unnecessary examinations, such as a CT scan, which exposes the patient to irradiation and increases costs. The false-negative findings delay diagnosis.

Ultrasonography is also important in antenatal evaluation for many congenital defects (Table 8.6), and thus improves pediatric surgical patients' perinatal care (Table 8.7).

Table 8.4 Ultrasonographic diagnostic values and pathological findings

Organ	Normal values	Pathological finding
Liver	Traversal length ≤16 cm Sagittal length ≤15 cm Portal vein width ≤11 mm ▪ Homogenous echo texture ▪ Sharp borders ▪ Anechoic lumen of portal vein branches with echogenic walls ▪ Anechoic lumen of the hepatic vein draining to the inferior vena cava	▪ Solid mass (tumors, metastases, hemangioma) ▪ Cystic (dysontogenetic, parasitic) ▪ Liquid mass (hematoma, abscess, bilioma) ▪ Hepatomegaly ▪ Fatty infiltration ▪ Portal hypertension ▪ Biliary atresia ▪ Budd–Chiari syndrome ▪ Trauma – intraparenchymal lesion
Gall bladder	Length 3–10 cm Thickness ≤4 cm Wall ≤3 mm Common bile duct ≤7 mm ▪ Sharp border ▪ Anechoic ▪ Postprandial contraction	▪ Stones (echogenic focus) ▪ Sludge phenomena (hepatization of the gallbladder) ▪ Hydrops, empyema (intraluminal gas) ▪ Ectopia ▪ Agenesis ▪ Thickening in inflammation ▪ Polyps, tumors ▪ Bile duct dilatation (obstructive, cystic) ▪ Atresia

Table 8.4 *(continued)* Ultrasonographic diagnostic values and pathological findings

Organ	Normal values	Pathological finding
Pancreas	Diameter 0.7–2.5 cm Duct diameter ≤1.3 mm ▪ Homogenous echo texture ▪ Superior mesenteric vein is a marker of division between head and body	▪ Solid mass (tumors) ▪ Cystic mass (dysontogenetic, pseudocysts) ▪ Pancreatic contusion ▪ Traumatic rupture ▪ Ductal stones ▪ Acute pancreatitis ▪ Fatty infiltration
Spleen	Length 11 cm Thickness 4 cm Width 7 cm ▪ Parenchymal, homogenous echo texture, consisting of low level echoes, with slightly lower reflexivity compare to the liver	▪ Splenomegaly (infection, sepsis, lymphoma, hematological diseases, portal hypertension) ▪ Solid mass (tumor, metastasis) ▪ Cystic (dysontogenetic, parasitic) ▪ Trauma (intraparenchymal and subcapsular hematoma, spleen rupture, free peritoneal fluid)
Kidney	Length 11 cm Thickness 5 cm Width 6 cm ▪ Echogenic ▪ High reflective renal pelvis	▪ Malformation (fetal lobulation, duplex collecting system, pancake and horseshoe kidney, cystic and polycystic kidney) ▪ Pathologic conditions (renal calculi, hydronephrosis) ▪ Solid masses (benign and malignant tumors – Wilms) ▪ Trauma – intraparenchymal and subcapsular hematoma, partial or total rupture
Adrenal glands	Length 3–5 cm Thickness 2–5 cm Width 1–3 cm ▪ Hypoechogenic ▪ Central thin and more reflective medulla	▪ Adenoma ▪ Neuroblastoma
Ureter	Visible when dilated	▪ Megaureter ▪ Ureterocele ▪ Vesicoureteric obstruction
Bladder	▪ Post micturition bladder volume <20–30 ml normal	▪ Cystitis (wall thickening >5 mm)

Table 8.4 (*continued*) Ultrasonographic diagnostic values and pathological findings

Organ	Normal values	Pathological finding
Testis	▪ Homogenous ovoid structure ▪ Size depends on age ▪ Head of epididymis adjacent to rete testis	▪ Atrophic testis (cryptorchidism, after herniotomy) ▪ Symptomatic hydrocele (torsion, epididymoorchitis) ▪ Trauma (intratesticular hematoma) ▪ Tumors (solid – lower echodensity, cystic – spermatocele)

Table 8.5 Postnatal ultrasonographic findings

Disease	Technical details	Ultrasonographic pattern/ pathologic values
Gastroesophageal reflux	Longitudinal scanning to the left of the aorta, demonstrating the gastroesophageal junction Recording reflux episodes through the cardia by feeding	More than 3–4 reflux episodes in 10 min
	Intraabdominal esophageal segment	<18 mm
Hypertrophic pyloric stenosis	Investigation in supine position with some fluid in the stomach	Muscle wall thickness > 4 mm Canal length > 15 mm
Ladd's syndrome		Whirlpool sign – spiral-like position of the small intestine, containing air bubbles
	Supine position	Target sign – concentric rings of high and low echogenicity, demonstrated as pseudokidney at the long axis
Appendicitis	Scanning in the point of maximal tenderness	Fluid-filled and rigid appendicular structure > 0.7 mm diameter Echogenic periappendicular mass

Table 8.6 Management of antenatal diagnosed malformations

Malformation	Procedure required
Anencephaly	**Pregnancy termination**
Hydranencephaly	
Bilateral pulmonary agenesis	
Bilateral renal agenesis	
Polycystic disease	
Severe skeletal anomalies	
Unilateral polycystic lung	**Correction after term normal delivery**
Small intact omphalocele/meningocele	
GI tract atresias	
Meconium ileus	
Intraabdominal benign cysts	
Hydronephrosis	
Small sacrococcygeal teratoma	
CDH	
Cystic hygroma	
Giant omphalocele	**Correction after cesarean delivery**
Gastroschisis	
Large/ruptured meningocele	
Large sacrococcygeal teratoma	
Large/ruptured meningocele	**Preterm delivery indicated**
Large sacrococcygeal teratoma	
Conjoined twins	
Hydrocephalus	
CDH	**Intrauterine correction possible (still experimental)**
Giant cystic adenomatoid malformation	
Airway obstruction	
Giant sacrococcygeal teratoma	

Table 8.7 Antenatal ultrasonographic findings

Malformations	Technical details	Ultrasonographic pattern
Cerebral malformations		
Hydrocephalus	Horizontal section through the lateral ventricles	Ventriculomegaly > 12–15 mm transverse
Meningomyelocele	Sagittal section through the spine	Irregular spine anatomy
Encephalocele	Sagittal section through the occipital region	Irregular cranio-cerebral anatomy
Spina bifida	Sagittal and horizontal section through the spine	Spine defect
Chest cavity		
CDH	Horizontal section through the thorax	Intestinal loops in the left thorax
CCAM		Dense, multicystic lesion of one or two lobes, mediastinal shifting
Esophageal atresia		Fluid-filled pouch in the upper thorax aperture; the stomach is not visible
Bronchogenic cyst		Simple cystic lesion within the lung tissue
Abdomen		
Duodenal atresia	Horizontal section through the abdomen	Double-bubble sign
Small-bowel obstruction		Bowel dilatation > 17 mm, mural thickness > 3 mm, increased peristalsis
Meconium peritonitis		Bowel dilatation, intraperitoneal calcification
Omphalocele/ Gastroschisis	Sagittal and horizontal section through the abdomen	Protruding liver in the amniotic sac and floating intestinal loops
Intraabdominal masses		Cystic or solid masses (ovarian or mesenteric cyst)
Anal obstruction	Sagittal section (transvaginal approach)	Dilated rectum and increased fluid with intraluminal calcifications when a recto-vesical fistula is present

Table 8.7 (*continued*) Antenatal ultrasonographic findings

Malformations	Technical details	Ultrasonographic pattern
Urinary tract anomalies		
Cystic lesions		Unilateral or bilateral cystic lesions with normal amniotic fluid or oligohydramnios
Obstructive lesions	Horizontal section through the abdomen	Proximal obstruction (hydronephrosis), distal obstruction (hydroureteronephrosis), with or without bladder dilation

Documentation of ultrasonography

- In an attempt to reduce errors due to inter-operator variability, there are guidelines for ultrasonographic imaging procedures (Table 8.8)
- The volume of an organ can be estimated from ultrasonographic images using the ellipsoid equation: $V_o = L \cdot W \cdot [(L_1 + L_2)/2] \cdot 0.523$, where, V_o is organ volume, L is length, W is width, L_1 is the longest longitudinal cross-section, and L_2 is the longest transverse cross-section

Table 8.8 Ultrasonography guidelines

Cross-section	View on the screen as seen by the examiner	
	Left	Right
Longitudinal	Cranial	Caudal
Transverse		
▪ Supine position	Right side	Left side
▪ Prone position	Left side	Right side
Cranium	Frontal	Occipital
Non-standardized cross-sections have to be defined by marking the image		

9 Expert Opinion

9.1 General Considerations

Prerequisites

- Specific request awarded (mandate)
- Expert report for health or other type of insurance, for the court or a lawyer
- The patient has to be informed and has to give authorization for the release of their medical information
- The physician has to be objective and possess professional competence

Structure of a medical expert report

- The document should include:
  - The name, the position, and the address of the expert
  - The date and time of the patient's examination
  - The name, date of birth, address, and occupation of the patient
  - Any patient-relevant file references as well as the date of accident or incident
  - The name of the constituent and the date, stating the reason for the expert's report
  - Documents used for the report should be listed (X-rays, hospital reports, etc.)
- A list of the diagnoses
- A list of the therapeutic procedures prescribed
- Description of the disease or accident, and the therapy prescribed

- Present symptoms and ailments
- The present results of the physical examination (i.e., anthropometric measures made using the neutral-0 method; Figs. 9.1–9.9)
- The results of present examinations (i.e., X-rays, ECG, MRI, etc.)
- A summary of the present findings
- An interpretation of the present situation including its relationship to the initial presenting disease
- Answers to specific questions asked by the patient

9.2 Examination of the Musculoskeletal Apparatus

Spine

- Inspection
  - Straight spine
  - Shoulders level with iliac crests
  - Physiological spinal curvatures
- Palpation
  - Mobile skin
  - Strong musculature
  - Normal pressure sensitivity
  - Spinal processes in a row
- Mobility
  - Spine freely mobile?
- Gait and stand
  - Freely mobile spine whilst walking and standing

Upper leg

- Inspection
  - Shoulders level
  - Symmetrical muscle mass
  - Normal skin color (after the arms have been hanging down for at least 5 min)

- Palpation
  - Mobile, tight skin
  - Adequate muscle tension
  - Normal pressure sensitivity
  - Pulses equal bilaterally?
  - Palpable lymph nodes?
- Mobility
  - Pain-free mobility
- Hand function
  - Symmetric
  - Homogenous skin color and temperature
  - Normal finger nails
  - Normal sensibility
  - Normal movement
  - Normal fist closure
  - Normal tweezers and clamp grip

Lower leg

- Inspection
  - Iliac crests level
  - Symmetrical muscle mass
  - Normal foot arch
- Palpation
  - Mobile, tight skin
  - Adequate muscle tension
  - Normal pressure sensitivity
  - Pulses equal bilaterally?
  - Palpable lymph nodes?
- Mobility
  - Pain-free mobility
- Gait and stand
  - Free gait on even terrain
  - Equal step size whilst walking
  - Symmetrical scrolling of the knee and ankle whilst walking
  - Secure standing on one leg

9.3 Anthropometric Measures

Neck

- Neck measurements made using the neutral-0 method are shown in Fig. 9.1
  - Lateral inclination right and left
  - Flexion/extension
  - Rotation

Spine

- Spine measurements made using the neutral-0 method are shown in Fig. 9.2
  - Axial rotation
  - Lateral inclination in the frontal plane
  - Active erection from the prone position
  - Schober sign
    - On an erect person mark a position on the skin over the spine 30 cm distal to C7, and another one 10 cm proximal of S1
    - The person should bend forwards with stretched knees. Measure the distance between C7 and the first mark ($30 + x$ cm) as well as S1 and the second mark ($10 + x$ cm) (Fig. 9.2)

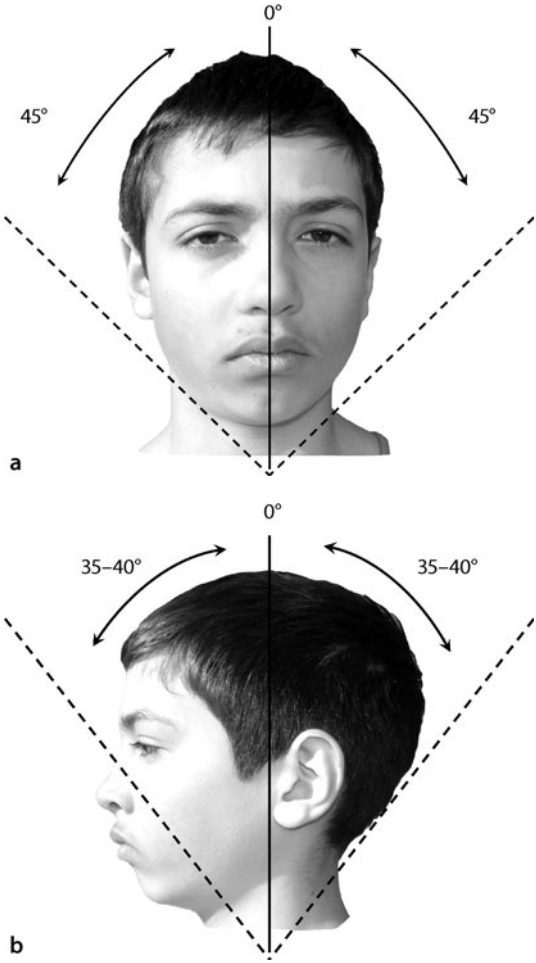


Fig. 9.1a–c Neck measurements (neutral-0 method). **a** Lateral declination of cervical spine 45°/0°/45°. **b** Forward, backward declination of cervical spine 35°–45°/0°/35°–45°. **c** Rotation of cervical spine 60°–80°/0°/60°–80°

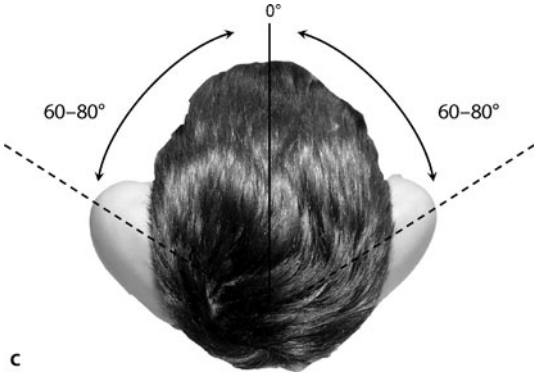


Fig. 9.1a–c (*continued*) Neck measurements (neutral-0 method). **a** Lateral declination of cervical spine 45°/0°/45°. **b** Forward, backward declination of cervical spine 35°–45°/0°/35°–45°. **c** Rotation of cervical spine 60°–80°/0°/60°–80°

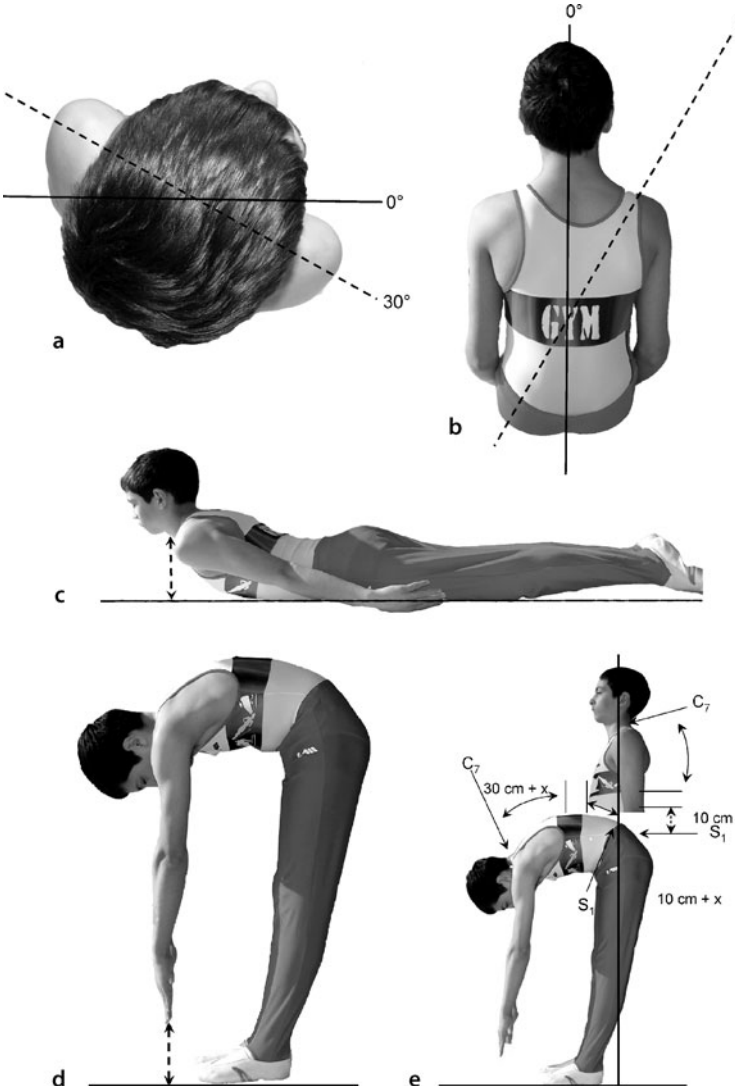


Fig. 9.2a–e Spine measurements (neutral-0 method). **a** Rotation of spinal column 30°/0°/30°. **b** Lateral declination of spinal column 30°–40°/0°/30°–40°. **c** Distance from the jugulum to the floor after lifting the head actively from a prone position. **d** Distance from the finger tips to the floor when bending with straight legs. **e** Schober sign

Shoulder

- Shoulder measurements made using the neutral-0 method are shown in Fig. 9.3
  - Sideward/toward the body
  - Toward the spine/forward
  - Outward/inward twisting (upper arm adjacent)
  - Outward/inward twisting (upper arm 90° abduction)

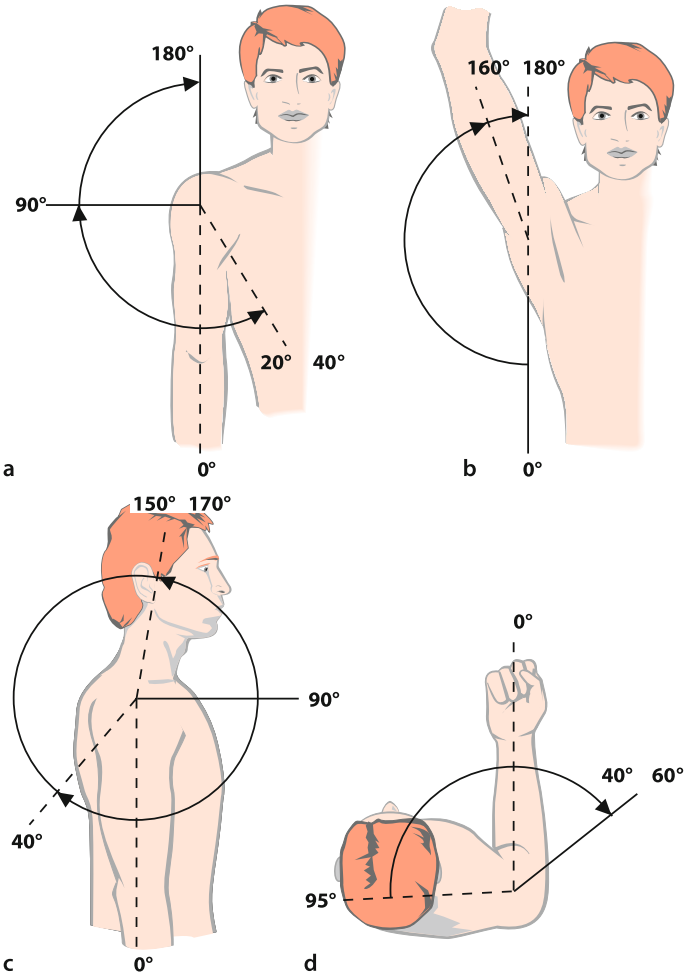


Fig. 9.3 Shoulder measurements (neutral-0 method)

Elbow

- Elbow measurements made using the neutral-0 method are shown in Fig. 9.4
  - Flexion/extension

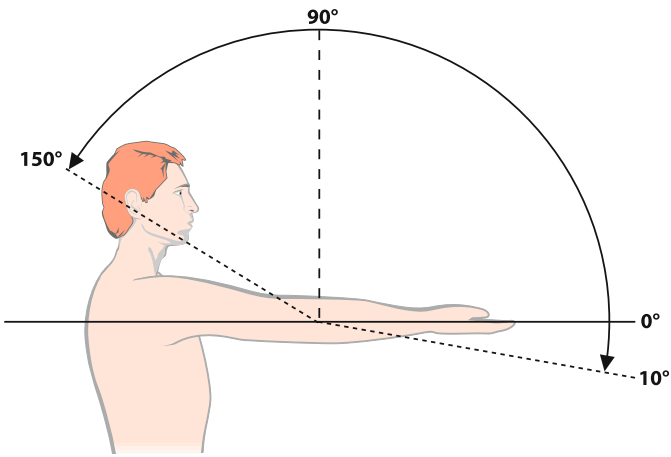


Fig. 9.4 Elbow measurements (neutral-0 method)

Forearm

- Forearm measurements made using the neutral-0 method are shown in Fig. 9.5
  - Outwards/inwards twisting

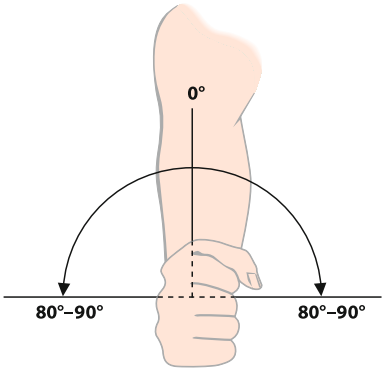
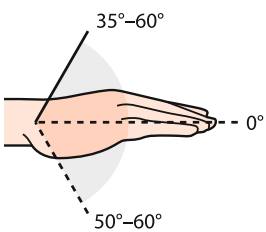


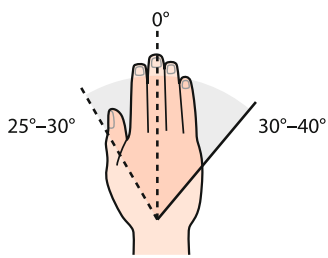
Fig. 9.5 Forearm measurements (neutral-0 method)

Hand

- Hand measurements made using the neutral-0 method are shown in Fig. 9.6
  - Pronation/supination
  - Flexion/extension
  - Sidewise
  - Lateral/medial bending



Towards back of hand/palmar



Towards ulnar/radius

Fig. 9.6 Hand measurements (neutral-0 method)

Hip

- Hip measurements made using the neutral-0 method are shown in Fig. 9.7
  - Flexion/extension
  - Abduction/adduction
  - Outward/inward twisting (knee bend at 90°)
  - Outward/inward twisting (thigh stretched)

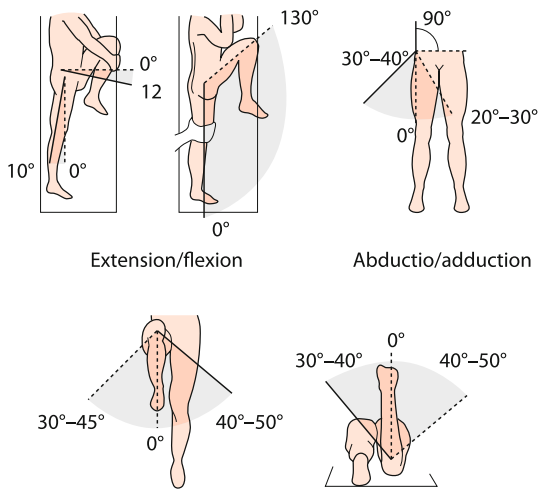


Fig. 9.7 Hip measurements (neutral-0 method)

Knee

- Knee measurements made using the neutral-0 method are shown in Fig. 9.8
  - Flexion/extension

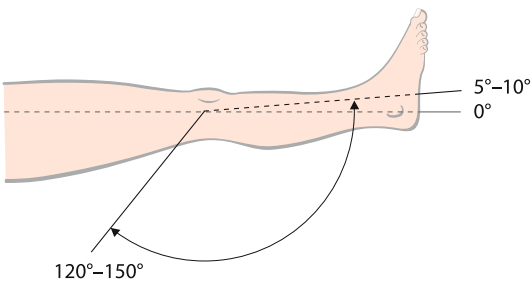


Fig. 9.8 Knee measurements (neutral-0 method)

Ankle joints

- Ankle joint measurements made using the neutral-0 method are shown in Fig. 9.9
  - Flexion/extension
  - Pronation/supination

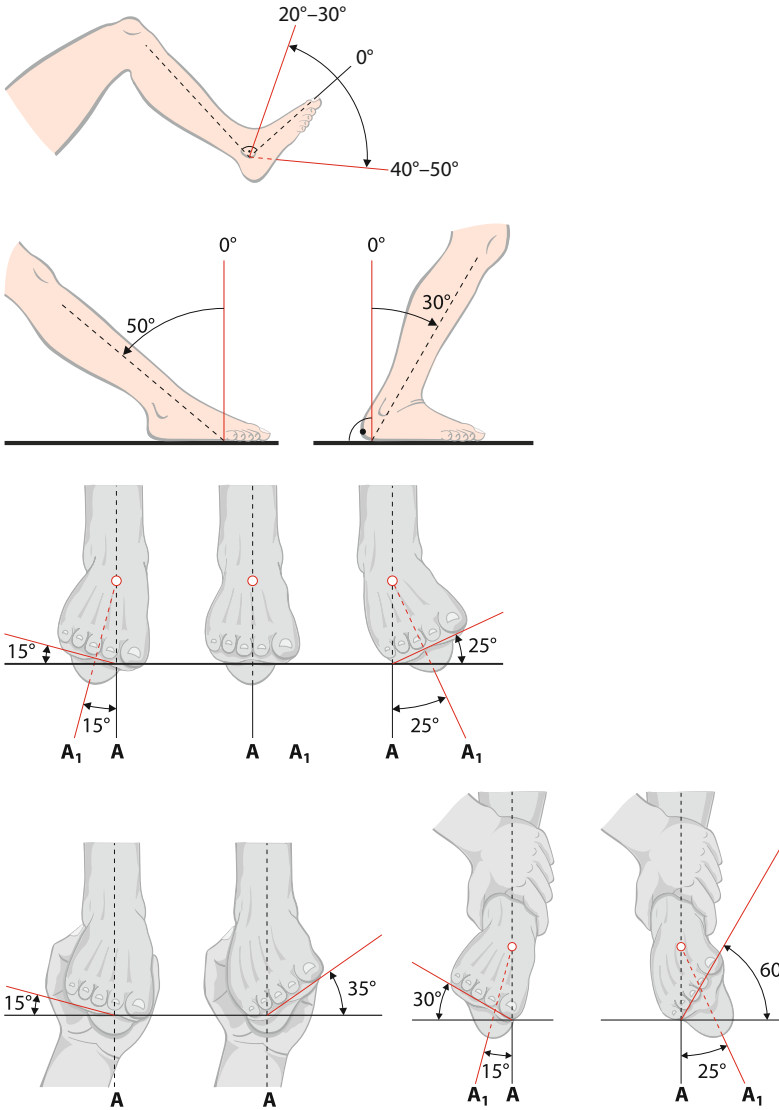


Fig. 9.9 Ankle joint measurements (neutral-0 method)

Size measurement

- Table 9.1 indicates where to measure the circumference of the extremities (see also Fig. 9.10)
- Table 9.2 shows where to measure their length

Table 9.1 Circumference measures of the extremities

Location	Point for the circumference measurement
Upper arm	15 cm above the lateral epicondyle of the humerus
Elbow	At the level of the elbow joint, arm in stretch position
Forearm	10 cm and 20 cm under the epicondylus of the humerus
Hand	Distal of the styloid process of the radius
Middle hand	At the level of the metacarpal joints II–V
Finger	In the middle of the phalanx of each finger
Finger joints	At the level of each finger joint
Thigh	10 cm and 20 cm above the knee joint space
Knee	At the level of the knee joint, leg in stretch position
Lower leg	15 cm below the medial part of the knee joint
Ankle	Directly over the ankle
Fore foot	At the level of the metatarsal joints II–V

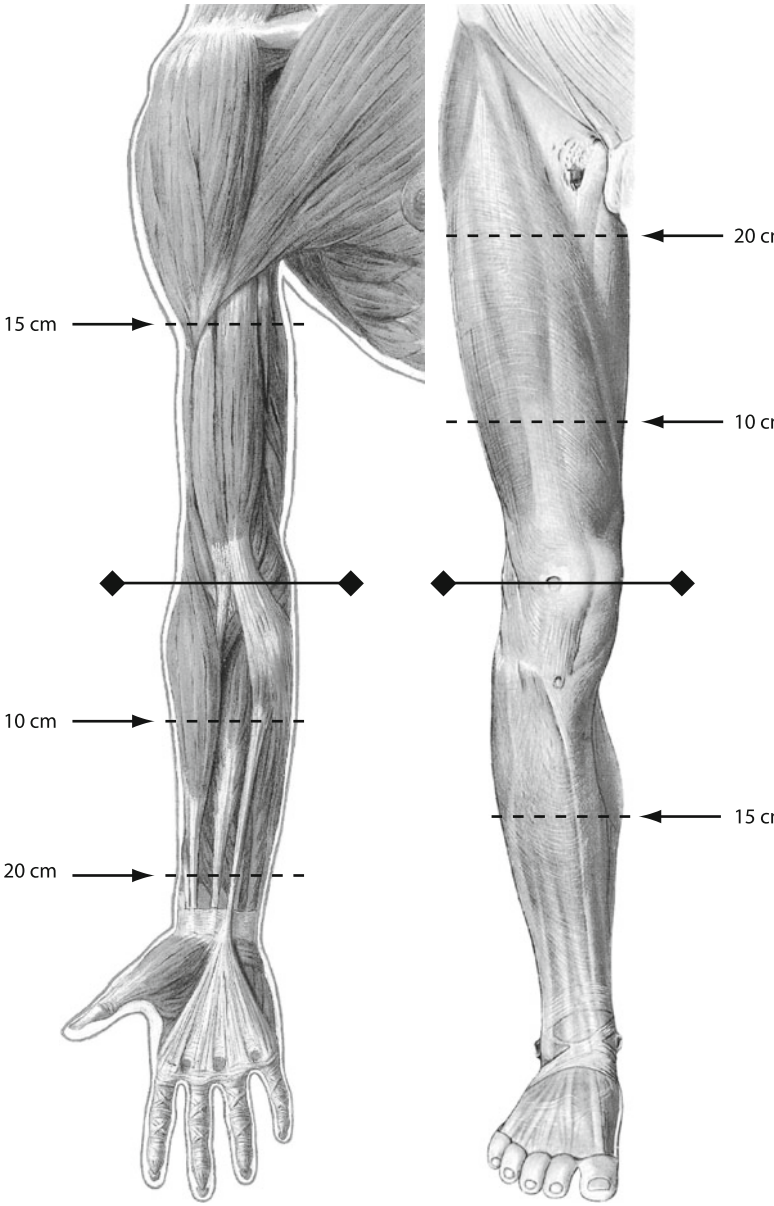


Fig. 9.10 Where to measure the circumferences of extremities

Table 9.2 Length measures of the extremities

Location	Distance from
Arm	Acromion to the styloid process of the radius
Upper arm	Acromion to the lateral epicondylus of the humerus
Forearm	Epicondylus of the humerus to the styloid process of the radius
Ulna	Olecranon to the styloid process of the ulna
Hand	Styloid process of the radius/ulna to D3 fingertip
Finger	Flexed hand (metacarpal) to the finger tips
Amputation stump	Acromion to the stump end (upper arm) Lateral epicondylus of the humerus to the end of the stump (forearm)
Leg	Anterior superior iliac crest to the lateral maleolus
Thigh	Major trochanter to the lateral knee joint space
Lower leg	Lateral knee joint space to the lateral maleolus
Foot	Heel to the toes
Amputation stump	Ischial tuberosity to the stump end (thigh) Lateral knee joint space to the stump end (lower leg)

Body surface area

- Normograms equating height, surface area, and weight are shown in Fig. 9.11

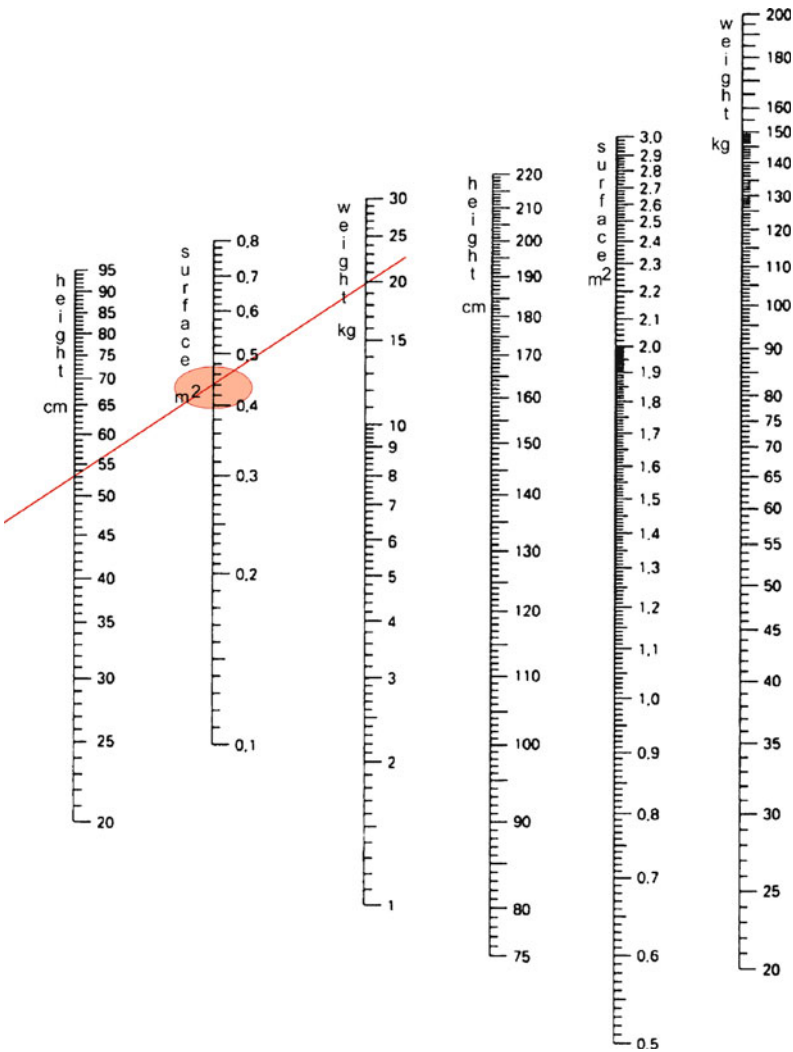


Fig. 9.11 Body surface area

10 Special Situations

10.1 The Polytraumatized Child

In the severely injured infant and child the following age-specific facts should be kept in mind:

- Larger head to body weight ratio (head often exposed to injury, neck injury)
- Larger body surface area to body volume ratio (hypothermia)
- More elastic thoracic wall (internal injury possible without external signs)
- Thinner abdominal wall where abdominal organs are below the rib cage (liver, spleen injury)
- Smaller total blood volume
- Narrow airways
- Long compensation of blood loss by tachycardia followed by rapid decompensation of circulatory status if left untreated

Definitions

- **Polytrauma**
  Two or more organ systems involved, where at least one injury or the combination of injuries is life-threatening
- **Multiple injury definition**
  Two or more injuries that affect the clinical course and treatment

Polytrauma protocol

- Every polytrauma protocol aims at fast evaluation and reconstitution of vital functions (treat first what kills first)
- While in general, Advanced Trauma Life Support (ATLS) guidelines (*ATLS Student Course Manual*, 7th edition., American College of Surgeon, Committee on Trauma, 2004) should be followed, local factors determine the composition of the trauma team and diagnostic pathways

Primary survey

Identification and simultaneous management of life-threatening conditions in the first few minutes, the next step (ABCDE) only starts when the previous step has been completed (Fig. 10.1):

- **A**irway maintenance with cervical spine protection
- **B**reathing and ventilation
- **C**irculation with hemorrhage control (see Table 10.1)
- **D**isability: neurological status (see Tables 10.2, 10.3)
- **E**xposure/**E**nvironmental control: completely undress the patient, but prevent hypothermia

Table 10.1 Blood volume loss evaluation

System	Mild blood loss (<30%)	Moderate blood loss (30%–45%)	Severe blood loss (>45%)
Cardiovascular			
▪ Heart rate	↑	↑↑	↑↑ then ↓
▪ Pulse peripheral	Weak, thready	Absent	Absent
▪ Pulse central	Normal	Weak, thready	Absent
▪ Blood pressure	Normal	Low normal	Hypotension
CNS	▪ Anxious ▪ Irritable ▪ Confused	▪ Lethargic ▪ Dulled pain response	▪ Comatose
Skin	▪ Mottled ▪ Cool ▪ Prolonged capillary refill	▪ Cyanotic ▪ Cool ▪ Markedly prolonged capillary refill	▪ Pale ▪ Cold
Urinary output	↓	Minimal	None

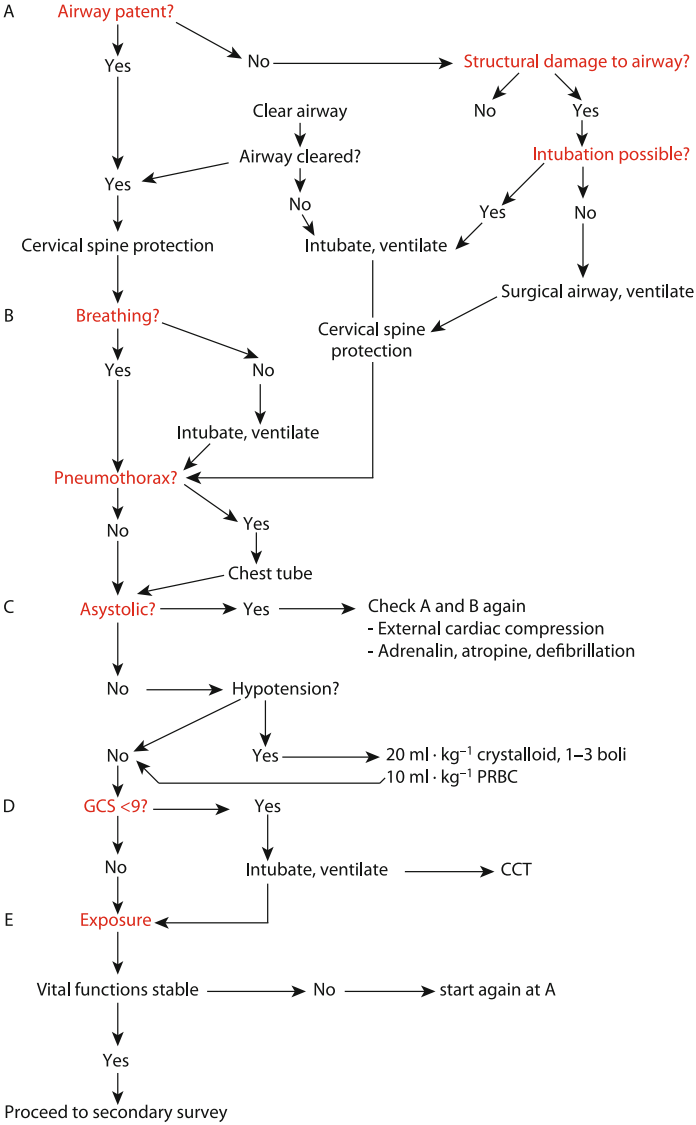


Fig. 10.1 Polytrauma algorithm (primary survey)

Table 10.2 Glasgow coma scale (GCS)

Parameter	Result	Score
Eye opening	Spontaneous	4
	▪ To voice	3
	▪ To pain	2
	▪ None	1
Verbal response child/adult	Oriented	5
	▪ Confused	4
	▪ Inappropriate words	3
	▪ Incomprehensible words	2
	▪ None	1
Verbal response infant	▪ Appropriate words or social smile	5
	▪ Fixes and follows	
	▪ Cries but consolable	4
	▪ Persistently irritable	3
	▪ Restless, agitated	2
	▪ None	1
Motor response	▪ Localizes pain	6
	▪ Obeys commands	5
	▪ Withdraws (pain)	4
	▪ Flexion (pain)	3
	▪ Extension (pain)	2
	▪ None	1

Table 10.3 Head injury (HI) severity according to GCS and presence of hemorrhagic shock

Injury type	GCS	Blood pressure (admission to hospital)	Mortality
Severe HI – shock	3–8	<5th age percentile	60%–70%
Severe HI	3–8	Normal	30%
Moderate HI	9–13	Normal	5%–10%
Mild HI	14–15	Normal	0%

Resuscitation

- Oxygenation and ventilation (if necessary orotracheal intubation with uncuffed tube, diameter approximates to the diameter of the child's little finger, infant: 40–60 breaths per minute, older child 20 breaths per minute, tidal volume 7–10 $ml \cdot kg^{-1}$)

- Pneumothorax decompression if indicated
- Placement of chest tube(s) if indicated
- Shock management and (two) IV lines
  - Peripheral vein including external jugular vein if accessible (two to three attempts)
  - Intraosseous device
  - Femoral or subclavian vein (percutaneous)
  - Saphenous vein (cut down at ankle)
- Warmed Ringer's lactate solution
  - One to three bolus infusions of 20 $ml \cdot kg^{-1}$
  - With third infusion consider giving 10 $ml \cdot kg^{-1}$ of type-specific or O-negative warmed packed red blood cells (pRBCs)
- Management of life-threatening problems identified in primary survey

Adjuncts to primary survey

- Monitoring (ECG, ventilatory rate, blood pressure, pulse oximetry, arterial blood gases; for normal parameters of physiologic function see Table 10.4)

Table 10.4 Vital functions: physiologic parameters in children of different ages

Age group (years)	Weight range (kg)	Heart rate (beats per minute)	Blood pressure (mmHg)	Respiratory rate (breaths per minute)	Urinary output ($ml \cdot kg^{-1} \cdot h^{-1}$)
Infant 0–1	0–10	<160	>60	<60	2.0
Toddler 1–3	10–14	<150	>70	<40	1.5
Preschool 3–5	14–18	<140	>75	<35	1.0
School age 6–12	18–36	<120	>80	<30	1.0
Adolescent >12	37–70	<100	>90	<30	0.5

- Urinary catheter (transurethral catheter contraindicated in suspected urethral injury)
- Gastric catheter (nasogastric tube contraindicated in suspected cribriform plate fracture)
- X-rays (chest, spine, pelvis), cervical CT (CCT) ± cervical spine CT, or spiral CT
- Abdominal ultrasonography (focused abdominal sonography for trauma, or FAST) rather than peritoneal lavage
- Draw blood [hemoglobin, glucose, electrolytes, partial thromboplastin time, aspartate aminotransferase (AST), alanine aminotransferase (ALT), lipase]

Secondary survey

- Head to toe evaluation, including reassessment of vital signs and complete neurologic examination
- Initiation of pain therapy and psychological support for child and parents
- Complete history (AMPLE)
  - **A**llergies
  - **M**edications currently used
  - **P**ast illnesses
  - **L**ast meal
  - **E**vents/**E**nvironment related to the injury
- Consider child abuse as a cause of major trauma especially in infants

Adjuncts to secondary survey

Examination-indicated specialized diagnostic procedures (after management of life-threatening injuries and after hemodynamic and ventilatory stabilization)

- CT scans
- Contrast X-rays
- Extremity X-rays
- Endoscopy
- Ultrasonography

Definitive care or transfer of patient
Evaluation of the institution's capabilities for definitive treatment and consultation of appropriate specialists. In cases where one or the other is lacking, early transfer of the patient to a different center is necessary.

10.2 Child Abuse

10.2.1 Epidemiology

Child abuse is frequent: about 3%–7% of children under 18 years suffer from child abuse in some manner. The incidence and prevalence depend on the development of a country's social service agencies and on the level of health professionals' awareness of the problem. The estimated number of unreported cases is high. Child abuse affects children of all socioeconomic, ethnic, and religious boundaries. There is no gender preponderance. It occurs at any age, but infants and toddler have a higher risk than older children.

10.2.2 Categories

Five main categories of child abuse are differentiated (Table 10.5). Each category requires its own individual approach to diagnosis and management but concomitant forms are common.

Table 10.5 Categories of child abuse

Category	Main characteristics
Physical abuse	Physical injuries (bruises, fractures, tissue disruption)
Sexual abuse	Contact sexual abuse (penetrating and non-penetrating injury) Non-contact sexual abuse (exhibitionism, voyeurism, sexual propositions or making of pornography)
Psychological abuse	Emotional deprivation, pathologic psychosocial environment, disturbed relationship between child and caregiver (psychosocial dwarfism, dissociative identity disorder)

Table 10.5 *(continued)* Categories of child abuse

Category	Main characteristics
Neglect (physical, educational, emotional, medical)	Failure to provide required age-appropriate care (shelter, food, clothing, education, affection, medical care)
Munchausen's Syndrome by Proxy	False history regarding a child who is not suffering from any of the reported symptoms, leading to unnecessary diagnostic procedures

10.2.3 Risk Factors for Child Abuse

Parental/caretaker risk factors

- History of abuse as children themselves
- Unrealistic developmental expectations of the child
- Young parents
- Single-parent home
- Character disorders (mental illness, depression, poor impulse control)
- Low self-esteem
- Addiction to drugs or other substances

High-risk child

- Prematurity
- Age (younger than 3 years)
- Physical disability (congenital anomalies, chronic illness)
- Developmental delay/disability
- Behavior problems (sleep or eating disorder, enuresis)
- Placement in foster care

Socioeconomic risk factors

- Poverty
- Unemployment
- Low education
- Social isolation
- Domestic violence

10.2.4 History

A thorough and complete medical history is mandatory, including present and past illness, review of systems, family medical history, and socioeconomic status.

Facts in the history that should alert to the possibility of child abuse

- Significant delay in seeking medical care
- No or insufficient explanation for the injury
- Changing history
- Injury and/or history incompatible with the development of the child
- History inconsistent with injury
- Doctor shopping

10.2.5 Physical Examination

It is of prior importance to examine the child carefully and completely. This permits assessment of the child's general well-being, appearance, behavior, development, nutritional status, and personal hygiene. Physical findings should be described in objective terminology and color photographic documentation should be used.

Physical findings compatible with child abuse

- Assault-like location (trunk, upper arms, upper legs, neck and face, perineal area)
- Obvious shape or pattern (marks of belts, hangers, hands, bite)
- Bruises in various stages of healing, indicative of repeated trauma
- Multiplanar injuries, such as bilateral or back and front together
- Most frequent forms of physical child abuse are listed in Table 10.6 and their signs are shown in Fig. 10.2

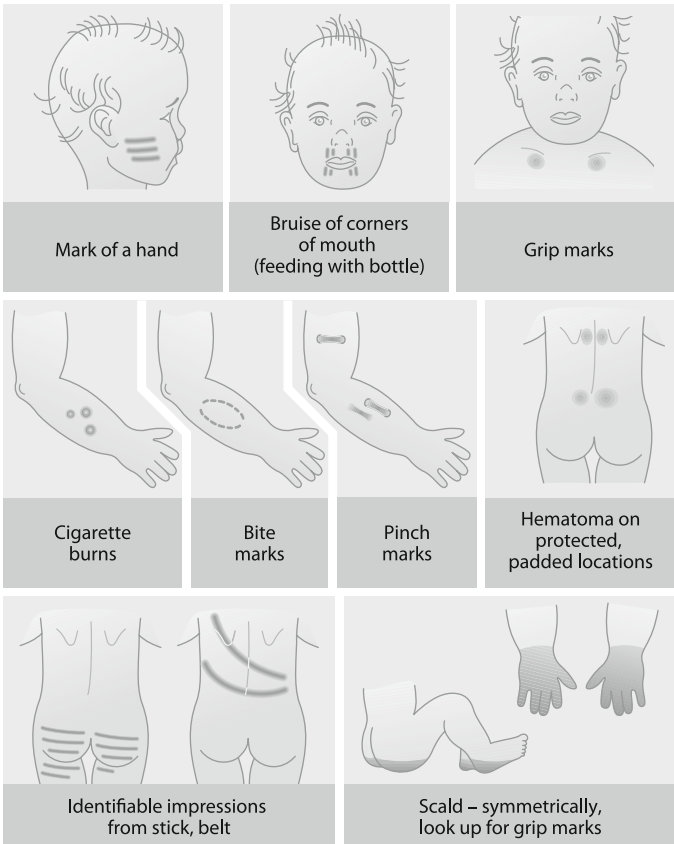


Fig. 10.2 Typical signs of child abuse

Table 10.6 *(continued)* Most frequent forms of physical child abuse

Injury	Characteristics
Bruises	▪ Very common presentation of child abuse ▪ Typical location (assault like) ▪ Almost always suspect in non-ambulating infant
Bite marks	Oval, tearing or crushing bite mark with a space between the canines of greater than 3 cm is likely to have be done by an adult human and indicates child abuse
Burns	▪ Contact injury: cigarette, hot iron (sharply defined pattern, deep) ▪ Scolds: hot liquids (often stocking or glove like) ▪ 10%–25% of burns in children are the result of abuse
Fractures	▪ Highly specific: metaphyseal fracture (bucket handle), posterior rib fractures ▪ Moderate specific: multiple (bilateral) fractures, fractures of different age ▪ Fractures occur in up to 30% of physically abused children, 75% of these fractures involve infants
Abdominal trauma	▪ Second most frequent cause of death in abused children ▪ Mostly young children (6 months to 3 years) ▪ Absence of visible external signs is common
Head injury: shaken baby syndrome	▪ Most frequent cause of death in abused children ▪ Mostly young children (usually younger than 2 years) ▪ Typical constellation of findings: retinal hemorrhages, intracranial trauma (subdural hemorrhage), diffuse axonal injury, cerebral edema, fractures of the ribs or metaphysis of long bones
Sexual abuse	▪ Girls are more frequently affected than boys ▪ Occurs at every age but mostly by adolescence ▪ The perpetrator is often a trusted family member or adult ▪ Lack of physical findings is very common

10.2.6 Differential Diagnosis

- Osteogenesis imperfecta
- Bleeding diatheses (idiopathic thrombocytopenic purpura, vitamin K deficiency, von Willebrand disease, Henoch–Schönlein purpura, hemophilia, leukemia)

- Accidental injuries
- Cutaneous mimickers (bullous impetigo, Mongolian spots, erythema multiforme minor)

10.2.7 Further Investigations

- According to the clinical presentation of the child abuse and to the differential diagnosis further investigations have to be considered

Laboratory studies

- Complete blood count including hematocrit
- Coagulation profile (prothrombin time, activated partial thromboplastin time, platelet count)
- Serology [liver transaminases, pancreatic amylase and lipase, calcium, phosphor, copper, ceruloplasmin, acylcarnitine, astroglial protein S-100, vitamin A, human immunodeficiency virus (HIV), syphilis]
- Urine analysis (hematuria, myoglobinuria, copper, organic amino acids)
- Stool guaiac
- Cerebrospinal fluid: blood, xanthochromia, neuron-specific enolase
- Gram stain of vaginal and/or anal discharge
- Genital, anal, and pharyngeal culture for gonorrhea
- Genital and anal culture for *Chlamydia*
- Wet prep of vaginal discharge for *Trichomonas vaginalis*
- Culture of lesions for herpes virus

Imaging studies

- Radiographic bone survey (two views of each long bone, of the skull, of the spine, chest, abdomen, pelvis, hands, and feet)
- CT scan of the head (rapid identification of CNS injury)
- CT scan of the abdomen
- MRI of the head (detecting and dating subdural hematoma and diffuse axonal injury)
- Radionuclide bone scan
- Photographs of skin lesions

Other examinations

- Fundoscopy by ophthalmologist
- Forensic evidence collection

10.2.8 Therapy and Management

- The medical care of the abused child depends on the injury and in serious cases may need reanimation and intensive care setting
- Hospitalization
- Refer to a child abuse specialist group for an interdisciplinary appreciation of the case. A medico legal examination can help to establish or corroborate the child abuse diagnosis
- Refer to child protection authorities
- According to legislation and depending on the severity of the child abuse, report to the appropriate law enforcement agency
- Protect siblings
- Offer psychological assistance to the abused child, the concerned family, and the perpetrator

Without appropriate management, the risk of recurrence of child abuse and sometimes escalating problems is estimated at 50%.

11 Birth Trauma

11.1 Skull

- Trauma to the skull can occur when the baby is born (Fig. 11.1, Table 11.1) and its treatment is outlined in Table 11.2

Table 11.1 General considerations of trauma to the skull at birth

Caput succedaneum	▪ Supraperiosteal edema ▪ Occasionally hemorrhage ▪ Secondary to compression during labor ▪ Crossing cranial sutures ▪ Disappears spontaneously after 1–2 days ▪ Subgaleal haemorrhage – spreads under galea but superficial to periosteum and blood tracks forward to periorbital fissues
Cephalhematoma	▪ Subperiosteal hematoma, not crossing the cranial sutures ▪ Most likely caused by repeated buffering of fetal skull against maternal pelvis or mechanical trauma (forceps/vacuum extractor) ▪ Most resolve spontaneously in 8–16 weeks ▪ Risk of super infection
Depressed fracture	▪ Depression of the soft bone like a table tennis ball

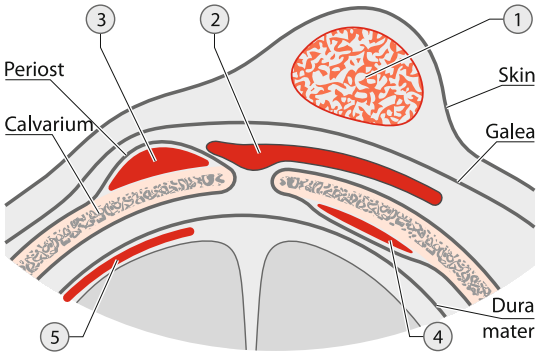


Fig. 11.1 Hematoma localization: *1* caput succedaneum, *2* subgaleal hemorrhage, *3* cephalhematoma, *4* epidural hemorrhage, *5* subdural hemorrhage

Table 11.2 Treatment of skull birth trauma

Caput succedaneum	No specific treatment required; if anemic, may need to give a blood transfusion
Cephalhematoma	No specific treatment required Aspiration contraindicated due to risk of introducing infection
Depressed fracture	Lifting of depression if depression is greater than the thickness of the calvarium

11.2 Extremities

- Trauma to the extremities can occur when the baby is born (Table 11.3) and its treatment is outlined in Table 11.4

Table 11.3 General considerations for trauma to the extremities at birth

Clavicle fracture	▪ Harmless fracture due to mechanical trauma during birth ▪ Sometimes iatrogenic ▪ Incidental diagnosis ▪ Crepitation or swelling likely
Humerus fracture	▪ Rare ▪ Caused by liberation of the arm ▪ Clinically swelling ▪ Malposition ▪ Relieving posture
Facial palsy	▪ Complication after forceps delivery ▪ Mouth is drawn to the normal side while crying ▪ Sometimes failure of eye closure (Bell's sign)
Brachial plexus paralysis	▪ Result from a difficult assisted delivery, e.g., shoulder dystocia, flaccid arm, hand in porter's tip posture (Erb's palsy)
Torticollis	▪ Fibrosis of the sternocleidomastoid muscle after trauma and hematoma (tumor) ▪ Tilted head ▪ Turning the chin to the non-affected side ▪ Only a small percentage of sternocleidomastoid tumors cause torticollis

Table 11.4 Treatment of birth trauma to the extremities

Clavicle fracture	Good prognosis, no treatment required, quick spontaneous healing with callus formation
Humerus fracture	Splinting to avoid malposition
Facial palsy	Spontaneous recovery; if failure of eye closure, protect eyes from drying
Brachial plexus paralysis	Physiotherapy, if not resolved by 6 months poor outcome
Torticollis	Physiotherapy, good prognosis

12 Skull

12.1 Congenital Osteocutaneous Defect

General considerations

- Regional defect (aplasia) of skin and bones, with an area >2–3 cm^2

Therapy

- Cover the defect with skin flaps

12.2 Encephalocele

General considerations

- Congenital herniation of the brain and the meninges through an opening in the midline of the skull
  - Approximately 60% are occipital
  - Frequently with involvement of the cerebellum
  - Incidence: approximately 10% of neural tube defects
- Encephaloceles are commonly covered with hairy thin skin, which may be pigmented
- Flammeus nevus or angiomatous changes are not rare
- Perforation danger in bulging encephaloceles
- Pulsatile mass that increases in size with crying is a sign of encephalocele
- May be identified antenatally by ultrasonography
- Differential diagnosis
  - Dermoid cyst
  - Hemangioma

- Nasal polyp (diagnosis may be difficult in anterior lesions)
- Teratoma
- Meningioma
- Neurofibroma

Preoperative work-up

- Ultrasonography and MRI (regarding brain deformity) → specification of prolapsed parts of brain
- X-ray → size and location of cranial defect

Operation

- Timing depends on:
  - Condition of the covering skin
  - Pressure
- Aims of the procedure:
  - Restoration of the brain to the cranium
  - Excision of extracranial non-functioning brain tissue
  - Closure of the dura
  - Coverage with periosteal flap
  - Subcutaneous and skin closure
- Simultaneous or subsequent therapy of hydrocephalus, if necessary

Complications

- Meningitis (more common with anterior encephalocele, connection to nose or pharynx)
- Cerebrospinal fluid (CSF) fistula

Prognosis

- Mortality of 20% by 1 year
- Approximately 30% of the survivors expect normal intelligence
- Approximately 30% of the patients with an occipital encephalocele are partially or completely blind
- Convulsions in 25%

12.3 Hydrocephalus

General considerations

- Presence of an excessive amount of cerebrospinal fluid (CSF) with ventricular enlargement, usually under increased pressure, due to an imbalance between production and absorption of CSF
- CSF mainly produced in choroid plexus of the lateral ventricles, a smaller amount originates from the extracellular fluid
- Drainage via the third ventricle, aqueduct of Sylvius, the fourth ventricle, and the spinal central canal
- The fluid escapes into the subarachnoid space via the foramina of Luschka and Magendie
- Absorption through arachnoid granulations and the sagittal sinus
- Normal pressure 5–15 cmH_2O
- Overall incidence 2 per 1000 births, excluding hydrocephalus associated with myelomeningocele, 0.4–1 per 1000 births
- Differential diagnosis
  - Chronic subdural hematoma (hygroma)
  - Macrocephaly/megalocephaly
  - Atrophic encephalopathy
  - Ventricular hemorrhage

Classification

- Communicating hydrocephalus
  - Free flow of CSF between the ventricles and the subarachnoid space
  - Malfunctioning of absorption after infection or hemorrhage
- Non-communicating hydrocephalus
  - Obstruction of CSF drainage in the area of the fourth ventricle caused by:
    - Neoplasm
    - Hemorrhage
    - Infection
    - Dandy–Walker malformation

  - Arnold–Chiari malformation (myelomeningocele)
  - Aqueductal stenosis
- Choroid plexus papilloma with excessive secretion – rare

Signs

- Rising pressure, which can be compensated by the gaping fontanels and the sutures in the neonatal period
- Enlargement of the neurocranium with "sun-set" appearance (superior sclera visible above the iris) due to pressure on the soft supraorbital plates in neonates
- Signs of raised intracranial pressure in the case of closed fontanels
  - Vomiting
  - Seizures
  - Opisthotonus
  - Decreasing level of consciousness
  - Visual disturbance
  - Hypertension
  - Bradypnea
  - Bradycardia
  - Mydriasis
  - Papilledema

Preoperative work-up

- Measure the head circumference and documentation on a centile chart
- Ultrasonography, if fontanel is still open
- Skull X-ray (osteoporotic changes, splaying of suture lines)
- MRI, in special cases CT (tumors, cysts, enlargement of ventricle, hemorrhage)
- Ophthalmoscopy (papilledema)
- Measurement of CSF pressure (Fig. 12.1)
- Scintigraphy scanning for measurement of CSF absorption (rarely needed)

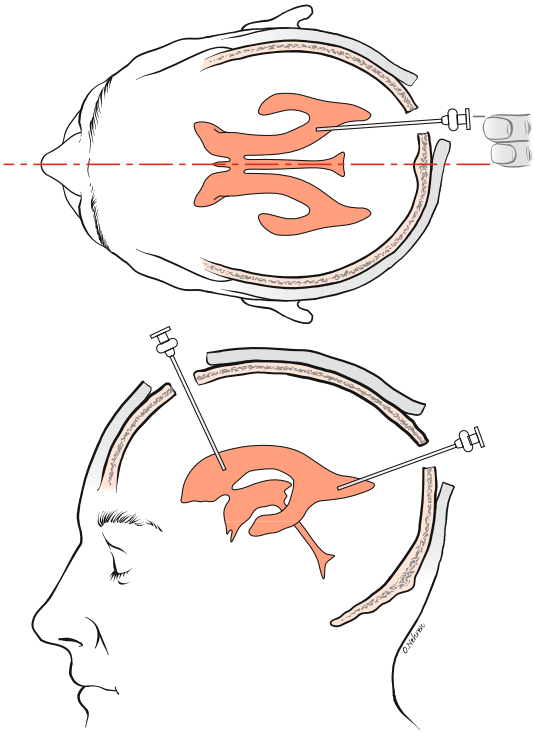


Fig. 12.1 Puncture of the ventricle system

Indications for operation

- Increasing ventricle volume with ballooning of lateral ventricle
- Increasing head circumference >97th centile
- CSF pressure >25 cmH_2O

Operation

- Like other prosthetic insertions, CSF shunts are prone to infections with common microbes from resident skin flora. The initial painting with antiseptics abolishes all vegetative organisms, but during the procedure further baceteria emanate from the pilosebaceous unit. These can infect a shunt system. A system of antisepsis has been shown to reduce infection rates with the offending *Staphylococcus epidermidis* to low levels

- The CSF is drained into the peritoneal cavity (ventriculo-peritoneal shunt = VP shunt) or occasionally the atrium of the heart is used (VA shunt)
- There are many types of shunts, e.g., Silastic slit valves, Holter, Pudenz–Heyer, Hakim-ruby-ball-valve (Codman–Hakim®)
- Ventriculostomy of the third ventricle
- Perioperative antibiotic prophylaxis to prevent wound infection
- Figure 12.2 indicates the surgical steps to treat hydrocephalus

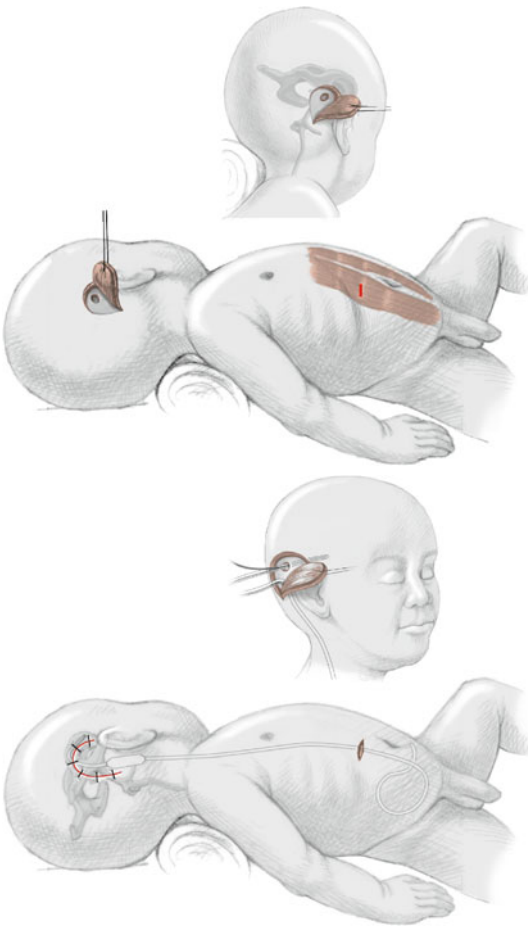


Fig. 12.2 Surgical steps in the treatment of hydrocephalus

Postoperative care

- C-reactive protein (CRP) estimation 6 weeks post surgery to pick up colonized shunts early
- Education of parents about signs of blocked/disconnected shunt, infection and over draining
- Life-time check-ups are required, but especially in the growing period

Complications

- Infection (peritonitis, ventricular sepsis), which can be reduced by taking great care during the insertion of the shunt (aseptic technique)
- Catheter blockage, disconnection, and breakage leading to signs of raised intracranial pressure
- Over-drainage (slit ventricle syndrome), subdural hematoma, craniosynostosis
- Catheter shortening due to patient growth
- Thrombosis of superior vena cava (VA shunt)
- Abdominal liquor cyst

Prognosis

- This depends on the primary situation, and is influenced by morbidity due to catheter-related complications

12.4 Craniosynostosis

General considerations

- The development of the cranium takes place along the cranial sutures, which close at different ages
- Due to abnormal and early cranial suture closure the skull becomes deformed and the intracranial pressure may rise

Classification

- Craniosynostosis may result in an increase of intracranial pressure when premature union of both coronal and sagittal sutures are involved

- Craniosynostosis should be differentiated from plagiocephaly, which is a condition in which the skull has an abnormal shape not always related to premature synostosis of the sutures but to the position of the head in utero
- Typical congenital craniofacial syndromes are Crouzon syndrome with pansynostosis and Apert syndrome with synostosis of the coronal sutures

Types

- Types of craniosynostosis are listed in Table 12.1

Table 12.1 Types of craniosynostosis

Form	Suture	Characteristics
Scaphocephaly	Sagittal	Elongated head
Brachycephaly	Coronal	Flattened anterior/posterior
Turricephaly	Coronal and sagittal	Tower shaped skull
Trigonocephaly	Frontal	Keel-shaped forehead

Signs

- Striking appearance
- Exophthalmia
- Developmental disorders

Preoperative work-up

- Clinical examination of the skull from the front, back, and always from above
- General neurological status
- X-ray a.p. and lateral
- CT
- Ultrasonography

Indications for operation

- Increase of intracranial pressure
- Cosmetic considerations

Operation

- According to the skull deformation
  - Release craniectomy or cranioplasty

Prognosis

- Very good aesthetic results if early surgery
- Good functional results depending on the severity and the time

12.5 Brain Injuries

General considerations

- Functional impairment of the brain after violent damage
- Anatomical specialties of the child's cranium
  - Open fontanels
  - Wide venous communication between the dura and the skull
  - Possibility of sub- and epidural hematoma without fracture
  - Meningeal arteries are not yet bone covered in a channel
  - Development of brain edema
- Rise of intracranial pressure due to:
  - Intracranial hemorrhage
  - Posttraumatic brain edema
- Skull fractures
  - Linear fissures
  - Non-dislocated linear fractures
  - Depressed fractures
  - Basal skull fractures
- Intracranial hemorrhage
  - Epidural hematoma
  - Subdural hematoma
  - Subarachnoid hematoma
  - Brain hemorrhage
- Always think of a battered child when you see a child with a brain injury

Classification

- Open skull injury: discharge of liquor, meninges or brain tissue
- Closed skull injury: injury of the brain and/or meninges without opening of the skull

Signs of fractures and hematomas

- Table 12.2 lists the signs of fractures and hematomas

Table 12.2 Signs of fractures and hematomas

Linear fissures	Slight signs according to degree of brain involvement
Linear fractures	Slight signs according to degree of brain involvement
Depressed fractures	Focal neurologic signs, signs of increased intracranial pressure
Basal skull fractures	Monocle hematoma, tympanum hematoma, blood and liquor, discharge out of nose, ear or throat
Epidural hematoma	Lucid interval, then increasing unconsciousness till coma, mydriasis on the side of the hematoma, contralateral paresis, positive Babinski, increased reflexes, irregular breathing
Subdural hematoma	Less dramatic than epidural hematoma with longer symptom-free interval (up to 3 days)
Subarachnoid hematoma	Acute headache, unconsciousness, coma

Evaluation

- Advanced Pediatric Life Support (APLS) assessment >11 points (Table 12.3): good prognosis
- APLS <8 points: intubation and respiration

Table 12.3 Glasgow Coma Scale (adapted as Children's Coma Scale <4 years)

Points	Check/condition
Best motor response (6 grades)	
6	Carrying out request (obeying command): the child moves spontaneously or to your request
5	Localizing response to pain: put gentle pressure on the patient's fingernail bed with a pencil then try supraorbital and sternal pressure: purposeful movements towards changing painful stimuli is a localizing response
4	Withdrawal to pain: pulls limb away from painful stimulus
3	Flexor response to pain: pressure on the nail bed causes abnormal flexion of limbs, decorticate posture
2	Extensor posturing to pain: the stimulus causes limb extension (adduction, internal rotation of shoulder, pronation of forearm), decerebrate posture
1	No response to pain
Best verbal response (5 grades) adapted for children <4 years	
5	Fixes and follows objects, is orientated to sounds, smiles, may cry but interacts
4	Crying, inappropriate fixation, irritable
3	Cries only to pain
2	Inconsolable, irritable, moans to pain
1	No response
Eye opening (4 grades)	
4	Spontaneous eye opening
3	Eye opening in response to speech: any speech, or shout, not necessarily request to open eyes
2	Eye opening in response to pain: pain to limbs as above
1	No eye opening to pain

Investigations

- Exclude other injuries
- Exclude skull fractures
- Neurological status
- Intracranial pressure signs – check-up

- Ultrasonography, if fontanel is still open
- X-ray
- CT according to clinical history and condition
- Consider repeat CT after a few hours or next day

Therapy for fractures and hematomas

- Treatment for fractures and hematomas is outlined in Table 12.4

Table 12.4 Treatment for fractures and hematomas

Cerebral concussion	Close observation, give parents head injury leaflet informing them on what they need to pay attention to or observe
Cerebral contusion	Admission and close observation
Cerebral compression	Admission and intracranial pressure prophylaxis
Linear fissures	Conservative management
Linear fractures	Usually conservative
Depressed fractures	If depression > thickness of calvarium and signs: operative
Basal skull fractures	Usually conservative
Epidural hematoma	Trepanation and operative removal
Subdural hematoma	Trepanation and early operative removal

General therapeutic concepts

Acute phase

- Chest up 15°–20°
- Normothermy
- Control of respiration; intubation and ventilation with slight hyperventilation
- Sufficient infusions to stabilize circulation and correct electrolyte imbalance
- Monitoring (ECG, pulse oximetry, blood gas analysis, blood tests)
- Treat shock with urine output monitoring
- Continuous re-check of Glasgow Coma Scale score
- Regular check of pupil status

- Temperature check (consider central hyperthermia)
- Nasogastric tube for aspiration prophylaxis and early high caloric nutrition – not with basal fracture
- Measurement of intracranial pressure if necessary
- Rehabilitation

Prognosis

- Very good for mild cerebral injuries
- Morbidity (e.g., epilepsy)

12.6 Skull Fractures

General considerations

- Of the skull fractures in childhood, 75% are linear temporoparietal fractures
- These fractures heal within 1–2 months without any residual complications
- The following must be excluded:
  - An epidural or other intracranial bleeding
  - A depression fracture
  - A leptomeningeal cyst (developing fracture)
    - When the dura ruptures and the meninges become interpositioned, and the fracture develops rather than heals

Signs

The following signs are not always present and may be signs of brain injury:

- Decreasing consciousness
- Headache
- Vomiting (may be a sign of high intracranial pressure)
- Bleeding or CFS from the ear is a sign of skull base fracture
- Liquorrhea from the nose

Investigations

- Careful clinical evaluation including Glasgow Coma Score
- Examination of liquid coming from the nose (liquor from a fronto-basilary fracture)
- Otoscopy
- Radiology and CT as indicated

Therapy

- Observation (pulse, blood pressure, pupillary reflex, etc.)
- Specific therapy needed depending on situation
- Operative therapy of depressed fracture and often of intracranial haemorrhage

Prognosis

- Simple fractures: very good
- According to severity

Prevention

- It is recommended that all females be commenced on folic acid before pregnancy which drops the risk of hydrocephalus significantly
- In some countries pregnant women are screened for α-fetoprotein in the serum and amniotic fluid. The affected fetus is then aborted with parental agreement

13 Spinal Column

13.1 Spina Bifida/Meningocele/Myelomeningocele

General considerations

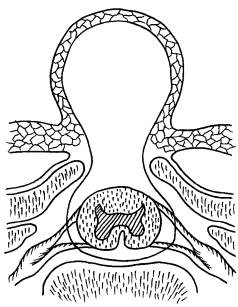
- Rachischisis or spina bifida is a developmental anomaly characterized by defective closure of the vertebral arch, through which the spinal cord may protrude
- Incidence of spina bifida cystica is 1–2 per 1000 live births with big geographic variations: higher risk in Caucasians and in lower socioeconomic groups
- Etiology still unknown; there is a relationship between spina bifida and genetics, folic acid metabolism, and antiepileptic agents
- Mostly lumbosacral
- Often hypertrichosis, hemangioma or other hamartomata overlying spina bifida occulta
- Differential diagnosis: teratoma
- Transfer to a center in the lateral decubitus position with the lesion covered in sterile saline-soaked swabs with clear plastic to prevent heat loss

Classification

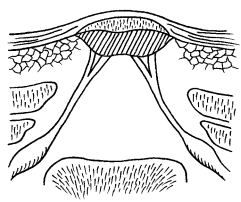
- Classification of spina bifida, meningocele and myelomeningocele is given in Table 13.1
- Meningocele and myelomeningocele are drawn in Fig. 13.1

Table 13.1. Classification of spina bifida, meningocele and myelomeningocele

Cleavage of			
the spine	= Spina bifida occulta		
the spine +	dura mater	= Meningocele	
the spine +	dura mater +	spinal cord	= Myelomeningocele



Meningocele



Myelomeningocele

Fig. 13.1 Meningocele and myelomeningocele

Signs

- Signs of spina bifida, meningocele and myelomeningocele are given in Table 13.2

Table 13.2. Signs of spina bifida, meningocele and myelomeningocele

Spina bifida occulta	Usually without signs, occasionally neurological defects caused by tethering of the cord
Meningocele	Usually not associated with neurological defects or hydrocephalus
Myelomeningocele	▪ Flaccid paralysis depending on the location of the defect; bladder and bowel involvement (incontinence, constipation, rectal prolapse) ▪ Skeletal abnormalities ▪ Sensory loss ▪ Hydrocephalus 80%–90%

Preoperative work-up

- Neurological and ophthalmological status (e.g., sensory levels, muscle charting, fundoscopy)
- Orthopedic status
- Urological status (e.g., renal ultrasonography, urodynamics)
- Cranial ultrasonography
- MRI, CT in special cases

Indication for operation

- Indication depends on the individual anatomical situation and other congenital anomalies
- Alternative is conservative management
- Parents should be informed and involved early in the decision-making process

Operation

- Meningocele
  - For cosmetic reasons
- Myelomeningocele
  - Operation, if possible at day 1 or within the first 72 h of life
  - Gentle replacement of the neural plaque, closure of the dura and the skin in layers
  - Incision of the skin at the border of the defect
  - Preparation of the borders of the neural plaque (skin rests have to be removed)
  - Dissection of the dura at the border to the skin
  - Tubularization of the dural layer with a running suture over the neural plaque
  - Muscle and skin closure
- The surgical treatment of spina bifida is illustrated in Fig. 13.2

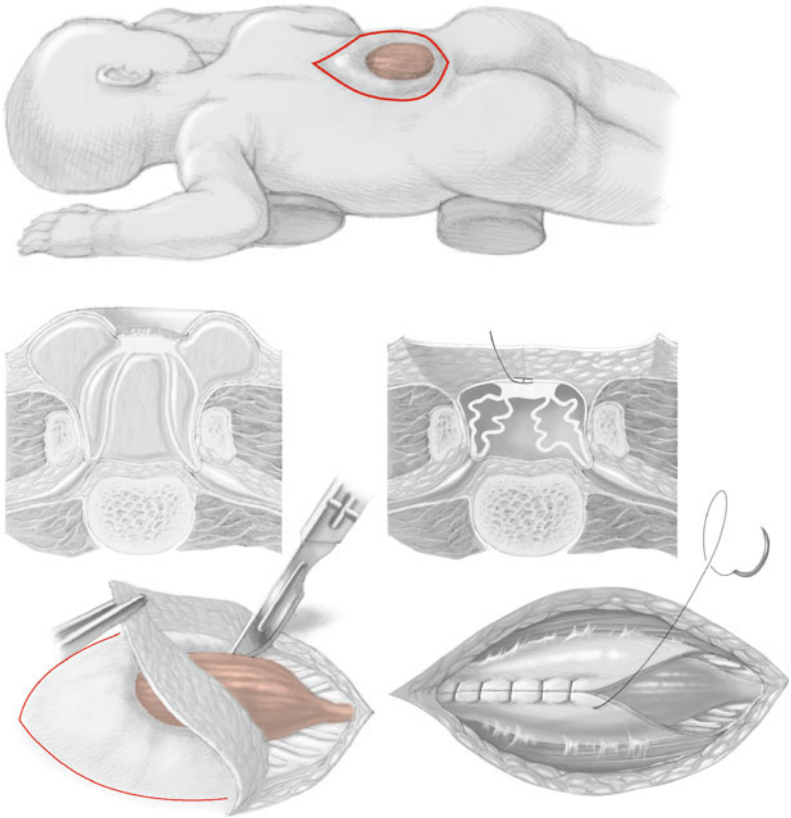


Fig. 13.2 Surgical treatment for spina bifida

Postoperative care

- Close follow-up
- Measurement of the head circumference and observance of intracranial pressure, possibly early shunting
- Urological investigations
- Orthopedic follow-up

Complications

- Wound infection
- Wound dehiscence
- CSF fistula

Prognosis

- Depends on the lesion itself and on optimum interdisciplinary cooperation between many specialties, i.e., pediatrics, disability specialists, pediatric surgeons and urologists, neurosurgeons, orthopedics, ophthalmologist, paramedical personnel, and the parents

14 Face

14.1 Oral Cavity

14.1.1 Tongue Tie

General considerations

- The frenulum of the tongue is relatively fibrotic and short and may restrict movement of the tongue tip – this is usually simply a cosmetic concern
- Displacement of the tongue tip over the lower teeth and upwards is restricted
- May impair sucking and feeding in the young infant
- Concern is raised about possible interference with articulation and speech sounds
- Often, only reassurance is needed, as the tongue tie releases with growth and tooth development

Operation

- Simple division of the anterior fibrous edge of the frenulum is sufficient
- Could be performed as a single snip without anesthesia, but a general anesthetic is preferable
- The frenulum can be clamped with a fine instrument for some seconds before cutting
- Bleeding occurs only occasionally, in which case a single suture is required
- Care should be taken to avoid injuring the frenular artery, which is usually well posterior

14.1.2 Gum Lesions

- Epulis
- Dentigerous cyst
- Alveolar abscess
- Eosinophilic granuloma

General considerations

- All appear as lesions on the gum margin
- X-ray helpful to exclude significant bony lesions

Operation

- Local excision preserving mucosa to cover defect
- Special care should be taken to avoid damaging the tooth buds
- Wide excisions are unnecessary as these lesions are benign

14.1.3 Ranula

General considerations

- Cystic swelling from sublingual glands
- Superficial, smooth, bluish, cystic lesion beneath tongue (ranula = frog)
- Related to multiple duct openings of the sublingual salivary gland
- Important to differentiate from lymphangioma, which is usually deeper

Operation

- Nasotracheal intubation preferable
- Packing in hypopharynx to avoid contamination
- Complete excision may be performed for smaller lesions
- Larger lesions treated by marsupialization – partial excision and cyst wall suture

14.1.4 Macroglossia

General considerations

- A large tongue may be true macroglossia or due to hypertrophy or infiltration with lesions such as hemangioma, lymphangioma or neurofibroma
- May cause respiratory obstruction, requiring relief with oral or nasal airway maintenance
- Protrusion from the mouth may lead to ulceration, infection, and hemorrhage
- Persistent macroglossia may interfere with maxillary, mandibular, and dental development
- Careful hygiene necessary

Operation

- Establishing a safe airway for anesthesia may be a challenge
- Tracheostomy may have to be considered
- Tongue reduction by extirpation or partial excision depending on the lesion

14.2 Face

14.2.1 Preauricular Defects

General considerations

- Skin tags and sinuses occur in the immediately preauricular position
- Skin tags and sinuses occur just anterior to the tragus of the ear
- Anomalies of first branchial arch development
- Skin tags may contain a cartilage rudiment
- Sinuses can extend down to the temporomandibular joint (TMJ)

Operation

- A small vertical skin ellipse is taken enclosing the tag or sinus
- Cartilage rudiment should be traced down and excised
- Sinuses should be completely excised down to the TMJ
- An infected or recurrent sinus may have complex ramifications and requires wider excision using a raised skin flap

14.2.2 Prominent Ears

General considerations

- Prominent (bat) ears may be uni- or bilateral
- Usually loss of superior anthelical fold and protrusion from the side of the head at an angle of greater than 30°
- Sometimes larger than normal and occasionally excessively vascular

Preoperative work-up

- Photographic documentation is advisable

Operation

- Operation best in children more than 5 years old, as younger children may be non-compliant with aftercare
- Local nerve block is a useful adjunct to general anesthesia
- Careful marking of incisions and proposed fold formation is important
- A postauricular elliptical incision exposes the conchal cartilage, which is retrocurved in the line of the superior anthelical fold by scoring, resection or suture plication after abrading the cartilage

Postoperative care

- A firm head bandage is used for compression, minimizing hematoma formation and possible infection. It also protects the ears and retains their position for up to 10 days

14.2.3 Dermoid Cysts

General considerations

- Caused by sequestration of epidermal/dermal cells beneath the skin surface usually at a line of embryonic fusion. The most frequent site is at the external angle of the eyebrow, but they can be found in the midline of the nose, palate, and orbit
- Firm, smooth, yellowish swelling about 1 cm in diameter at the lateral end of the eyebrow
- Usually mobile in the subcutaneous plane but occasionally fixed to bone
- Nasal dermoid may be associated with a small midline nasal sinus – the possibility of extension deep into the anterior cranial fossa should always be checked by X-ray
- Large nasal dermoids raise suspicion of an anterior encephalocele – CT scan needed

Preoperative work-up

- Ultrasonography to determine size and infiltration of bone
- X-ray if the ultrasonography indicates that the cyst is destroying bone
- CT scan advisable if the cyst is fixed, to exclude an intracranial (dumb-bell) extension through the frontozygomatic suture

Operation

- Short incision in the shaved lateral eyebrow line in the plane of the hair shafts
- A midline nasal biconvex incision is used to incorporate a nasal sinus and dermoid
- The cyst should be mobilized and removed intact to avoid leaving skin cells for possible recurrence

14.2.4 Choanal Atresia

General considerations

- Choanal atresia is a rare problem of bony or membranous obstruction of one or both choanae (nasal passages)
- Bilateral atresias can lead to severe respiratory distress
- Unilateral atresia can remain unidentified for long periods

Signs

- Presentation is usually immediately postnatal with respiratory obstruction in bilateral cases
- The child oxygenates well when crying with their mouth open, but becomes cyanotic after closing their mouth
- May present as part of a complex condition (e.g., CHARGE association, i.e., **c**oloboma, **h**eart defects, choanal **a**tresia, **r**etarded growth, **g**enitourinary abnormalities, and **e**ar anomalies)

Immediate management

- An oral airway tube (McGill) encourages the newborn, who is often an obligate nose-breather, to breathe through the patent oral airway
- Artificial ventilation may be needed if respiratory problems persist

Preoperative work-up

- Nasogastric tube in both nostrils to check the passage
- X-ray with contrast of the nostrils
- CT

Operation

- Perforation of membrane (transnasally) or resection of bone (transpalatal approach)
- Insertion of tubes through both choanae (or U-tube with posterior window) to maintain patency for 3–4 weeks

14.2.5 Cleft Lip and Palate

Cleft lip and palate is one of the most frequent major congenital anomalies, occurring in 1:600 newborns. In recent years they have sometimes been recognized on prenatal scans. Early and well-informed counseling is an important start to the comprehensive, multidisciplinary plan of management, which will often extend throughout childhood into early adult life. Clefting may be an isolated abnormality but up to 50% of cases are also associated with other anomalies expressing a multitude of syndromes.

Classification

Clefts of the primary palate involve the lip and alveolus whereas secondary palate clefts involve the hard and soft palates. Clefts can be incomplete or complete, unilateral or bilateral. Left-sided clefts are more frequent than right-sided ones and boys are more often affected than girls. Clefts of the palate alone constitute up to 50% of all clefts and may be associated with lower jaw hypoplasia.

Immediate care and investigation

- Respiratory support in Pierre Robin syndrome
- Feeding advice and supplementation – modified teats and feeding bottles
- Consideration of genetic assessment
- Good quality standardized photographs for planning and audit purposes
- Assessment by plain X-ray, CT or MRI scan when craniofacial anomaly suspected
- Orthodontic management to improve alveolar alignment if necessary

Operation

A schedule of operations is usually outlined at the outset of treatment and may consist of:

- Lip repair and primary nasal correction at about 3 months
- Hard and soft palate repair at about 6 months
- Pharyngoplasty, if needed for speech, at 4/5 years
- Alveolar bone grafting at time of eruption of second dentition (9/10 years)
- Secondary surgery (cosmetic, orthognathic) in adolescence
- Unilateral cleft lip repair is illustrated in Fig. 14.1
- Bilateral cleft lip repair is illustrated in Fig. 14.2
- Cleft palate repair is illustrated in Fig. 14.3

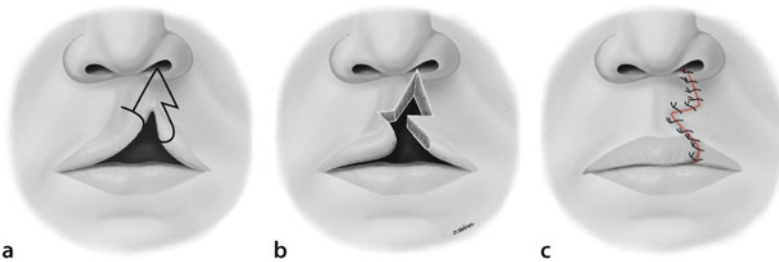


Fig. 14.1a–c Surgical repair of unilateral cleft lip

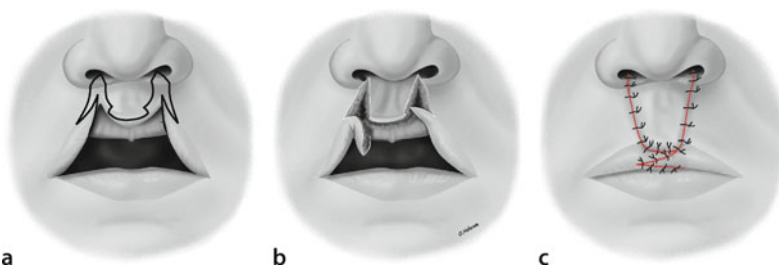


Fig. 14.2a–c Surgical repair of bilateral cleft lip

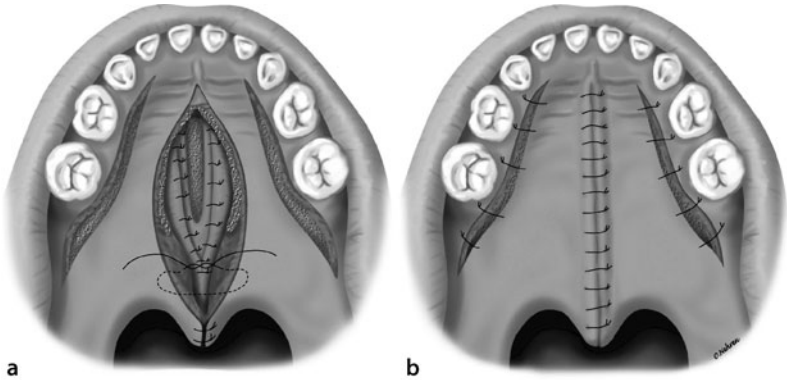


Fig. 14.3a,b Surgical repair of cleft palate

Further management

- Ongoing management throughout childhood in the context of a multidisciplinary clinic is essential in order to achieve optimal outcomes. The professionals involved are: cleft surgeon, orthodontist, speech therapist, specialist cleft nurse, geneticist, audiometrist, otorhinolaryngologist, pediatric dentist, and psychologist

14.2.6 Facial Fractures

General considerations

- The facial skeleton is susceptible to fractures giving rise to characteristic signs and symptoms

Classification

- Facial fractures are classified using the Le Fort system (Fig. 14.4, Table 14.1)

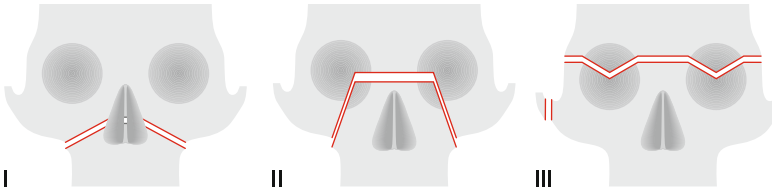


Fig. 14.4 Le Fort fracture classification

Table 14.1 Le Fort fracture classification

Grade	Fracture
I	Horizontal fracture through alveolar process of maxilla
II	Pyramidal maxillary fracture
III	Oblique fracture through lateral orbitae

Signs

- Signs of facial fracture relate to the site of the fracture (Table 14.2)

Table 14.2 Signs of facial fracture

Signs	Fracture
Bleeding from mouth Jaw asymmetry and displacement	Maxillary Mandibular
Damage/dislocation of teeth Bleeding/CSF from nose	Mandibular Maxillary Nasal bone Ethmoid plate fractures
Bleeding/CSF from ear	Temporal mastoid bone
Bruising and swelling around mastoid Facial nerve paralysis	Temporal Mastoid
Periorbital hematoma unilateral and bilateral Diplopia	Orbita

Preoperative work-up

- Detailed facial X-rays
- Chest X-rays for dislocated teeth, bone fragments
- CT scan needed to define fracture pattern

Conservative therapy

- Establish adequate airway patency
- Treatment of possible infection with antibiotics
- Treatment of possible aspiration of blood and/or teeth

15 Neck

15.1 Median and Lateral Neck Masses

The differential diagnoses of median and lateral neck masses are shown in Table 15.1.

Table 15.1 The differential diagnoses of median and lateral neck masses

Lateral	Median
Lymph node enlargement (inflammatory, systemic, neoplastic)	Submental lymph node
Neurogenic tumor (neuroblastoma)	Dermoid
Vascular tumor	Ectopic salivary tissue
Parotid/salivary gland	Thyroglossal cyst
Laryngocele	Ectopic thyroid tissue
Branchial cyst	Thyroid swelling
Thyroid swelling	Ectopic thymus
Mesenchymal tumor	
Teratoma	
Lymphangioma (cystic hygroma)	

15.2 Congenital Malformations

15.2.1 Thyroglossal Cysts and Fistulas

General considerations

- Thyroglossal fistulas/cysts are found along the line of the embryological descent of the thyroid gland in the neck from its site of origin at the foramen cecum of the tongue to the hyoid bone
- They rarely extend beyond the hyoid bone
- Smooth, firm, rounded swellings sitting in the midline over the hyoid bone
- Occasionally extend inferiorly
- If infected, they become enlarged and inflamed and may discharge
- Differentiation from midline dermoid cyst, lymph gland or aberrant thyroid is required
- A thyroid scan or thyroid hormone analysis is rarely needed

Signs

- Usually asymptomatic but patients wish resection for cosmetic reasons
- Become painful and tender if infected
- Move superiorly on extrusion of tongue

Preoperative work-up

- Antibiotic cover (against oral flora) is advised

Operation (Sistrunk's procedure)

- Transverse (collar) incision over the hyoid bone
- A skin ellipse is incorporated if the cyst is noticeably adherent to skin
- The central segment of the hyoid is removed with the cyst by dividing the hyoid using a bone-cutter
- The tract of the fistula is excised with a midline strip of hyoglossus muscle up to the foramen cecum
- The dissection is made easier if a surgical assistant places a gloved finger into the patient's mouth
- The operation is illustrated in Fig. 15.1

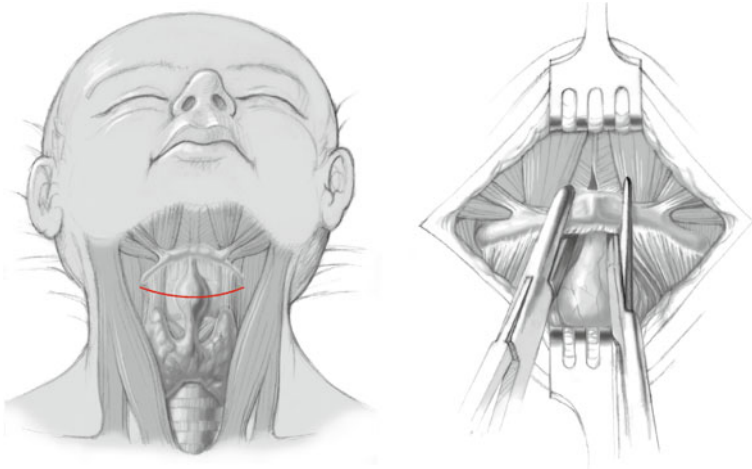


Fig. 15.1 Operative steps: thyroglossal cyst

Complications

- Bleeding/hematoma formation may lead to swelling and upper airway obstruction
- Infection is a risk when the cyst is initially infected or when the fistula tract is opened
- Recurrence rates are low (5%) when the fistula is excised, but are higher (20%) for cyst removal alone

15.2.2 Branchial Arch/Cleft Remnants

General considerations

- Branchial remnants are almost always related to the second arch/cleft
- They are located along the anterior edge of the sternocleidomastoid muscle around its midpoint
- They may extend from the tonsillar fossa of the pharynx to the skin surface

- Other arch remnants are very rare
- First arch remnants occasionally present as cysts and fistulas at the angle of the jaw and around the external auditory meatus
- Third and fourth arch remnants arise at the level of the thyroid

Signs

- May appear as a smooth, asymptomatic swelling at the anterior sternomastoid border
- Infection causes inflammation and tenderness and may lead to suppuration, abscess formation, and discharge
- A fistula or sinus presents as a small skin point with viscid mucous discharge
- In atypical cases, differentiation from other cervical masses may be aided by ultrasonography or CT scanning

Preoperative work-up

- Palpation of the cyst from outside and from the pharyngeal opening with the finger in the mouth
- Ultrasonography
- CT for third and fourth remnants
- Antibiotic cover is needed if there is any evidence of infection

Operation

- A probe may be inserted into a sinus/fistula to aid in defining the direction
- Dye (methylene blue) may be instilled but any spillage/leakage stains normal tissues and may obscure dissection
- An incision is made over the swelling or elliptically around a sinus orifice in the line of the cervical skin creases
- Excision of deep components and extensions is necessary to avoid recurrence
- Dissection should be carried upwards between the two main carotid artery branches to the pharyngeal wall
- A second higher transverse incision may be needed to complete excision
- The operation is illustrated in Fig. 15.2

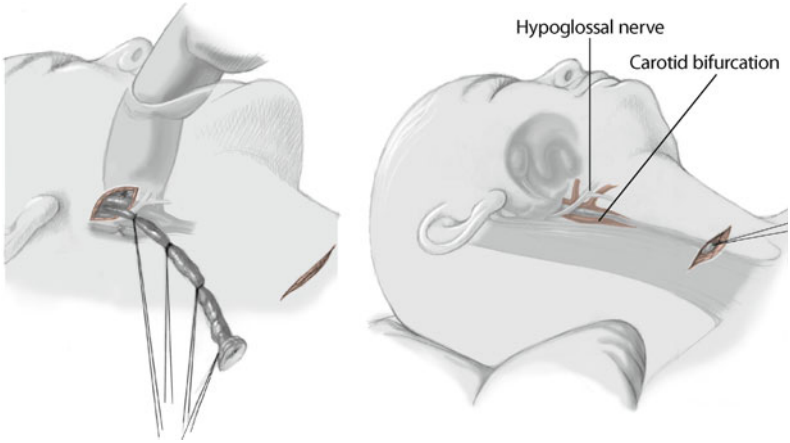


Fig. 15.2 Operative steps: brachial arch/cleft remnants

Complications

- Recurrences of second branchial arch remnants are rare and imply incomplete excision

15.2.3 Congenital Torticollis/Sternomastoid Tumor

General considerations

- The sternomastoid muscle is prone to localized thickening (endomysial fibrosis) and swelling due to trauma and hematoma caused by difficult parturition or due to hamartoma development
- This leads to shortening, torticollis (wry neck) and, eventually, to an asymmetric head shape (plagiocephaly)

Signs

- Usually asymptomatic but noticed by parents or at routine examination
- Thickened, tight sternomastoid with olive-shaped central segment
- Head tilted to the side of the lesion, with restriction of rotation to that side and of lateral deviation (flexion) to the opposite side

- When the head is viewed from above, the frontal area is prominent on the side of the lesion and the occipital area on the opposite side; the ear is relatively forward on the side of the lesion
- Secondary facial growth asymmetry is apparent when diagnosis is delayed
- Ultrasonography will define thickened muscle

Therapy

- Physiotherapy and instruction of parents in neck manipulation exercises in the first 6 months
- Resistant or delayed cases will need surgical release

Operation

- Skin incision just above clavicle gives access to sternomastoid tendon insertions
- Division of the tight sternomastoid using a tenotomy knife or an open technique
- In delayed cases, release of fibrous tissue in the cervical fascia may be needed

Postoperative care

- Postoperatively a collar support is initially helpful but early physiotherapy is important
- Parents/patient should be warned about transient diplopia due to ocular imbalance

Prognosis

- Very good

15.3 Infections

15.3.1 Cervical Lymphadenitis

General considerations

- Enlargement of cervical lymph nodes is common
- Often it is a non-specific reactive hyperplasia due to viral etiologies; jugulodigastric and deep cervical nodes are commonly involved
- Mouth, ears, nose, and scalp are primary sites for secondary cervical lymphadenitis

Signs

- Palpable, tender neck mass – may be multilocular
- Inflammation and fluctuation suggest abscess formation
- Fever and systemic illness not often marked

Differential diagnosis

- Viral – upper respiratory viruses
- Cat scratch disease
- HIV
- Toxoplasmosis
- Actinomycosis
- Acute suppurative – bacterial infection, e.g., *Staphylococcus aureus*, *Streptococcus hemolyticus*
- Chronic bacterial tuberculous, atypical mycobacterial (MAIS complex, i.e., *Mycobacterium avium, M. intracellulare, M. scrofulaceum*)
- Neoplastic – Hodgkin and non-Hodgkin lymphomas

Preoperative work-up

- Ultrasonography may elucidate the presence of pus
- Antibiotics if patient present with systemic illness or is a young child

- Needle aspiration (with antibiotic instillation) may be diagnostic and therapeutic
- Occasional retropharyngeal extension poses risks for intubation anesthesia

Therapeutic strategy

- Abscess may be encouraged to discharge spontaneously or to point when using warm poultice
- When pointing, incision and drainage (with packing if large cavity) is definitive
- Atypical mycobacteria infection is insensitive to antituberculous drugs and requires complete excision
- Solid masses may need biopsy for tissue diagnosis

15.3.2 Thyroid Enlargement

General considerations

- Thyroid enlargement in children is unusual and is most commonly due to simple goiter
- Thyroiditis, neoplasia, acute inflammation, cysts and Graves' disease (hyperthyroidism) all cause enlargement
- The anatomy of the thyroid gland is shown in Fig. 15.3

Differential diagnosis

- The differential diagnosis of thyroid enlargement is given in Table 15.2

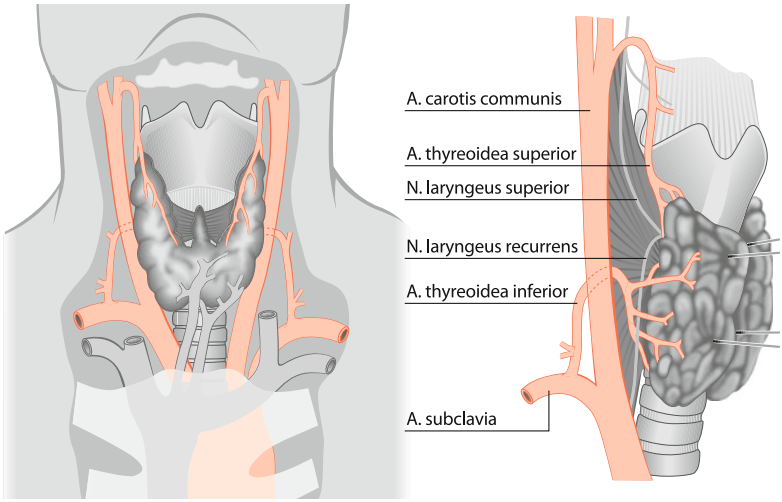


Fig. 15.3 Thyroid anatomy

Table 15.2 Differential diagnosis of thyroid enlargement

Thyroid enlargement	State	Age	Description
Congenital hypothyroidism	Euthyroid	Newborns	Usually not enlarged, but may have an ectopic gland
Simple colloid goiter	Euthyroid	Adolescence	Smooth, uniform or nodular gland enlargement
Chronic lymphocytic thyroiditis (Hashimoto's)	Euthyroid but can progress to hypothyroid	Adolescence	Autoimmune disease, firm gland
Subacute viral thyroiditis (de Quervain's)	Euthyroid	Any age	Usually tender and painful, mildly thyrotoxic
Acute suppurative (bacterial) thyroiditis	Euthyroid	Any age	Acute swollen and painful, sepsis
Toxic goiter (Graves')	Hyperthyroid	Any age	Diffuse enlargement
Thyroid nodules and cysts	Hypothyroid	Any age	Adenoma most common but risk of malignancy
Thyroid carcinoma	Both	Adolescence	More common in girls, usually papillary

Preoperative work-up

- Palpation of gland – size, symmetry, firmness (hard-neoplasia), tenderness (inflamed)
- Thyroid state – eu-/hypo-/hyperthyroid – based on physical signs in addition to thyroid-stimulating hormone (TSH) and free thyroxine (T_4) estimations in plasma
- Imaging may include ultrasonography, radionuclide scintigraphy (^{131}I) and ^{99m}TC-scan
- Biopsy by fine needle aspiration (FNA)
- Consider possible association with multiple endocrine neoplasia (MEN) syndromes

Therapy

- Antithyroid medications for thyrotoxicosis
- Radioactive gland ablation – possible long-term cancer risk

Operation

- Thyroidectomy – subtotal or total – recurrent laryngeal nerves at risk

15.3.3 Hyperparathyroidism

- The four parathyroid nodules are located posterior to the lateral thyroid lobes and are derived from the third and fourth branchial arches
- Hyperparathyroidism (HPT) may present with urolithiasis, peptic ulcer, constipation, and depression

Vitamin D/parathormone metabolism

- The metabolism of these two substances regulates the calcium concentration in blood (Fig. 15.4)

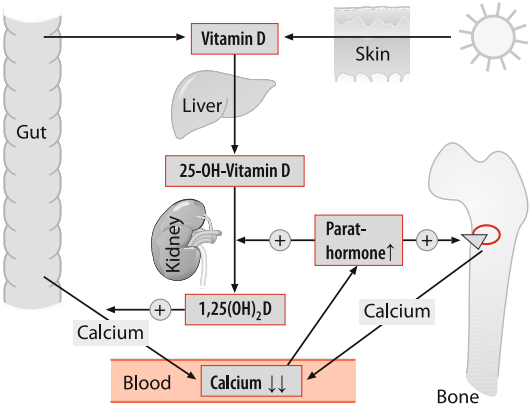


Fig. 15.4 Vitamin D/parathormone metabolism

Classification

- Hyperparathyroidism is classified as primary, secondary or tertiary (Table 15.3)

Table 15.3 Classification of hyperparathyroidism (*HPT*)

Primary HPT	Secondary HPT	Tertiary HPT
▪ Hyperplasia ▪ Pituitary adenoma	▪ Vitamin D deficiency (rickets) ▪ Chronic renal disease ▪ Maternal hypoparathyroidism	▪ Chronic hypocalcemia

Laboratory investigations and findings

- Laboratory investigations vary according to the type of HPT (Table 15.4)

Table 15.4 Laboratory investigations and findings according to the type of HPT

Investigation	Primary HPT	Secondary HPT
Parathormone	Raised	Normal, raised
Serum calcium	Raised	Normal, lowered
Urine calcium	Normal, raised	Raised
Serum phosphate	Normal, lowered	Normal, lowered, raised
Urine phosphate	Raised	Lowered
Alkaline phosphatase	Normal, raised	Raised

Preoperative work-up

- Neck ultrasonography
- Localization of tumor by selective thyroid vein sampling and parathormone estimation
- MRI scan

Operation

- Resection of adenoma or selected glands

16 Thorax

16.1 General Considerations

16.1.1 Anatomy

- The respiratory system (Fig. 16.1) arises as an out-pouching from the primitive lung bud at the 3rd week of embryonic life. Between the 6th and 16th weeks of gestation, rapid bronchial division beyond the subsegmental level occurs
- Alveoli begin to develop at the 24th week of gestation by out-pouching, so that by 8 years of age the adult number of approximately 300 million alveoli is attained
- Each lung is divided into 10 segments (Fig. 16.2). For certain localized lesions segmental and subsegmental resections rather than lobectomy should be performed
- The pulmonary lymph nodes are located at points of division of segmental bronchi or at the bifurcations of the pulmonary artery. The hilar nodes are allocated along the main bronchi. The interlobar nodes are situated in the angles of the main bronchi into the lobar bronchi
- The child's thorax is more flexible than that of adults

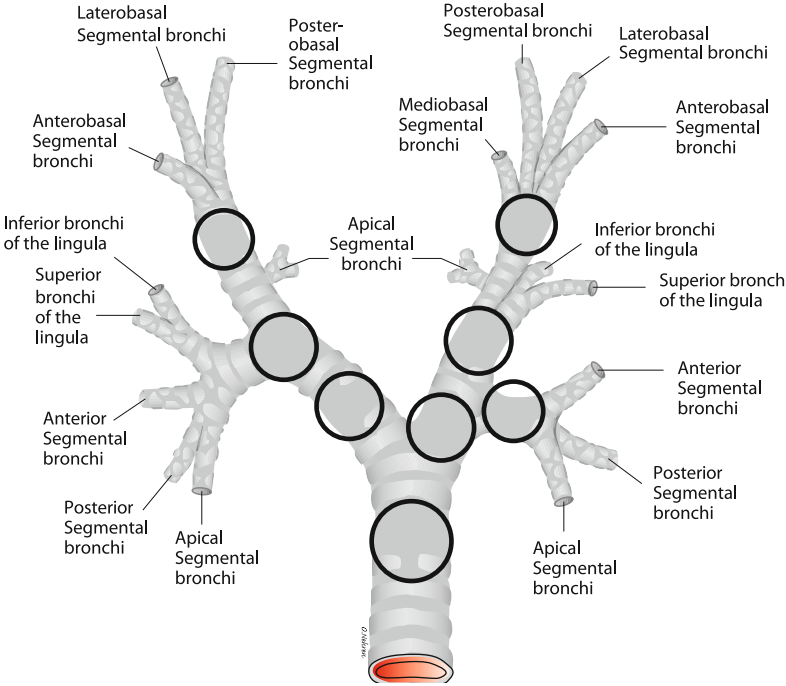


Fig. 16.1 Anatomy of the tracheobronchial tree

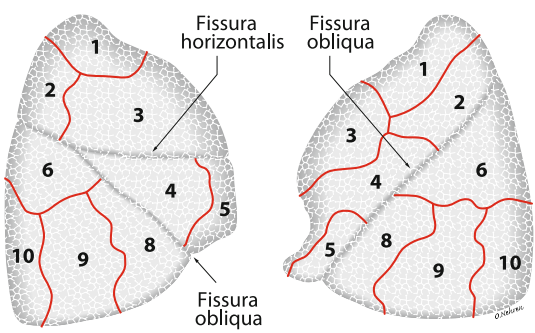


Fig. 16.2 Lung segmentation

16.1.2 Tracheostomy

General considerations

- Tracheostomy is occasionally an emergency procedure but is more usually semi-elective or elective
- A tracheostomy offers direct and established access to the major airways, reduces airway dead space, overcomes upper airway obstruction, and avoids interference with the vocal cords and larynx

Indication for operation

- Long-term mechanical ventilation (approx. 4–6 weeks, e.g., post cardiac surgery)
- Congenital laryngotracheal malformations (tracheal membrane, subglottic stenosis, micrognathia)
- Acute infection (acute epiglottitis, acute laryngotracheobronchitis)
- Upper airway injuries and edema (cervical trauma, inhalation burn injuries)
- Laryngeal edema following intubation
- Functional obstruction (vocal cord paralysis – recurrent laryngeal nerve damage)
- Laryngotracheal tumors (lymphangioma, hemangioma, papilloma)
- Respiratory insufficiency due to muscle dystrophy
- Preparatory to planned surgery on pharynx or larynx

Operation

- General anesthesia
- Hyperextended neck (shoulder roll)
- Vertical or horizontal incision midway between the cricoid and the suprasternal notch
- Division of superficial cervical fascia and platysma
- Subcutaneous preparation and division of the strap muscles in the midline

- Two U-stay retraction sutures 3-0 (2-0) through the skin on either lateral side of the proposed tracheal incision (3rd and 4th tracheal ring; caution because of the endotracheal tube)
- A single midline incision is created
- Removal of the tracheal cartilage is not recommended
- Insertion of appropriate diameter tracheostomy tube as the endotracheal tube is withdrawn slowly and the tracheostomy tube (size approx. the diameter of the patient's 5th finger) is pushed in without allowing the trachea to collapse
- Two additional sutures are required to fix the skin to the tracheal wall. For larger tubes inflate the cuff carefully (less than 20 mmHg). The tracheostomy tube should remain unchanged until at least the postoperative day 5
- Secure fixation of the tracheostomy tube using tapes tied around the neck with or without suture to skin
- The operation is illustrated in Fig. 16.3

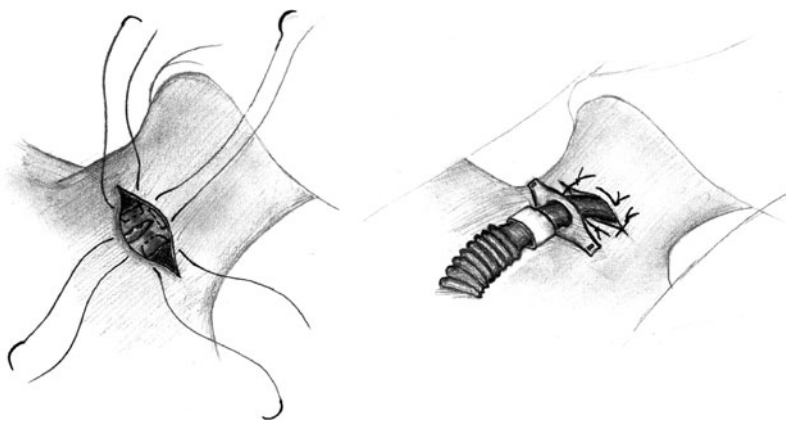


Fig. 16.3 Operative steps for tracheostomy

Postoperative

- Chest X-ray
- Flexible bronchoscopy if necessary

Complications

- Tube obstruction
- Granulation tissue
- Tracheomalacia (local inflammatory destruction)
- Severe hemorrhage (from the innominate artery especially in tracheotomies below the fourth tracheal ring, urgent median thoracotomy)
- Stricture
- Stenosis, delayed spontaneous closure of the tracheostomy by a skin flap

Tracheotomy care

- Check chest X-ray immediately after the operation
- Warm, humidified, filtered air/O_2 necessary
- Keep spare tracheotomy tube and tracheal dilator readily available
- Regular tube irrigation (0.9% NaCl) and aspiration
- Careful antiseptic technique of tube changing
- Change tube at regular intervals (2–3 days)
- Instruct the parents thoroughly

16.1.3 Thoracotomy

Median sternotomy

- Indicated for access to both lungs (metastases), the heart, and the anterior mediastinum (thymoma)
- Mobilization (digitally, long blunt clamp) of the retrosternal connective tissues from the suprasternal notch and the subxiphoidal region
- Median (sometimes difficult) sternal division using a chisel or saw. Mechanical ventilation should be stopped for a short time during this procedure to avoid making lesions in the pleura sacs and lungs
- Full exposure using the Finochietto retractor
- Closure using resorbable sutures (0, 1, 2) or surgical wire

Lateral thoracotomy

- Lateral thoracotomy is frequently used in pediatric surgery
- The patient is positioned overextended on his or her left or right side (Fig. 16.4), with support under the chest and the arm elevated. After surgical covering, the mammilla and the inferior angle of the scapula (palpation) should be visible for orientation
- The incision is made, starting far behind the mammilla and going around the inferior angle of the scapula or along the middle axillary line
- The latissimus dorsi muscle and the thoracodorsal fascia have to be divided (care should be taken to stay below the inferior angle of the scapula); the serratus muscle is partially divided
- The 4th intercostal space is used to enter the thorax. Therefore, a longitudinal periosteal incision is performed using a diathermy knife, in the middle of the 5th rib. The upper half of the periosteum is mobilized step by step from the rib starting dorsally and moving ventrally (Fig. 16.5)
- For operations on the diaphragm it is advisable to use the 5th or 6th intercostal spaces
- Rib resections are not necessary
- Make an incision in the partly mobilized periosteum
- The Finochietto retractor is inserted and opened
- Access to the mediastinum (e.g., esophageal atresia) should be performed extrapleurally (pleural lesions after blunt dissection are mainly located in the area of the anterior chest wall)
- At the end of the operation pleural drainage should be ensured (Fig. 16.6), usually with two chest tubes (short intrathoracic tube in the posterior position, long tube in the anterior position)
- Closure is achieved with resorbable sutures around the two ribs either side of the opened intercostal space, ensuring that they are not tied too firmly
- Closure of the muscle layers and the fascia thoracodorsalis as well as the subcutaneous tissues is made using resorbable sutures
- Skin closure

Fig. 16.4 Positioning for lateral thoracotomy

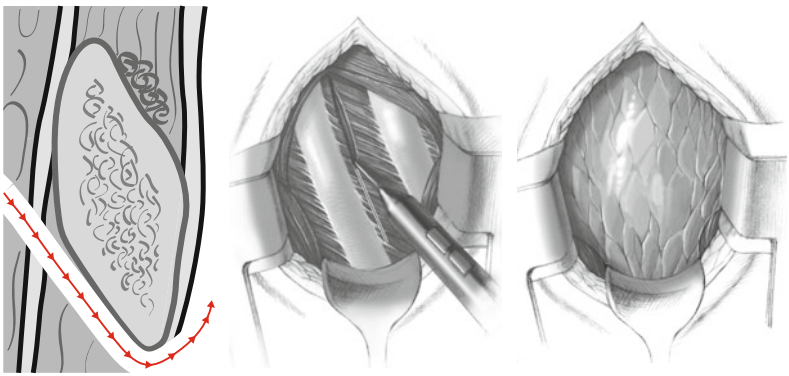


Fig. 16.5 Operative steps for lateral thoracotomy

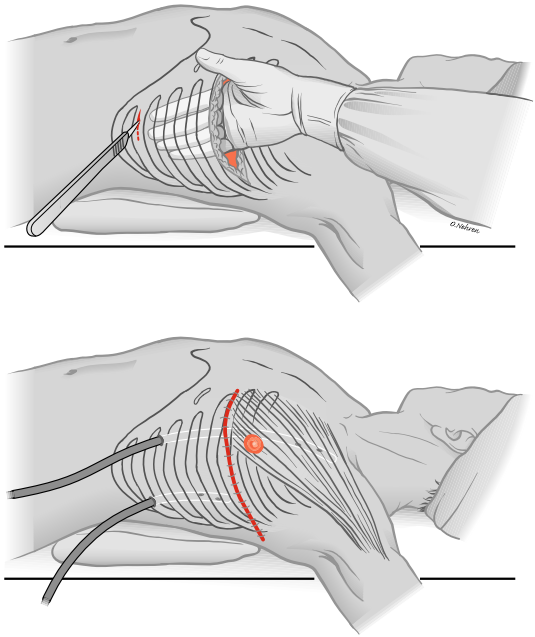


Fig. 16.6 Postoperative drainage of the thorax

Axillary thoracotomy

- This is only performed when limited exposure of the upper hemithorax is needed (i.e., biopsy, pleural abrasion)
- The skin incision parallels the course of the third rib in the axilla and extends from the anterior to the posterior axillary folds
- Division of the latissimus dorsi or the serratus muscle is not necessary

16.1.4 Bronchus Closure

Surgical procedure

- Closure of large bronchial stumps with commercial surgical stapling devices (Endo-GIA) is appropriate, especially in older children
- Closure of small and middle bronchi and of large bronchi in small children is best achieved with a simply sewn monophilic resorbable suture (e.g., PDS®, Monosyn®)

- The distance between the stitches should be about 3 mm
- During suturing intermittent atraumatic bronchus occlusion using a wet peanut swab on a clamp should be performed
- The procedure is illustrated in Fig. 16.7

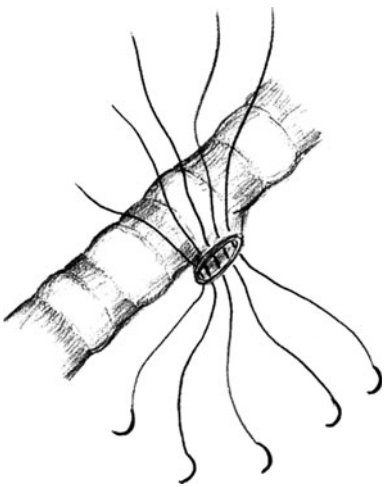


Fig. 16.7 Bronchus closure

16.1.5 Large Vessel Closure (Pulmonary Arteries and Veins)

Surgical procedure

- Non-resorbable sutures (Prolene®, Gore®) are used
- A double closure technique is applied with a simple ligature centrally and a suture ligature peripherally

16.1.6 Lobe/Segmental Resection

Surgical procedure

- The operation is illustrated in Fig. 16.8
- Lateral thoracotomy (incision of the pleura using the scalpel)
- The Finochietto retractor is inserted and gradually opened taking care not to clump the lung; a rib resection is not necessary in children
- Careful intrathoracic exploration (e.g., look for pleural adhesions)
- The visceral pleura is carefully incised exposing the hilar structures
- Careful preparation of the vessel branches (pulmonary artery, pulmonary vein) and the bronchus (sometimes difficult because of enlarged lymph nodes)
- Identification of all structures before dissecting them after exposing the segmental branches of the vessels
- Vessel loops are helpful, as intermittent occlusion of the segment bronchi with soft vascular clamps (e.g., bulldogs) at the same time as lung inflation helps to detect the borders between the segments
- If the lesion is limited to lung segments, just segmental resection may be chosen
- After segment resections or after division of interlobar parenchymal bridges the lung surface may be sealed, if necessary with fibrin glue or a fibrin-coated collagen patch
- For lower lobe resection, preparation begins in the interlobium with an incision of the visceral pleura followed by preparation of the pulmonary artery and its branches
- In lower lobe resections, the pulmonary ligament can be dissected (use diathermy for hemostasis) to allow the lobe to be lifted up easily
- In upper lobe resections surgical preparation should start at the anterior aspect of the hilus
- The pulmonary veins are better exposed from the dorsal aspect of the hilus
- The arteries are closed before the veins with non-resorbable sutures
- In right lower lobe resection attention must be paid to the origin of the middle lobe bronchus. It may be necessary to cut the right main bron-

chus below the origin of the middle lobe bronchus and the bronchus of the lower apical segment separately

- Parenchymal bridges between the lungs may be divided using a commercial stapling device (Endo-GIA) or mattress sutures (eventually reinforced with pledges)
- Occlusion of the bronchus in small children is usually made using long-term resorbable sutures (in older children stapling devices may be used)
- Air leaks may be controlled by filling the thoracic cavity with warm saline solution, whilst the anesthetist inflates the lung; no air bubbles should arise
- Hemostasis should be checked
- Two thoracic tubes are placed in the chest, a long one at the front for air drainage and a short one at the back for fluid drainage

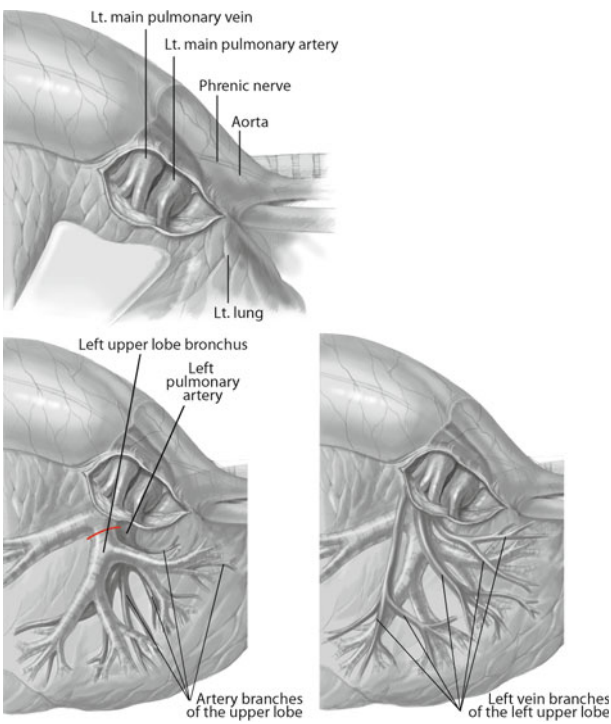


Fig. 16.8 Operative steps for lobectomy

16.1.7 Minimally Invasive Procedures

Indication for minimally invasive procedures

- Diagnostics and biopsy results (lung, lymphatic tissue, mediastinal masses)
- Debridement of fibrinopurulent pleural effusions
- Emphysematous blebs are usually found in the apical part of the upper lobes
- Benign mediastinal masses or pleurectomy, avoiding making lesions in the sympathetic chain especially in the apical area of the chest

Patients and team position

- Patient is usually in the standard lateral decubitus position
- Arm is elevated over the head
- Straight line principle (surgeon – target – monitor) (Fig. 16.9)

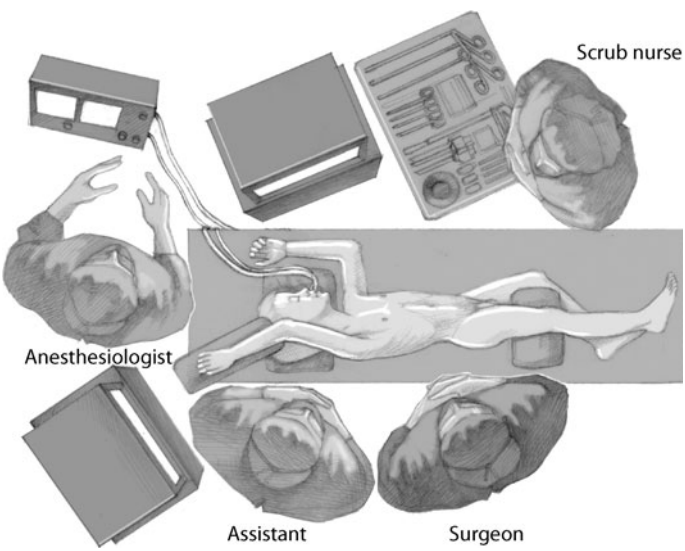


Fig. 16.9 Theater setting for thoracoscopy

Ventilation

- Single lung ventilation (Carlens double lumen tube; smallest tube: 28 F; cannot be used in children below 30 kg bodyweight)
- Not mandatory for small children, but, if necessary, blockage of the ipsilateral main bronchus can be achieved using a Fogarty catheter (Fig. 16.10)
- Collapse of the lung can be achieved by CO_2 insufflation into the pleural cavity (max. flow: 1 l·min^{-1}; maximum pressure: 4–6 mmHg, 4 mmHg is better)

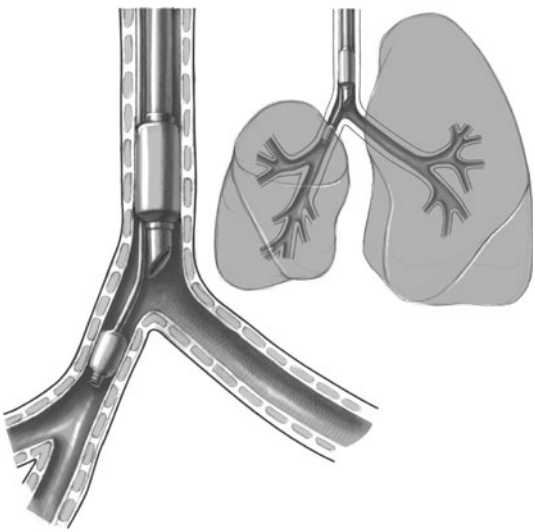


Fig. 16.10 Single bronchus ventilation using a Fogarty catheter

Port position

- Insert the telescope port (5 mm, 30°) in the middle axillary line, preferably opposite the area of interest to allow for the widest initial survey
- Two to three working ports are inserted in the anterior and posterior axillary line, usually between the 4th and 8th interspaces (5–10 mm; valved trocars) (Fig. 16.11). Avoid liver and spleen puncture
- Thoracic tubes can be inserted at the end of the procedure using the inferior port access ports

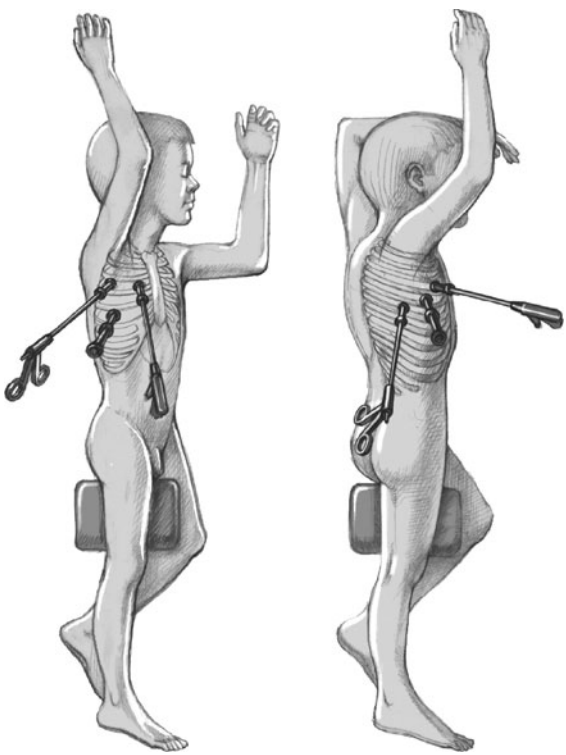


Fig. 16.11 Port positions for minimally invasive procedures in the thorax

16.2 Chest Wall

16.2.1 Breast Diseases

Developmental breast disorders

- Polythelia; additional nipples and areolas may develop anywhere along the milk line from axilla to pubis (most common location of accessory nipples is on the chest below the actual breast)
- Breast hypoplasia
- Breast aplasia in Poland syndrome (absent or diminished underlying chest wall structures)
- Asynchronous thelarche: One breast bud may appear weeks to months ahead of the other (biopsy never indicted, just observation)
- Premature thelarche: breast development starts before the age of 8 (just observation)
- Swelling of the breast in the neonatal period (just observation)
- Virginal hypertrophy
- Unilateral hypertrophy

Gynecomastia

General considerations

- Unilateral or bilateral benign enlargement of the male breast mainly at puberty (greatest prevalence at 13–14 years; Fig. 16.12)
- Imbalances between estrogen and androgen concentrations
- Microscopically a proliferation of dense periductal connective tissue and marked hyperplasia of the ductal linings
- Enlargement may also occur in response to an excess of estrogen or decreased androgen production (Klinefelter syndrome, Leydig cell tumor, rarely because of Sertoli cell tumor, prolonged liver failure cirrhosis, sometimes seen in cases of hermaphroditism, in cases of severe malnutrition)
- Pseudogynecomastia due to adipositas
- Physiological slight hypertrophy is seen in the neonatal period (days 3–8) and during puberty

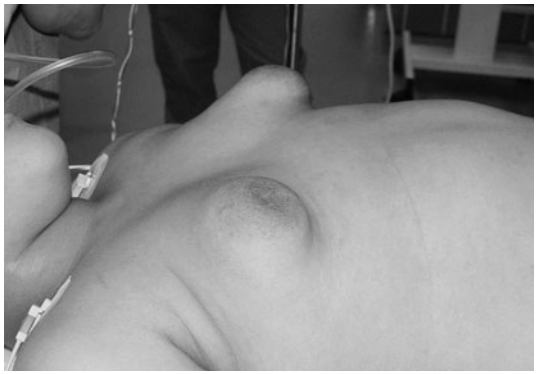


Fig. 16.12 Adolescent boy with gynecomastia

Signs

- Unilateral or bilateral enlargement of the male breast
- Palpation reveals a feeling of a disk of rubbery tissue (not palpable in pseudogynecomastia)
- Painful swelling is typical in adolescent boys (swelling of the gland, protrusion of the nipple, friction on the shirt)
- Pubertal gynecomastia generally resolves within 1 year

Preoperative work-up

- Ultrasonography of the testis and the liver
- Investigation of the testis, the liver, endocrinological work-up [human chorionic gonadotropin (hCG), luteinizing hormone (LH), testosterone, estradiol], chromosome analysis (Klinefelter syndrome), and history of drugs
- In cases of physiological gynecomastia, reassurance and follow-up at 1, 3, and 6 months are required to observe regression

Indication for operation

- Severe pain, tenderness
- Psychological distress

Operation

- Subcutaneous total mastectomy (reaching the fascia of the pectoral muscle) through a semicircular periareolar incision
- In severe cases mammary reduction plasty is necessary (McKissock technique)
- Suction drainage may be indicated

Postoperative care

- Remove suction drainage on the second postoperative day
- Remove skin sutures on the tenth postoperative day

Prognosis

- In cases of complete mastectomy, no recurrence

Mastitis

General considerations

- Bacterial (*Staphylococcus*, *Streptococcus*, *Escherichia coli*), unilateral inflammation of the breast
- Mastitis occurs more commonly after the thelarche
- Abscess formation is common

Signs

- Inflammatory painful enlargement of the breast

Preoperative work-up

- Palpation, fluctuance in cases of abscess formation
- Ultrasonography
- White blood cell count, C-reactive protein (CRP)

Indication for operation

- Rarely in mastitis of newborns (usually responds to intravenous antibiotics)
- Abscess formation

Operation

- Careful drainage via paramamillary small incision trying not to destroy breast parenchyma, especially in girls
- Material preservation for bacteriological work-up
- Careful debridement of necrotic tissue (sharp spoon)
- Rinsing with physiological sodium saline
- Insert a small rubber tube to keep incision open

Postoperative care

- Intravenous antibiotics
- Remove rubber drainage on the second postoperative day

Prognosis

- Good

Breast tumors

- Masses in prepubertal breast are nearly always benign
  - Fibroadenoma of the breast (in adolescence they are benign, solitary, firm, non-tender nodules; best therapy is local resection)
  - Hemangioma of the breast
- Phyllodes tumors and cancer are very rare (nipple discharge)
- Bone and soft tissue sarcomas of the chest wall

Preoperative work-up

- CT, MRI or positron emission tomography (PET)
- Exclude inflammatory lesions

Operation

- Malignant tumors should be resected en-bloc (muscles, ribs, pleura)
- Closure of the chest, performing a thoracoplasty (using a corium skin graft, an artificial resorbable net or a non-resorbable PTFE–Gore® patch)

Postoperative care

- Postoperative radiation and chemotherapy may be indicated

16.2.2 Pectus Excavatum

General considerations

- Condition in which the distance from the sternum to the vertebrae is decreased, giving a funnel-like appearance; located mostly in the distal part of the sternum (Fig. 16.13)
- Etiology is still unclear. There is a contiguity to a mechanical instability of the sternum and an overgrowth of the rib cartilage
- 90% are noticeable within the first year of life
- Occurs more frequently in boys (ratio 3:1)
- In the asymmetrical funnel the depression is deeper on the right side and the sternum is twisted
- In older patients (adolescents, adults) posterior angulation of the most anterior portion of the osseous part of the ribs occurs. This late stage should be avoided and surgical correction carried out before it occurs
- Mitral valve prolapse is frequently seen and sometimes also mild valve insufficiencies (compression and shifting of the heart to the left)
- 15%–20% show additional musculoskeletal abnormalities such as scoliosis
- 37% have family histories

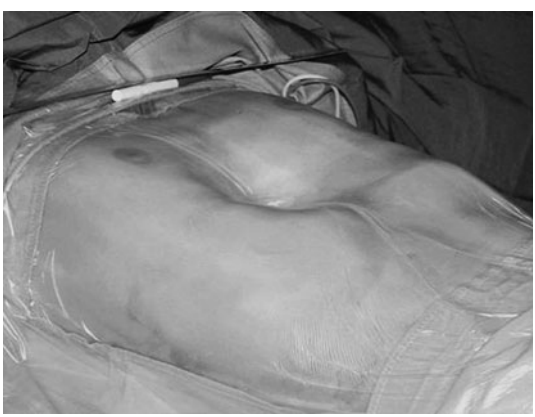


Fig. 16.13 Typical funnel chest

Signs

- Pectus excavatum is well tolerated in infancy and childhood
- Psychological stress
- Palpitations
- Arrhythmias and tachyarrhythmia
- In cases of severe heart shift there is reduced stroke volume. Increased cardiac output is achieved primarily by increasing the heart rate because of the limited stroke volume
- Pain in the area of the funnel
- Exercise intolerance

Preoperative work-up

- X-ray a.p. and lateral with contrasted border of the funnel on the skin
- Determination of a funnel chest index
  - Vertebral index [VI = (vertebral diameter × 100)/(sagittal diameter+vertebral diameter); normal value 15–25] (Fig. 16.14)
  - Fronto-sagittal index (FSI = sagittal diameter × 100/frontal diameter; normal value 36–56)
    - Vertebral diameter (VD) = the diameter of the body of the vertebra directly behind the deepest point of the funnel
    - Sagittal diameter (SD) = the distance between the deepest point of the funnel behind the sternum and the ventral wall of the vertebra
    - Frontal diameter = the biggest distance between the ribs in the frontal plane
- CT scan (Fig. 16.15) or MRI
- Heart ultrasonography to detect insufficiencies or mitral valve prolapse
- 24-h ECG in different positions
- Pulmonary function
- Psychological evaluation (degree of psychological distress caused to the child)

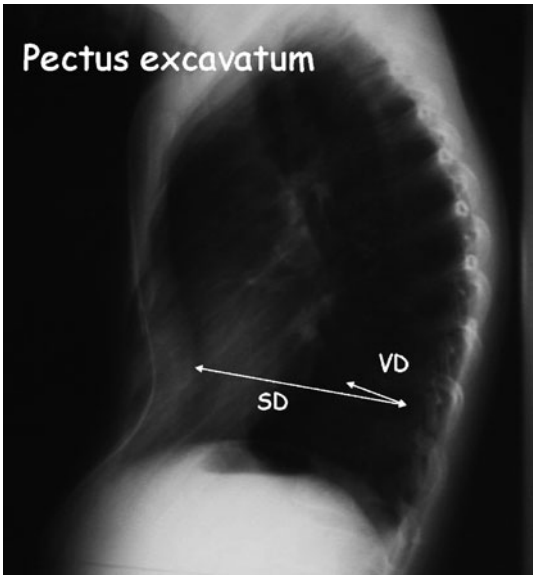


Fig. 16.14 X-ray for determination of the vertebral index

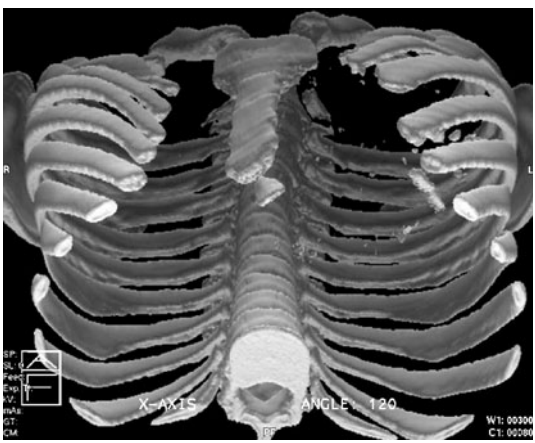


Fig. 16.15 CT 3D funnel chest reconstruction

Indication for operation

- Psychological distress
- Arrhythmias, valve insufficiencies
- Exercise impairment
- Vertebral index >26

Operation

Reconstruction of the thorax wall can be performed by different operations. Minimally invasive methods are more accepted nowadays but do not always offer the expected result. Compared to the minimally invasive technique, the Ravitch–Welsh–Rehbein method is a more radical approach and its acceptance is limited. However, there are still patients suffering from a highly asymmetrical and/or very stiff funnel chest (e.g., adult patients) who benefit from this technique. Especially in mixed forms (pectus carinatum and excavatum) the techniques may be combined (resection of rib cartilage in the carinatum area and elevation using the retrosternal bar in the excavatum area).

Nuss procedure

- This is a minimally invasive surgical correction (Fig. 16.16) using an individually curved C-shaped steel bar with lateral stabilizers to elevate the sternum
- Via two lateral incisions the bar is placed behind the sternum at the area of the deepest point of the depression
- In older patients two bars as well as a partial horizontal sternotomy at the upper median edge of the funnel may be necessary
  - If possible, meet the requirements for single lung ventilation
  - The length of the pectus bar is determined by measuring the distance from the right to the left midaxillary line minus 1–2 cm to allow for the fact that the bar passes under the sternum
  - Two bilateral short incisions are made in the midaxillary lines followed by mobilization of the subcutaneous tissue

- A thoracoscopy (e.g., 5 mm port) is inserted usually from the right lateral incision to visualize the deepest point of the funnel and the introducer is inserted
- For optimal bar placement the intercostal space should be perforated within the edges of the funnel using the introducer. Take care to avoid making lesions to the internal thoracic artery, the lungs, the pericardium or the heart
- A strong tape is tied to the tip of the introducer, which is pulled through the thorax behind the sternum
- The individually prepared C-shaped bar is then bound to one side of the tape and the bar is positioned behind the sternum with both ends facing anteriorly, i.e., the convexity facing posteriorly
- Then the bar is rotated 180° to raise the sternum and anterior chest wall, and is then fixed with sutures to the lateral thoracic muscles (M. serratus) using a single stabilizer plate on one side. The latest generation of single-piece pectus bars have one integrated stabilizer lash which cannot be dislocated
- No chest tubes are normally necessary, however one could use them

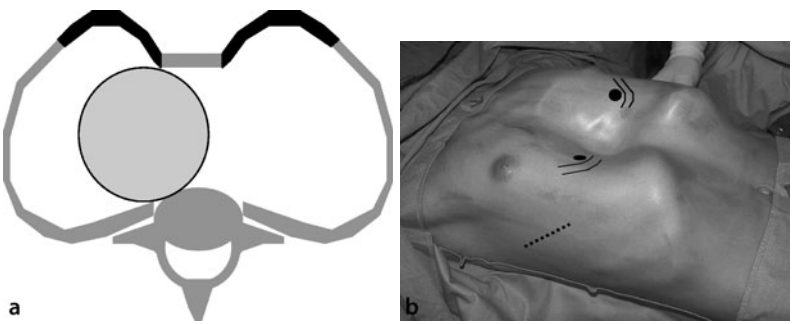
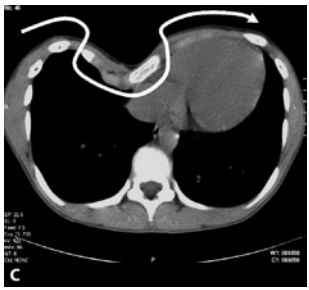
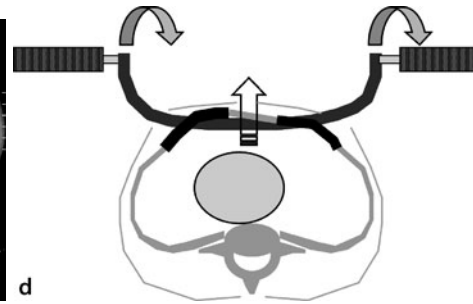
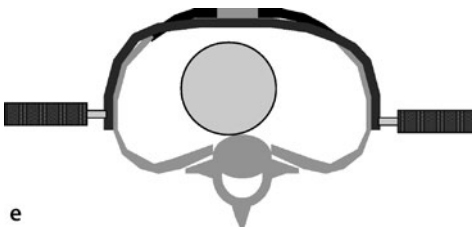
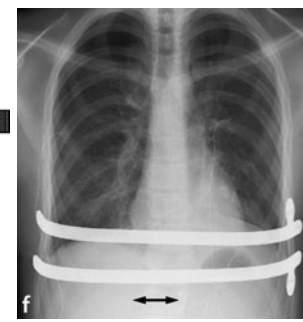


Fig. 16.16a–g Operative steps for the Nuss procedure. **a** The patient has a symmetrical funnel chest. **b** Marking the preparatory landmarks: *black spots* are intercostal port sites. **c–g** *see next page*









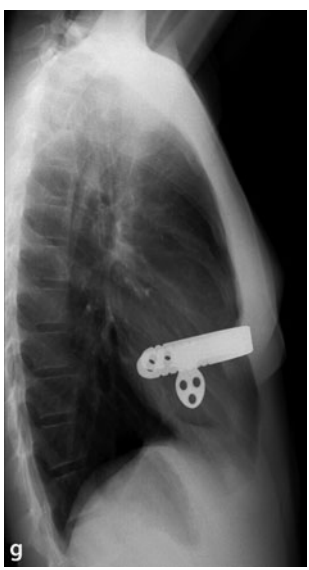


Fig. 16.16 *(continued)* Operative steps for the Nuss procedure. **c** The intercostal perforations are within the edge of the funnel. **d** The C-shaped bar is rotated 180° to raise the sternum and anterior chest wall. **e** Final aspect. **f** X-ray (a.p.) after implantation of the bar. **g** Profile X-ray after implantation of the C-shaped bar

Rokitansky modifications

- For simple funnel chest
  - The above steps for the Nuss procedure are followed, however a single-piece pectus bar with an integrated stabilizer lash is used
- For stiff curved sternum
  - A horizontal partial sternotomy at the upper edge of the funnel is recommended
  - A small, third, median incision below the xiphoid process is made to introduce a sharp bone retractor (Rochards retractor) that elevates the sternum over a long period and helps to mobilizes the retrosternal adhesions between the sternum and pericardium. A mediastinoscopy could be performed
  - Where the thorax is extremely stiff, lateral thoracoscopy should be performed in order to conduct chondrotomies (the dissection or surgical division of cartilage) (Fig. 16.17) on involved ribs using electrocautery
  - Sometimes the Nuss procedure has to be completed by cartilage resections

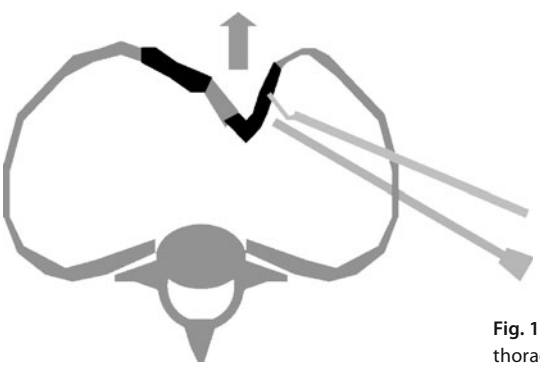


Fig. 16.17 Operative steps for thoracoscopic chondrotomy

Postoperative care

- Analgesics are necessary particularly in the first postoperative week (piritramide 0.1 mg·kg^{-1} bodyweight·h^{-1} or preferably patient-controlled analgesia, PCA)
- Peridural or epidural pump-controlled anesthesia is useful in the first days postoperatively
- The patient should start breathing exercises as soon as possible
- Chest tubes should be taken out as soon as possible
- The bar should stay in place for at least 2–3 years
- The advantages of this technique over the Ravitch–Welsh–Rehbein method are the short duration of the operation, the short hospital stay, and the relatively short postoperative restriction on physical activity
- Difficulties may occur if this technique is used on patients suffering from an extremely asymmetric funnel (it gives a cosmetically poor result because of protrusion on the parasternal area of the left thorax)

Ravitch–Welsh–Rehbein procedure

- The sternum is elevated by partial bilateral resection of the elongated rib cartilages (ribs 3–7), sternotomy (wedge osteotomy), and stabilization using Rehbein metal struts (Fig. 16.18)
- This operation should not be performed below the age of 4 years (possible impairment of chest growth caused by a lesion at the costochondral junction)
  - A horizontal or sagittal incision is made in the central area of the funnel
  - The skin and pectoralis major are mobilized synchronously and elevated
  - The rib cartilage to be resected is determined and incisions made anteriorly in the perichondral tubes using electocautery. Especially in young patients complete resection of cartilage should be avoided, as the costochondral junction is important for growth
  - Pleural lesions occur most commonly on the 3rd or 4th right ribs
  - Take care to avoid making a lesion in the internal mammary vessels

- The xiphoid is divided from the lower sternum
- A partial horizontal wedge osteotomy at a level above the last deformed cartilage is performed
- A fibrin-coated collagen patch may be used to achieve hemostasis
- Fixation of the osteotomy using wire suture, inducing elevation of the sternum
- Bilateral insertion of the Rehbein struts into the bone marrow of one or two pairs of ribs (e.g., 3rd or 4th ribs). They are medially joined to each other by wire and/or non-resorbable sutures creating a metal arch anterior to the sternum
- The sternum is sewn to the artificial Rehbein strut arch to secure it in a forward-facing position
- The xiphoid is fixed again to the sternum and the pectoral muscles are approximated and sewn at the middle above the elevated sternum. If tension prevents complete approximation, a patch (e.g., Gore soft tissue®) should be used
- A submuscular and/or subcutaneous suction drain may be placed for 2 days postoperatively

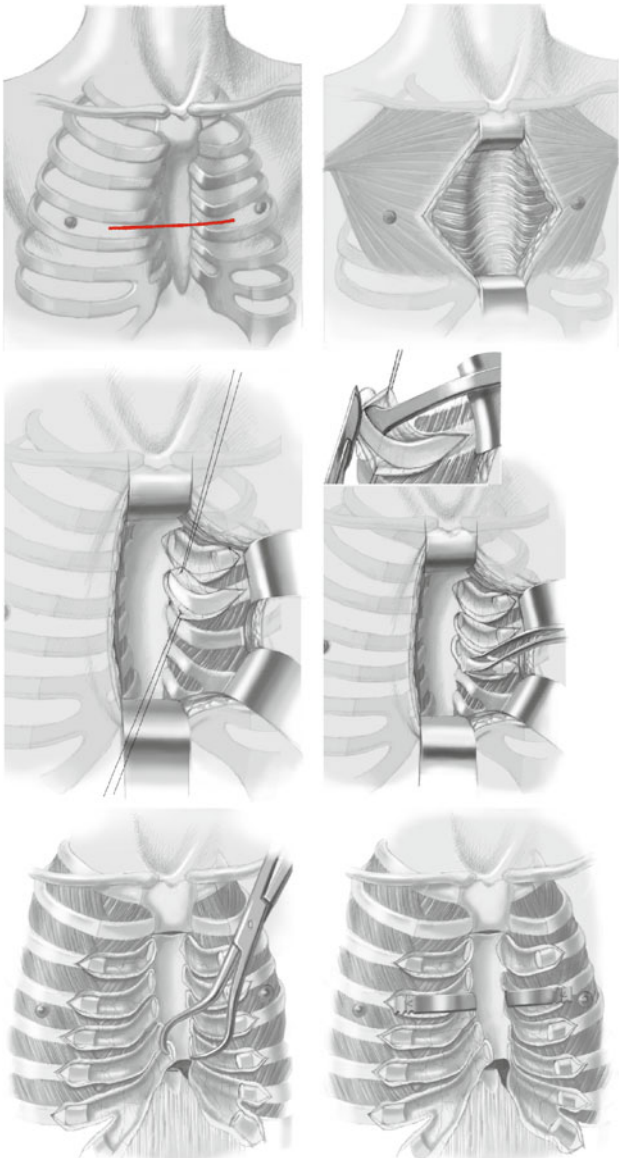


Fig. 16.18 Operative steps for the Ravitch procedure

Postoperative care

- Remove pleural drainage in the first postoperative week (when the quantity of drained fluid is less than 100 $ml \cdot day^{-1}$) and the suction drainage around the second postoperative day
- Intravenous analgesics (e.g., piritramide 0.1 $mg \cdot kg^{-1}$ $bodyweight \cdot h^{-1}$) and antibiotics
- Start respiratory exercises as early as possible
- Full muscular training can start after complete healing once a mechanically resistant scar has developed (especially for thoracic and abdominal muscles; swimming is recommended), starting the second postoperative month
- The Rehbein struts should stay in place for at least 6 months

Silicon cushion

- A plastic cushion may be implanted in the funnel for cosmetic reasons, but due to growth the results are uncertain. This technique cannot be recommended because it does not resolve the symptoms and the physical as well as the respiratory and hemodynamic problems

Prognosis

- Recurrence rate is up to 5% or 10% (higher in patients with Marfan syndrome)
- Delaying surgery until the child has begun the pubertal growth spurt is beneficial

16.2.3 Pectus Carinatum

General considerations

- Protrusion deformity of the sternum (Fig. 16.19); less frequent than pectus excavatum
- Etiology remains unclear
- The deformity appears during adolescence and is noted very rarely at birth

- Occurs more frequently in males (ratio 3:1)
- Cardiopulmonary effects have not been demonstrated
- 15% show additional musculoskeletal abnormalities such as scoliosis

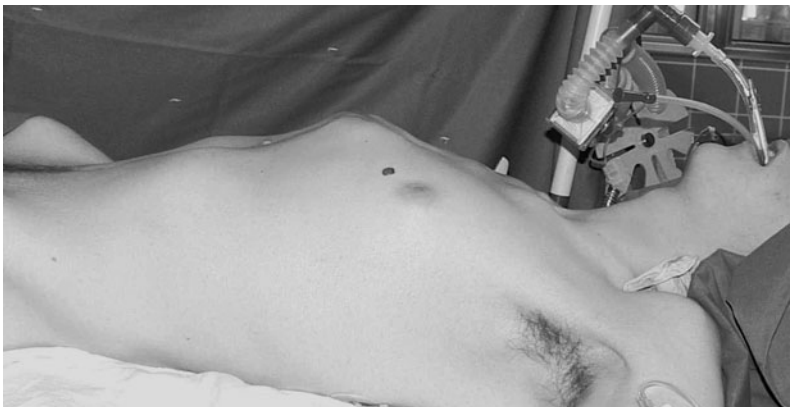


Fig. 16.19 Typical pectus carinatum

Signs

- Psychological stress
- Pain in the area of the protrusion (frequent local trauma)

Preoperative work-up

- X-ray a.p. and lateral
- CT scan or MRI

Indication for operation

- Psychological stress
- Local pain

Operation

- Depression of the sternum is achieved by partial bilateral resection of the elongated rib cartilages and sometimes additional sternotomy (wedge osteotomy)
  - A horizontal or sagittal incision is made in the central area of the defect
  - The skin and pectoral muscles are mobilized and elevated
  - The rib cartilages to be resected are determined and an incision made into the perichondral tubes anteriorly using electocautery. Especially in young patients complete resection of the cartilage should be avoided as the costochondral junction is important for growth
  - Pleural lesions most commonly occur on the 3rd or 4th right ribs
  - Take care to avoid making a lesion on the internal mammary vessels
  - A partial horizontal wedge osteotomy and fracture of the anterior cortex of the sternum is sometimes necessary
  - For blood hemostasis fibrin-coated collagen patches may be used

Postoperative care

- The rib cartilage is remodeled after a period of about 2 months
- Start respiratory exercises as early as possible
- Full muscular training can start after complete healing has occurred, once a mechanically resistant scar has developed (especially thoracic and abdominal muscles; swimming is recommended), starting the second postoperative month

Prognosis

- Good results with rare recurrence

16.2.4 Sternal Cleft

General considerations

- Developmental defect of the sternum with incomplete fusion of the sternal bars in the midline
- Compared to pectus excavatum, this is a very rare deformity

- Upper, lower, partial as well as complete clefts have been described
- Upper defects are the most frequent
- Combination with ectopia cordis is possible

Signs

- Partial defects may be asymptomatic
- Protrusion from the defect during crying
- Depression of the defect during inhalation
- In large defects paradox breathing and respiratory insufficiency may occur

Preoperative work-up

- Thoracic X-ray, CT scan or MRI
- Cardiac ultrasonography examination

Indication for operation

- Signs of respiratory insufficiency
- Lower clefts of the sternum with the risk of cardiac trauma
- Sternal cleft with ectopia cordis
- Large clefts with signs of herniation of thoracic organs

Operation

- If repair is considered it should be within the first year of life
- Repair is performed in order to achieve mechanical stability and provide protective coverage of the heart. Artificial patches may be useful (e.g., Gore® dual mesh or soft tissue patch)
- Sternal cleft closure with low tension may be achieved by making additional oblique incisions through the costal cartilages as well as a wedge resection of the sternal cartilage at the sternal end of the defect to allow approximation of the two sternal halfes

Postoperative care

- The child should be kept on artificial ventilation for 1 week

Prognosis

- Excellent results when approximation is possible with minimal tension

16.3 Pleura and Pleural Cavity

16.3.1 Pneumothorax

General considerations

- Air within the pleural space
- Spontaneous pneumothorax is especially common in male teenagers, caused for example by rupture of a small lung bubble without any lung disease
- Risk of recurrence is 16% after the first and 80% after the third episode
- Pneumothorax may be caused by trauma (lung injured by broken ribs), a penetrating chest wall injury (sucking chest wound), injury to the tracheobronchial tree, a severe asthma attack, pulmonary infections with development of an air fistula, artificial ventilation, resuscitation, or by a congenital cystic lung disease
- Induced by a valve-like mechanism, tension pneumothorax is caused by increasing accumulation of air within the pleural cavity leading to a mediastinal shift which develops into a dangerous situation

Signs

- Mild dyspnea or no signs in cases of mild spontaneous pneumothorax
- Chest pain and shortness of breath
- Varying degrees of respiratory distress
- Reduced or absent breath sounds on the side of the pneumothorax
- In patients suffering from tension pneumothorax (in addition to respiratory insufficiency) hemodynamic deterioration (neck vein distension in normovolemic patients) occurs

Preoperative work-up

- Chest X-ray (misinterpretation of medial margin of the scapula with the lung surface)
- CT scan if necessary

Therapy

- Observation in cases of minimally closed stable pneumothorax. Supplemental oxygen may be necessary

Chest tube insertion

- If significant signs occur insert a chest tube [2nd or 3rd intercostal space in the midclavicular line (classic technique) or in the midaxillary line at the level of the breast nipples] to provide a water seal drainage (Bülau drainage)
  - Make a small skin incision with the patient under general anesthetic
  - Perforate the intercostal space slowly via the upper edge of the rib with the tip of a clamp
  - Remove the clamp and insert the chest tube (reinforced by a trocar) through the prepared canal
  - Remove the trocar and fix the tube with sutures (size: 3-0 to 1). A second purse suture is placed to close the skin after the chest tube has been removed
  - Connect the chest tube to the water-sealed drainage system (Bülau system)
  - Induced by breathing movements, air bubbles should pass through the water-sealed drainage system

Operation

- Surgical therapy should be considered under the following conditions:
  - If the air leak is persistent over a period of 1 week of water-sealed drainage
  - If the CT scan shows an underlying lung disease
  - In the case of a second episode
  - If full lung expansion is not possible

- Surgical methods
  - Closure of the air leak (suture or stapling with bleb resection) and/or parietal pleurectomy (apical and anterolateral areas) via thoracotomy or thoracoscopic surgery. Pulmonary blebs may be overlooked when using just the thoracoscopic approach
  - In cases of multiple recurrence, intrapleural instillation of tetracycline (for pain control instill 2% lidocaine into the chest tube 30 min beforehand) to obliterate the pleural cavity (pleurodesis) may be indicated

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes decrease to below 20–50 ml within a 24-h period
- Start respiratory exercises and physiotherapy as soon as possible

Prognosis

- Lung function may be permanently impaired if the lung is not completely re-expanded
- Children younger than 9 years of age are unlikely to develop a recurrence
- After simple drainage therapy older children have a mean recurrence rate of about 50% (16% after the first, 80% after the third episode)
- Thoracotomy, resection of the blebs, and pleurectomy together form the most suitable surgical method to avoid recurrence

16.3.2 Empyema

General considerations

- Bacterial infection of a pleural effusion following pneumonia. Pneumococci and staphylococci remain the predominant organisms
- In two-thirds of cases this follows failure of medical therapy for parapneumonic effusions
- Empyema can be due to thoracic surgery or trauma
- A bronchopleural fistula may be involved (infected fluido-pneumothorax)

Classification

- There are three stages of development, distinguished by radiographic, clinical and pleural fluid characteristics (Table 16.1)

Table 16.1 The three stages of development of empyema (exudative, fibrinopurulent, and organizing). (*LDH* Lactate dehydrogenase, *WBC* white blood cell count)

Phase	Laboratory findings	Characteristic
Exudative	▪ Low WBC ▪ Low LDH ▪ Normal glucose ▪ Normal pH levels	▪ Fluid in the pleuropulmonary space
Fibrinopurulent	▪ Increased WBC count ▪ Increased LDH (>1000 $U \cdot l^{-1}$) ▪ Low glucose (<40 $mg \cdot dl^{-1}$)	▪ Fibrin depositions producing a "peel" ▪ Limited lung expansion
Organizing	▪ High WBC count ▪ High LDH ▪ Very low glucose	▪ Thick fibrous inelastic "peel" with scar formation ▪ Lung entrapment ▪ Erosion of the lung or the chest wall

Signs

- Tachypnea, dyspnea
- Fever
- Productive cough
- Tachycardia
- Chest pain
- Reduced mobility of the chest wall at the affected site
- Decreased breath sounds and dullness to percussion on the involved side

Preoperative work-up

- Chest X-ray
- CT scan or MRI in order to differentiate between empyema and lung abscess

- Ultrasonography to determine the stage of empyema (Table 16.1) according to detection of the thick pleural peel
- Ultrasonography- or CT-guided needle thoracocentesis
- Chest tube drain (see "Thoracoscopy" below)
- Aspirated fluid should be analyzed (Gram stain, aerobic and anaerobic cultures, search for mycobacteria, and fungi)
- White blood cell count (WBC), glucose level, lactate dehydrogenase (LDH), pH
- Non-specific leukocytosis, drastically increased C-reactive protein levels

Therapy

- In the early stages (exudative phase of parapneumonic empyema) administration of antimicrobial agents alone may be sufficient
- Conservative therapy has to be successful within the first 72 h

Indication for operation

- Late stage II (fibrinopurulent effusion)
- Effusion of more than 3 cm on the X-ray that does not improve despite aggressive treatment (i.v. antibiotics, multiple drainage) over a period of 72 h
- Bronchoppleural fistula

Thoracoscopy

- Especially in young children, for whom anesthesia is necessary in order to insert a chest tube, it is better to perform a thoracoscopy instead
  - Make a small skin incision after induction of general anesthesia
  - Introduce the trocar
  - Aspirate as much effusion as possible
  - Visualize the thoracic cavity
  - The fibrinous septa can be destroyed with the scope to make the thorax one cavity
  - Irrigate with at least 2 l of warm saline
  - Place the tube

Operation

- Most empyemas require invasive interventions (large size chest tube drainage, thoracotomy with decortication)
- It is better to drain the chest and sometimes more than one chest tube is necessary to ensure adequate drainage
- If simple drainage fails intrapleural fibrinolytic agents (250,000 units of streptokinase or urokinase in 100 ml of sterile saline; chest tubes remain clamped for at least for 4 h) may be useful. Afterwards continuous suction on the chest tubes is necessary to increase the drained fluid volume and to expand the lung
- Thoracoscopy is useful in the treatment of multiloculated empyema when there is no thick visceral pleural peel. Simple debridement in the middle exudative phase is possible
- Open drainage, lavage of the pleural cavity via thoracotomy combined with decortication is indicated in the late fibrinopurulent and the organizing phases. The peel on the visceral pleura becomes a barrier to expansion, entrapping and immobilizing the lung
- Decortication with removal of fibrin leaving the visceral pleura largely intact allows re-expansion of the lung
- In cases of bronchopleural fistula thoracotomy, open drainage and resection of the involved lung parenchyma is always necessary (usually a lobectomy). Bronchus closure may be reinforced by a dorsally based intercostal muscle flap

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes fall below 20–50 ml within a 24-h period
- Antibiotic management based on cultures should be continued until the chest tubes are removed and the pulmonary infection is controlled

Prognosis

- A lung entrapment from empyema is uncommon if the patient receives proper and timely (in terms of the three phases) treatment

16.3.3 Hemothorax

General considerations

- Blunt (rib fractures) or penetrating trauma may produce a hemothorax
- Frequently accompanied by a pneumothorax
- Bleeding from the intercostal vessels, internal mammary vessels, or great thoracic vessels
- A mediastinal shift is possible

Signs

- Chest pain
- Decreased or no breath sounds on the involved side
- Dullness to percussion on the involved side
- Respiratory distress
- Depending on the blood loss (25% loss: prehypotensive; 40% loss: hypotensive – hypovolemic shock) there may be an increased heart rate
- Shock signs, i.e., unconsciousness, decreased capillary refill, cold extremities, skin color, decreased urinary output

Preoperative work-up

- Physical examination
- Monitor the respiratory and circulatory status (blood gases, blood pressure, red blood cell count, RBC)
- Blood examination (RBC, match the patient's blood type)
- Ultrasonography, looking for effluent
- Chest X-ray in an upright position (hemothorax may be missed in the supine position)
- Repeated chest X-ray (CT) to control drainage

Therapy

- Immediate volume resuscitation is necessary initially with 20 $ml \cdot kg^{-1}$ of crystalloid. The aim is to decrease the heart rate and increase the urine output (1–2 $ml \cdot kg^{-1} \cdot h^{-1}$)
- Red blood cell transfusion (initial 10 $ml \cdot kg^{-1}$)
- Chest tube insertion (early placement, large size for full drainage)

Indication for operation

- Ongoing hemorrhage, more than 20% of child's blood volume drained (blood volume: 90 ml·kg^{-1} in a newborn, 80 ml·kg^{-1} in 1 year old, 70 ml·kg^{-1} in early adolescence)
- Failure of complete drainage after several days

Operation

- Limited thoracotomy
- Evacuation of the clot
- Removal of pleural peel
- Two chest drains should be placed

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes fall below 20–50 ml within 24 h
- Antibiotics

Prognosis

- Persistent blood in the thorax increases the risk of empyema and lung entrapment

16.3.4 Chylothorax

General considerations

- Accumulation of chyle in the pleural space
- Physiologically, the chyle from the right and left lumbar trunk flows into the left subclavian vein via the cisterna chyli and the thoracic duct (aortic hiatus, at the level of the 5th thoracic vertebra inclining to the left)
- The thoracic duct has smooth muscle in its wall and valves
- Malformations of the thoracic duct, birth trauma or an unclear etiology are commonly given as causes of pleural effusion in the first few days of life

- Insertion of a central venous line, previous cardiac surgery or operations on the mediastinum, chest tubes, blunt thoracic trauma, pneumonia, and neoplasms may also induce chylothorax
- Impaired immunological status due to chronic loss of T-cells
- Fluid and electrolyte imbalance, malnutrition
- Possible mediastinal shift

Signs

- Tachypnea
- Decreased or no breath sounds and dullness to percussion on the involved side
- Lymphocytopenia as a consequence of chyle loss

Preoperative work-up

- Physical examination
- Monitoring of the respiratory and circulatory status (blood gases, blood pressure, RBC)
- Ultrasonography
- Chest X-ray in an upright position
- Thoracocentesis (aspiration of a milky fluid after oral nutrition, chylomicrons, elevated numbers of lymphocytes)

Indication for operation

- Operative therapy is not always successful and difficult, so waiting reduces the need for surgical intervention
- If chyle flow continues (more than 15 $ml \cdot kg^{-1} \cdot day^{-1}$) for a period of 1 month (patient on total parenteral nutrition)

Therapy

- Repeated thoracocentesis or chest tube to expand the lung
- Total parenteral nutrition (no oral intake) to drastically decrease the chyle flow
- Feedings restricted to medium chain triglycerides

Operation

- Detect the lesion by thoracoscopy or thoracotomy
- Place surgical clips or multiple sutures, possibly ligation of the thoracic duct (main duct) above and below the leak
- A pleuroperitoneal shunt may be effective
- In resistant cases pleurodesis may be helpful

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes fall below 20–50 ml
- No suction on the chest tubes
- Antibiotics

Prognosis

- Most chylothoraces resolve spontaneously after some weeks

16.4 Laryngotracheal Lesions

16.4.1 Laryngomalacia

General considerations

- Immaturity of the larynx with problems in infancy in 60% of cases
- Male preponderance
- Shortened aryepiglottic folds, omega- or tubular-shaped epiglottis, anteriorly and medially aligned collapsing movements of the arytenoid cartilages, excessive tissue in the supra-arytenoid area
- Increasing inspiratory effort followed by increasing obstruction

Signs

- Stridor (inspiratory fluttering noise, especially in the supine position), sometimes only during exercise. Expiration is usually noiseless
- Respiratory difficulties usually present within the first 2 weeks of life and signs may increase during the first months

- Condition exacerbated by crying
- Sometimes feeding problems
- Signs usually resolve at the age of 2 years with a benign clinical course

Preoperative work-up

- Physical examination
- Respiratory function (blood gases, oxygen saturation monitoring)
- Direct flexible laryngo-bronchoscopy (with the child under topical or general anesthesia, but breathing spontaneously, the scope is passed through the nose and the epipharynx). In severe cases, the vocal cords cannot be seen because of supraglottic collapse

Indication for operation

- Respiratory distress
- Phases of obstructive apnea
- Failure to thrive

Operation

- Tracheostomy
- Dissection (CO_2 laser) of the shortened right and left aryepiglottic fold
- Supraglottoplasty: unilateral or bilateral removal of supraglottic obstructive tissue

Postoperative care

- Antibiotics

Complications

- Postoperative supraglottic stenosis after removal of too much tissue

Prognosis

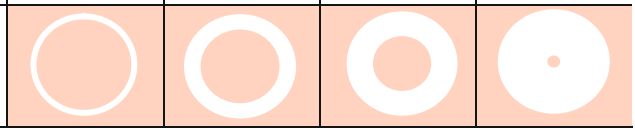
- Usually good, self-limiting condition

16.4.2 Laryngotracheal (Subglottic) Stenosis

General considerations

- Congenital malformation
- Acquired, usually after long-term artificial ventilation in neonates by a tube-related mucosal lesion in the area of the cricoid cartilage (impaired blood perfusion due to a tight fitting tube)

Classification

Obstruction	None	50%	70%	99%
View				

Signs

- Stridor and respiratory distress
- Recurrent croup-like infections
- Conjoined gastroesophageal reflux is possible

Preoperative work-up

- Respiratory function (blood gases, oxygen saturation monitoring)
- Direct flexible laryngo-bronchoscopy in a spontaneously breathing child. Intubation should be avoided especially when high-grade stenosis is suspected because of the risk of complete obstruction due to mucosal swelling. Examination under conditions for urgent tracheotomy. Endoscopy should be repeated every 6 months
- Plain X-rays
- CT or MRI is better than X-rays, especially for measuring the length of the stenosis

Differential diagnosis

- Hemangioma
- Rarely lymphangioma

Indication for operation

- Respiratory distress, phases of obstructive apnea and failure to thrive
- Stenosis grade III and more

Operation

- Tracheotomy under jet ventilation (this allows the child time to grow, it allows the stenosis to mature, however language skills may be delayed)
- Laser incision/ablation in highly selected cases of low-grade stenosis due to possible induction of scar tissue. Four quadrant incisions followed by dilatations
- Gentle dilatations in early stages including systemic steroids
- Grade III and IV lesions require open reconstruction:
  - Anterior cricoid split
  - Anterior and/or posterior cricoid split, extended 5–10 mm into the tracheal wall
  - Costal cartilage expansion, if required distraction is >3 mm
  - Perichondrium should be removed from the outer surface and preserved at the inner surface for improved epithelialization

Postoperative care

- Repeated endoscopies after 4 weeks, 3 months, 6 months, 12 months, and 24 months
- Antibiotics for 3–4 weeks
- Treat gastroesophageal reflux if necessary

Prognosis

- Good in grade I and II
- Moderate in grade III and IV

16.4.3 Benign Laryngeal Tumors

Papilloma

General considerations

- The most common benign neoplasm of the larynx
- Multiple locations (vocal cords and the anterior commissure), with possible extension into the distal airway
- No histological differences between the juvenile and adult forms
- Frequently detected between the ages of 2 and 5 years
- Caused by the human papilloma virus (HPV-6, HPV-11)
- The mothers of 50% of children with recurrent respiratory papillomas had active cervical papillomas at the time of the child's birth

Signs

- Hoarseness
- Respiratory distress

Preoperative work-up

- Respiratory function (blood gases, oxygen saturation monitoring)
- Direct flexible laryngo-bronchoscopy

Indication for operation

- Maintenance of an open airway (20%–30% require tracheotomy)

Therapy

- Antiviral agents (interferon α) with variable results

Operation

- Laser ablation/vaporization (complications: scar tissue, fixation of the vocal cord)
- Tracheotomy

Prognosis

- Recurrence is typical over a certain period
- There is no universally effective treatment

Subglottic Hemangioma

General considerations

- Rarely present at birth; develops within the first months of life
- 50% also have cutaneous hemangiomas
- Rapid growth in the first few months
- Regression by the age of 1 year (complete resolution by the age of 5 years)
- Usually a history of complete resolution

Signs

- Stridor (in the early stages: inspiratory stridor)
- Respiratory distress

Preoperative work-up

- Respiratory function (blood gases, oxygen saturation monitoring)
- Direct flexible laryngo-bronchoscopy showing a compressible lesion, usually arising from the posterolateral aspect of the subglottis
- Abdominal ultrasonography to detect liver hemangiomas
- Cardiac ultrasonography

Indication for operation

- Maintenance of the airway

Therapy

- Steroid (1 $mg \cdot kg^{-1} \cdot day^{-1}$)

Operation

- Tracheotomy to bypass the lesion and wait for regression
- Intralesional laser vaporization (Nd:YAG laser) with postoperative intubation for about 1 week

Prognosis

- Mostly spontaneous involution

16.4.4 Tracheomalacia

General considerations

- Intrinsic defect of the trachea. Inability of the cartilages to keep the trachea open during respiration
- Unusually an obstruction; due to positive intrathoracic pressure the flattened trachea collapses during expiration
- In patients after corrected esophageal atresia type IIIb, the site of collapse is usually above the entry of the esophagotracheal fistula
- Unusually severe signs as long as more than 20% of the lumen remains open during the breathing cycle

Differential diagnosis

- Compression from outside, due to vascular malformations (aberrant innominate artery, double aortic arch, pulmonary artery sling)
- Compression from outside, due to cardiac malformations (left pulmonary artery, left atrium, right pulmonary artery)

Signs

- Expiratory stridor
- Respiratory distress

Preoperative work-up

- Respiratory function (blood gases, oxygen saturation monitoring)
- Direct flexible laryngo-bronchoscopy (with the child under local or general anesthetic, but breathing spontaneously)
- CT or MRI

Indication for operation

- Maintenance of the airway

Operation

- Tracheotomy with an extra long cannula to bypass the lesion
- Aortopexy
  - Left thoracotomy
  - Resection of the left thymus lobe
  - The aortic arch is sutured to the sternal periosteum with three sutures making several attachments to the aortic adventitia. The sutures are tied while the sternum is manually pressed towards the heart so that when they are released the anterior tracheal wall stretches
- Endoluminal stents (problematic because of difficulties with removing and changing the device, and with granulation tissue; in future perhaps resorbable stents could be the solution)

Prognosis

- Good in cases treated with simple aortopexy

16.5 Lung

16.5.1 Foreign Body Aspiration

General considerations

- Patients present aged between 1 and 3 years, most commonly at age 2 (toddlers)
- Aspirated material: peanuts, popcorn, seeds, pieces of meat, or other small objects
- Larger objects may lodge within the glottic opening (cause of death, especially in the group around the age of 1 year)
- Smaller objects are usually inhaled into the more distal branches of the airways (frequent location: right or left main bronchus, right and left bronchi of the lower lobes)
- Significant local inflammation usually starts 2–3 days after aspiration

Signs

- Coughing, wheezing, suprasternal retractions
- Respiratory distress, cyanosis
- In cases of minor aspiration, unspecific signs comparable to those seen in respiratory infections
- Pneumonitis after prolonged occlusion
- Atelectasis after complete occlusion
- Tracheal deviation, mediastinal shift

Preoperative work-up

- Full history (extraordinary episode of coughing, peanut ingestion)
- Chest X-ray with overinflation of the involved lung (air entrapment due to a ball-valve effect), possible mediastinal shift (Fig. 16.20)

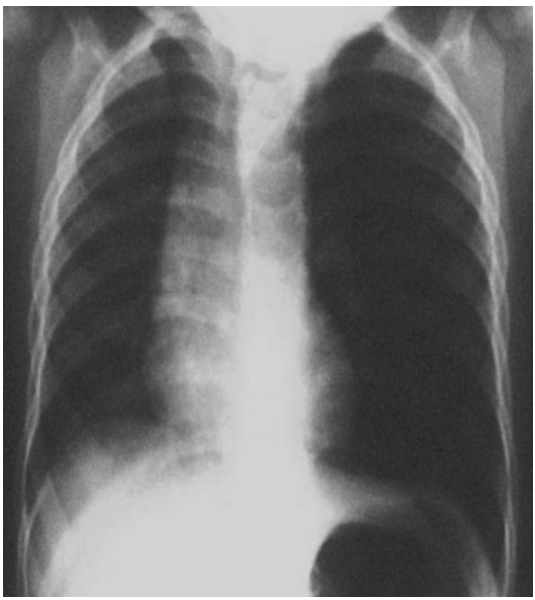


Fig. 16.20 Overinflated left lung due to aspiration

Therapy

- Heimlich maneuver in children older than 1 year (designed to force the diaphragm upward, which generates increased intrathoracic pressure) to dislodge a large foreign body from the larynx

Operation

- Rigid laryngo-bronchoscopy under general anesthesia, avoiding high ventilatory pressures which could drive the foreign body further into the airway
- Extraction of the foreign body (effective in 95% with a complication rate of less than 1%) with forceps, alligator forceps, peanut forceps, balloon catheter, basket forceps, suction and lavage catheters
- Very rarely thoracotomy, bronchotomy or sometimes localized lung resection is necessary

Postoperative care

- Plain chest X-ray
- In questionable cases repeated bronchoscopy

Prognosis

- Good, directly related to timely diagnosis and treatment

16.5.2 Shock Lung (Acute Respiratory Distress Syndrome)

Definition of acute respiratory failure

- Inadequate oxygenation (PaO_2 <55 mmHg, FiO_2 >0.5)
- Inadequate ventilation ($PaCO_2$ >50 mmHg)
- Acute lung injury (ALI): PaO_2/FiO_2 <200 mmHg
- Acute respiratory distress syndrome (ARDS): PaO_2/FiO_2 <300 mmHg, static compliance <40–50 $l \cdot cmH_2O^{-1}$

Classification

- Hypoxemic respiratory failure
  - Inadequate oxygenation (low or normal $PaCO_2$)
  - Acute lung injury
  - Systemic inflammatory reaction (SIRS)
  - Extracorporeal circulation
  - Shock of any cause, trauma
  - Systemic infection: sepsis
  - Pulmonary inflammation: pneumonia, inhalation injury, aspiration
  - Pulmonary infection: pneumonia
  - Lung contusion
- Hypercapnic respiratory failure (ventilatory failure)
  - Hypercapnia plus acute respiratory acidosis
  - $PaCO_2 = kV\dot{C}O_2/V_T\ (1 - V_D/V_T)$
  - CO_2 production ($V\dot{C}O_2$) $= V_A \times F_ACO_2$
  - Fractional concentration of CO_2 in the alveolar gas (F_ACO_2)
  - Alveolar volume (V_A) $= V\dot{C}O_2/F_ACO_2$
  - Tidal volume (V_T) $= V_D + V_A$
  - Minute ventilation ($= V_T \times RR$)
  - Dead space (V_D) $= V_E \times [(PaCO_2 - P_ECO_2)]/PaCO_2$
  - ↑ CO_2 production ($V\dot{C}O_2$): fever, sepsis, pain
  - ↑ V_D/V_T (increased V_D): ARDS, bronchoconstriction
  - ↓ Minute ventilation
  - Ventilatory pump dysfunction
  - Central respiratory drive (traumatic brain injury, sedatives, anesthetics, etc.)
  - Abnormal respiratory efferents (spinal cord injury, etc.)
  - Abnormal chest/abdominal wall: pleural fluid, ascites, scoliosis, etc.
  - Upper air way obstruction

Pathophysiology

- Diffuse alveolar injury
- Heterogeneous alveolar injury (different time constants)

- Alveolar consolidation (atelectasis, dependent lung regions, ↓ functional residual capacity)
- Alveolar overstretching (non-dependent regions)
- Shear trauma (between consolidated and overstretched lung areas)
- Pulmonary hypertension: vasoconstriction, microthrombi, obstruction of the airways with mucous that is then infiltrated with leukocytes to form a plug (leuko-plugging), interstitial edema
- ↓ Hypoxic pulmonary vasoconstriction: ventilation/perfusion mismatch
- ↑ Intrapulmonary shunting (no significant improvement with FiO_2 1.0)
- Endothelial dysfunction: mediator imbalance, inflammation, procoagulatory state
- Epithelial injury: surfactant deficiency, fluid and ion flux across the membrane
- Alveolar–capillary barrier lesion
- Bronchial obstruction (edema, secretions, terminal airway instability, spasm)
- ↑ Extravascular lung water (EVLW): permeability, decreased lymphatic flow
- ↓ Pulmonary compliance (surfactant dysfunction, edema, hyaline membranes) $C = \Delta V / \Delta P$
- ↓ Chest wall compliance (edema, injury)
- ↓ Abdominal wall compliance (abdominal compartment syndrome)
- Ventilator-induced lung injury (shear trauma, overstretching)
- Air leak (shear trauma, over distension)
- Fibroproliferative alveolitis

Signs

- Cardiorespiratory
  - Tachypnea, dyspnea, ↑ labored breathing
  - Pallor, cyanosis, stridor, retractions
  - Coarse lung crackles
  - Tachycardia, hemodynamic instability, poor skin perfusion

- Distant organ dysfunction (systemic inflammatory response)
  - Disseminated intravascular coagulation (DIC)
  - Encephalopathy (agitation, altered mental status)
  - Acute renal failure
  - Acute liver failure
  - Sepsis (gut: bacterial translocation, lung: ventilator-induced lung injury)
  - Hyperglycemia

Investigations

- Arterial blood gases (ABGs): hypoxemia, hypocapnia, acute respiratory alkalosis or acute metabolic acidosis
- Chest X-ray (bilateral infiltrates, pleural effusions)
- CT scan of the thorax, lung, abdomen
- Cultures (sputum, blood, effusion)
- Respiratory mechanics

Therapy

- Non-invasive correction of hypoxemia, hypercapnia, O_2 supplementation, CPAP, BiPAP (nasal airway, face mask)
- Mechanical ventilation
  - Open the lung and keep the lung open, inspiratory alveolar recruitment: plateau pressure (P_{pl}), tidal volume (V_T) expiratory alveolar recruitment: PEEP
  - Preventing lung injury: $V_T \leq 6$ ml·kg^{-1}, P_{pl} <3.5 kPa (36 cmH_2O) (prevents lung overinflation) V_T, peak inspiratory pressure (PIP) minimizing (overstretching, shear injury) PEEP optimization (early sufficient expiratory recruitment). IRV (inverse ratio ventilation, I:E > 1:1): increasing – inspiratory time (Ti), ↑ mean airway pressure (P mean), ↓ PIP, auto-PEEP
  - PCV (pressure control ventilation), PSV (pressure support ventilation) better than volume controlled MV

- Early spontaneous breathing (CPAP/ASB, BiPAP)
- FiO_2 reduction before pressure reduction PIP/PEEP (alveolar stability)
- Permissive hypercapnia ($PaCO_2$: 50–60 mmHg, arterial pH >7.25)
- Permissive hypoxemia (SpO_2: 88%–92%)

- High-frequency ventilation, early indication if FiO_2 >0.5/4 h on conventional mechanical ventilation (CMV), percussive (VDR4) or oscillating ventilation
- Kinetic therapy
- Selective pulmonary vasodilatation: inhalation of NO, prostacyclin
- Anti-inflammatory therapy
  - Prostacyclin inhalation
  - Steroids
  - Ibuprofen
- Antiproliferative therapy with steroids
  - Surfactant
  - Reduction in EVLW: negative fluid balance
  - β-agonist (bronchodilatation, ↑ interstitial and alveolar fluid transport)
  - Extracorporeal membrane oxygenation (ECMO) support if shunt >30%, FiO_2>60%, compliance <0.5 $ml \cdot cmH_2O^{-1} \cdot kg^{-1}$
  - Diagnosis and treatment of complications (air leak, pneumothorax)
- Supportive therapy
  - Hemodynamic optimization: oxygen delivery (DO_2) preload, cardiac output, SvO_2 (mixed venous oxygen saturation)
  - Drainage of pleural fluid, ascites (intra-abdominal pressure)
  - Bronchoscopy (lavage, source of bleeding)
  - Intestinal therapy
  - Early enteral feeding (bacterial translocation)
  - Stimulation of bowel motility
  - Closed tracheobronchial suction system (high respiratory support)
  - Nutritional support (early enteral feeding)

Prognosis

- In children better than adults
- Early mortality: multi-organ failure rather than lung failure, oxygen utilization defect not hypoxemia
- Long-term pulmonary dysfunction, broncho-pulmonary dysplasia (neonate), greater susceptibility to bronchial obstruction and airway infection

16.5.3 Lung Contusion

General considerations

- In children often without rib fracture
- Caused by severe shearing force and serial rib factures
- Frequently pneumothorax, hemothorax
- Additional injuries: abdomen, cervical spinal cord

Investigations

- Chest X-ray with a fluffy infiltrate that progresses in extent and density over a period of 24–48 h
- CT of the thorax is recommended early in the course to detect consolidation areas, injury of the lung and other organs
- Abdominal ultrasonography examination (liver or spleen ruptures, free abdominal fluid)
- Echocardiography: pericardial effusion, myocardial contractility, injury of great vessels

Therapy

- Early intubation and mechanical ventilation in cases of obvious respiratory insufficiency: SpO_2 <85% (PaO_2 <50 mmHg, $PaCO_2$ >50 mmHg) with FiO_2 0.21
- Temporary assisted high-frequency ventilation should be preferred

Operation

- Bronchoscopy: initially often without a result but in many cases helpful for guided pulmonary lavage and suctioning of blood plugs as well as defining the source of bleeding. Topical injection of surfactant
- Sufficient drainage: pneumothorax/hemothorax
- Continued or uncontrollable hemorrhage and/or massive air leak generally mandates an early thoracotomy

16.5.4 Lung Abscess

General considerations

- Result of a necrotizing pneumonia, e.g., after aspiration of gastric juice, gastroesophageal reflux
- Streptococci, *Staphylococcus aureus*, *Klebsiella pneumoniae*, *Pseudomonas aeruginosa*, or other Gram-negative enteric organisms
- Frequent in children with neurological impairment, seizure disorders, and immune suppression
- Location: posterior segment of the right upper lobe and the superior segments of the right and left lower lobe
- 10% of the affected children have more than one abscess
- Abscess formation related etiologically to an unknown pre-existing localized pulmonary malformation (congenital cystic adenomatoid malformation or CCAM, bronchogenic cyst, lung cyst, infected sequestration)

Signs

- Respiratory distress with tachypnea and cough
- Fever
- Decreased or no breath sounds and dullness to percussion on the involved side
- Pulmonary infiltrate, cavity with fluid level
- Quick development of pleural effusion
- Perforation into the pleural cavity leads to empyema usually combined with pneumothorax

Preoperative work-up

- Chest X-ray a.p. and lateral (Fig. 16.21)
- CT or MRI
- Bronchial lavage guided by flexible fiber optic bronchoscopy or thoracocentesis is useful for obtaining culture material used to determine specific antibiotic treatment

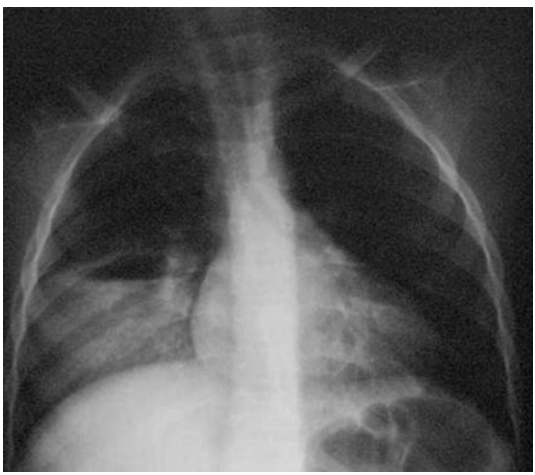


Fig. 16.21 X-ray findings in lung abscess

Indication for operation

- Bronchopleural fistula
- Operation frequently needed in younger and more debilitated children
- Large abscesses (approx. more than 5 cm in diameter) with fluid levels, especially located near the lung surface, unresponsive to aggressive conservative treatment
- No complete expansion of the lung over a period of 2 weeks
- Superfical lung abcess leading to partial pleura necrosis

Therapy

- Intravenous antibiotic management continued orally

Operation

- Care must be taken on anesthetic induction or when positioning the patient to prevent spillage of the abscess's contents into the contralateral lung (bronchoscopically guided suction)
- Closed drainage (multiple chest tubes sometimes necessary)
- Open drainage (including decortication) of the pleural effusion, usually with two large chest tubes
- Resection, usually of the complete involved lobe (covering the bronchial closure additionally with a dorsally based intercostal muscle flap)

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes decrease below 20–50 ml within 24 h
- Antibiotic management continued orally after discharge from hospital

Prognosis

- Good
- Resolution of a sufficiently drained abscess needs several weeks

16.5.5 Pneumatocele

General considerations

- Thin-walled, air-filled cyst usually after a necrotizing *Staphylococcus aureus* pneumonia (other germs involved: *Streptococcus*, *Hemophilus influenzae*, *Klebsiella*, *Escherichia coli* and *Pseudomonas*)
- Endotoxin released from the staphylococcal organisms contributes to the extremely destructive inflammatory process
- Mechanically ventilated patients are at increased risk of developing pneumatocele
- Adjacent structures may be compressed or a mediastinal shift may occur when a tension pneumatocele develops
- 25% of the pneumatoceles rupture, causing a, usually insignificant, pneumothorax

Signs

- Respiratory distress

Preoperative wok-up

- Chest X-ray
- CT scan
- Ultrasonography

Indication for operation

- Rapidly enlarging pneumatocele producing mediastinal shift (tension pneumatocele)

Therapy

- Most pneumatoceles require no treatment, only observation
- Percutaneous needle aspiration or chest tube for drainage of large cysts

Operation

- Thoracotomy, suture or resection is rarely necessary

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes decrease below 20–50 ml within a period of 24 h

Prognosis

- Good. About 50% resolve within 6 weeks and the remaining within 12 months

16.5.6 Chronic Atelectasis (Middle Lobe Syndrome)

General considerations

- Absent lung expansion over a period of at least 2–3 weeks due to bronchial stenosis (slit fashioned bronchi, malformation of the bronchial wall, compression from outside by enlarged lymph nodes or vascular anomalies) or intraluminal bronchial obstruction (foreign body)

Signs

- Asymptomatic at the beginning
- Recurrent signs of pulmonary infections
- Chronic cough

Preoperative work-up

- Chest X-ray (Fig. 16.22)
- CT scan
- Bronchoscopy
- Ventilatory and perfusion scintigraphy

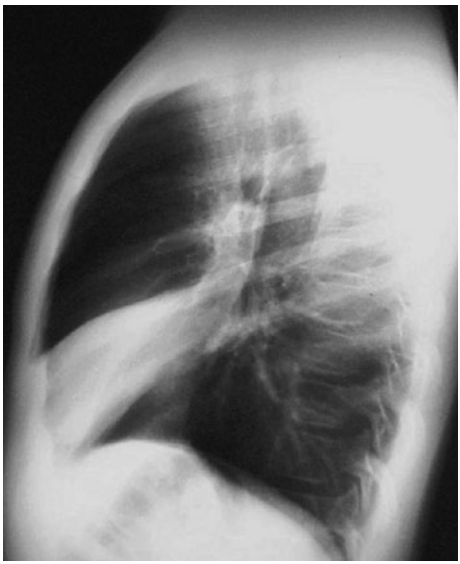


Fig. 16.22 Typical right middle lobe atelectasis

Indication for operation

- No improvement after intensive conservative treatment over a period of 8 weeks

Operation

- Middle lobe lobectomy

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes decrease below 20–50 ml within 24 h

Prognosis

- Good

16.5.7 Bronchiectasis

General considerations

- Low incidence in industrialized nations
- Permanent localized or diffuse abnormal dilatation (fusiform, cylindrical, saccular) of the segmental and subsegmental bronchi and their branches
- Principal areas of involvement: lower lobes most often affected, followed by the right middle lobe and the lingula
- Etiology: chronic suppurative lung disease, repeated pulmonary infections and poor clearance of lung secretions leading to destruction of the bronchial wall tissue with bronchomalacia and muscular hypertrophy. Bronchus obstruction may be worsened by compression due to enlarged lymph nodes
- Congenital bronchiectasis is rare (Klippel–Feil short neck, rib anomalies or Kartagener syndrome with ciliary dyskinesia, situs inversus, pansinusitis and bronchiectasis, infertility)
- Cystic fibrosis is the most common underlying disease causing bronchiectasis
- A foreign body may also cause local bronchiectasis

Signs

- Fever, cough, significant amounts of purulent sputum
- Physical activity (change of position) stimulates paroxysmal coughing
- Musical rales may be detected by auscultation
- Hemoptysis may occur
- Clubbing of the fingers in the late stages

Preoperative work-up

- High-resolution CT scan
- Bronchoscopy and biopsy to determine ciliary morphology
- Ventilatory/perfusion lung scans
- Investigation for cystic fibrosis

Indication for operation

- Recurrent lung infections in relation to localized bronchiectasis

Conservative therapy

- In early stages, conservative treatment (antibiotics, mucolytic, thorough pulmonary toilet)

Operation

- Lobe or segment resection. The operation may be difficult because of post infectious pleural adhesions and lymph node hypertrophy
- In patients with diffuse bronchiectasis lung resection may be required
- Cystic fibrosis of the lungs is the main indication for lung transplantation in childhood

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes decrease below 20–50 ml within 24 h

Prognosis

- Good
- Relapse may occur in patients with poor clearance of lung secretions

16.5.8 Bronchogenic Cysts

General considerations

- Arises from parenchymal cells that have been isolated during budding and branching to form a mass of non-functioning pulmonary tissue. Central cysts are of early embryonic origin. They are solitary and usually asymptomatic until infection occurs
- Located near the pulmonary hilum or in the mediastinum (near the esophagus)
- The lesions appear as solid masses or they are air filled (fluid level) when they communicate with the airways
- Rapid enlargement of a tension cyst may produce sudden respiratory distress
- Malignant transformation has been described

Signs

- Usually asymptomatic
- If the lesion becomes infected symptoms such as fever, hemoptysis, and cough with purulent secretions occur

Preoperative work-up

- X-ray
- High-resolution CT scan

Indication for operation

- The asymptomatic cyst is also a clear indication for excision

Operation

- Thoracotomy, cyst excision or lobectomy

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes decrease below 20–50 ml within 24 h

Prognosis

- Good

16.5.9 Pulmonary Cysts

General considerations

- Lesion develops between the 6th and 16th weeks of gestation. In contrast to the central (bronchogenic) cyst, they may be multiple and extensive
- Bronchial communication is more common
- A typical X-ray image is shown in Fig. 16.23

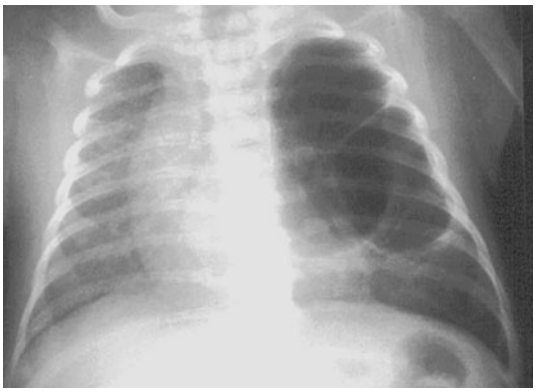


Fig. 16.23 Congenital pulmonary cysts with mediastinal shift

Differential diagnosis

- CCAM
- Congenital lobar emphysema
- Pneumothorax (sharp costophrenic angle)
- Congenital diaphragmatic hernia
- Arteriovenous lung aneurysm (Fig. 16.24)

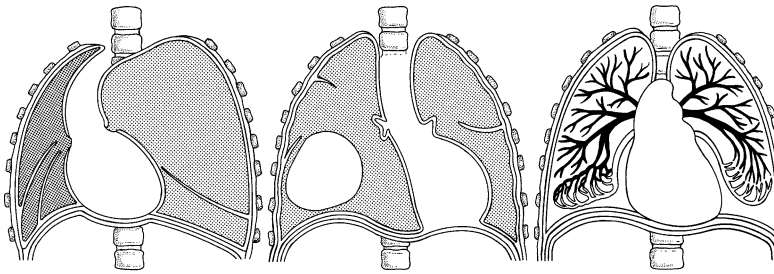


Fig. 16.24a–c Differential diagnosis of congenital lung malformations. **a** Congenital lobar emphysema, **b** pulmonary cysts, **c** arteriovenous lung aneurysm

Signs

- Respiratory distress shortly after birth

Preoperative work-up

- Chest X-ray
- CT scan

Indication for operation

- Clear indication for resection

Operation

- Thoracotomy, segmentectomy or lobectomy

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes decrease below 20–50 ml within 24 h

Prognosis

- Good

16.5.10 Congenital Cystic Adenomatoid Malformation (CCAM)

General considerations

- Non-cystic or multi-cystic mass of pulmonary tissue lined by cuboidal or columnar epithelium (sometimes skeletal muscle can be detected in the cyst wall). There is an overgrowth of bronchioles with suppression of alveolar development
- Usually only a single lobe is affected
- The lesion can enlarge because of fluid and air trapping. Over-distension of the involved lobe may lead to mediastinal shift, anasarca, hydramnion, hypoplasia of the ipsilateral and contralateral lung, fetal death, or respiratory distress after birth
- Spontaneous regression is possible
- Risk of pneumothorax, infection and malignant change (small risk)

Classification

- Classification of CCAM is given in Table 16.2

Table 16.2 Classification of CCAM

Type	Incidence	Description
I	50%–75%	Single or multiple cysts more than 2 cm in diameter, lined by ciliated pseudostratified columnar epithelium with thick smooth muscle and elastic tissue walls
II	10%–40%	Multiple small cysts less than 1 cm in diameter, lined by ciliated cuboidal or columnar epithelium. Respiratory bronchioles and distended alveoli may be present between these cysts. This type is frequently associated with other congenital anomalies such as renal agenesis, heart defects
III	10%	Non-cystic adenomatous solid mass. Poor prognosis

Signs

- May be asymptomatic
- Respiratory distress in types II and III

Preoperative work-up

- Prenatal serial ultrasonography examinations (detectable in the 12th to 14th gestational weeks)
- Repeated X-ray
- CT scan (Fig. 16.25) or MRI

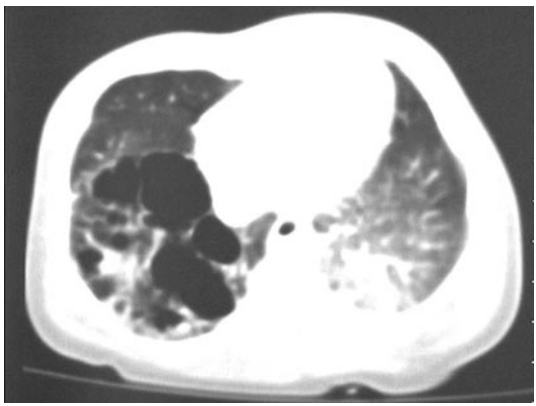


Fig. 16.25 CT scan of CCAM in the right lung

Indication for operation

- Fetal intervention (before the 32nd week) in severe cases with mediastinal shift: thoracocentesis of large cysts or resection of the lesion in utero
- Persistent (no signs of regression after 3 months of observation)
- Growing lesion, even if the patient is asymptomatic

Operation

- Thoracotomy and usually lobectomy within the first year of life

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes decrease below 20–50 ml within 24 h
- Problem of pulmonary hypertension and shunting may provoke ECMO (after successful resection) support

Prognosis

- Macrocystic lesions (>5 mm): good prognosis
- Microcystic lesions (<5 mm): poor prognosis

16.5.11 Congenital Lobar Emphysema

General considerations

- Postnatal overdistension of one or more lobes of a histologically normal lung, as a result of the collapse of bronchi
- Cartilaginous deficiency in the tracheobronchial tree with possible obstruction with air trapping or extrinsic pressure (anomalous pulmonary artery) on the airway or idiopathic causes
- The normal lobes are compressed and the mediastinum is shifted away from the affected side
- The upper lobes are more commonly involved, especially the left
- Males are more affected than females
- Associated anomalies (heart, kidney), in about 40% of the cases

Signs

- Respiratory failure with cyanosis
- Sometimes dramatic presentation due to overdistension of the affected lobe mimicking a tension pneumothorax

Preoperative work-up

- Chest X-ray
- CT scan
- Ventilation/perfusion lung scans

Indication for operation

- Conservative therapy is useless in congenital forms, which are a clear indication for resection

Operation

- Urgent lobe or segment resection
- Placement of a chest tube into emphysematous lobe can have catastrophic results (air leak, bleeding)
- In rare cases, the cause of the bronchial obstruction can be operatively treated

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes decrease below 20–50 ml within 24 h

Prognosis

- Good

16.5.12 Pulmonary Sequestration

General considerations

- Mass of non-functioning lung tissue that lacks an obvious communication with the tracheobronchial tree and receives its arterial blood from the systemic circulation. The venous drainage is either into the pulmonary vein (large shunts may develop >30% of cardiac output) or the azygos vein
- More than two-thirds receive their arterial blood supply from the abdominal aorta with the large vessel passing through the diaphragm in the aortic hiatus
- More than two-thirds are located in the lower lobes
- Two-thirds of the sequestrations are on the left side
- More than two-thirds have only one arterial supporting vessel
- Extralobar sequestrations in particular are associated with other abnormalities such as diaphragmatic hernia

Classification

- The interlobar sequestrations are classified according to the arterial blood supply and the ventilation pattern (Table 16.3)

Table 16.3 Pryce classification of interlobar sequestrations

Interlobar	The sequestration is located within the normal lung
Type I	Regularly ventilated lung tissue perfused by two arterial blood supplies (pulmonary artery, systemic artery)
Type II	Irregularly ventilated (atelectatic) lung tissue perfused by two arterial blood supplies on the margins (pulmonary artery, systemic artery)
Type III	Lung tissue not ventilated and perfused only by the systemic artery blood supply
Extralobar	
	The sequestration is completely separated as an accessory lobe from the normal lung (Rokitansky lobe). The arterial blood supply usually originates from the abdominal aorta, the venous blood usually returns into the azygos vein

Signs

- Interlobar sequestrations
  - Tend to provoke infections in the non-functioning lung tissue and the compressed surrounding lung
  - Children with unexplained recurrent pneumonias must be suspected of having sequestration
- Extralobar sequestrations (Fig. 16.26)
  - Small sequestrations may be completely asymptomatic
- Inter- and extralobar sequestrations
  - Hemothorax
  - Hemophthisis
  - Cyanosis
  - Clubbing of the fingers
  - Dyspnea

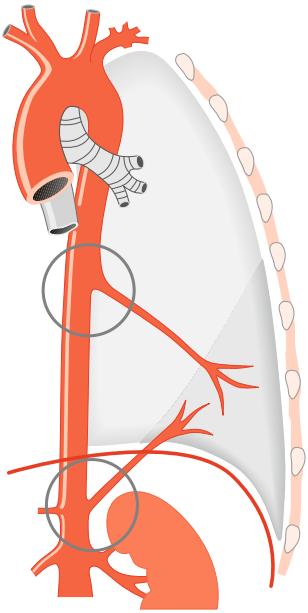


Fig. 16.26 Extralobar sequestration with blood supply from the thoracic or abdominal aorta

Preoperative work-up

- Prenatal ultrasonography
- The most specific diagnostic criterion for sequestration is the atypical artery, which is detected by high-resolution CT and CT angiography at the level of the diaphragm
- Doppler ultrasonography shows the abnormal arterial blood supply
- MRI
- Angiography

Indication for operation

- Always indicated as prophylaxis for recurrent pneumonias
- When recurrent pneumonias occur

Operation

- Lobe resection in cases of interlobar sequestration
- Sequester resection in cases of extralobar sequestration
- The transection of this transdiaphragmatic artery has to be performed meticulously as the ruptured vessel could retract into the abdomen causing massive bleeding in a compartment that is not very accessible. A splitting incision of the diaphragm is then necessary to access the abdomen and control the bleeding vessel
- In cases of obvious abdominal aorta vessel laparoscopic ligature of the vessel prior to resection is possible
- Thoracoscopic resection of extralobar sequesters is feasible

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes decrease below 20–50 ml within 24 h

Prognosis

- Good

16.5.13 Pulmonary Arteriovenous Aneurysm

General considerations

- Vascular malformation producing a high flow shunt between the pulmonary artery and the pulmonary vein
- High shunt volumes may lead to a hyperdynamic cardiac failure
- Location mainly in the right lung

Signs

- Cyanosis
- Clubbing of the fingers
- Polyglobulia
- Right heart insufficiency

Preoperative work-up

- CT angiography
- Angiography (Fig. 16.27) of the pulmonary artery
- Heart ultrasonography (increased cardiac output; hypertrophic or dilated ventricles)
- ECG (hypertrophy)

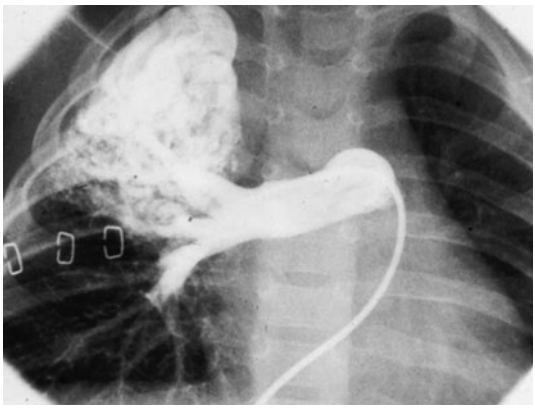


Fig. 16.27 Angiography of an arteriovenous lung aneurysm

Indication for operation

- Clear indication for resection on an urgent basis

Operation

- Thoracotomy with segmentectomy or lobectomy

Postoperative care

- Chest tubes may be removed if the lung is fully expanded and drainage volumes decrease below 20–50 ml within 24 h

Prognosis

- Good if cardiac insufficiency is reversible

16.6 Mediastinum

16.6.1 Patent Ductus Arteriosus

General considerations

- The ductus arteriosus is a vascular connection between the pulmonary artery and the aortic arch, a physiologic situation in the fetal circulation
- The duct should close within the first few days after birth
- If not closed an effective hemodynamic shunt between the pulmonary artery and the aorta is maintained

Signs

- High cardiac output failure
- Respiratory insufficiency, dependence on mechanical ventilation
- Elevated pulsatility index

Preoperative work-up

- Heart ultrasonography (increased cardiac output; hypertrophic right ventricle)

Indication for operation

- Hemodynamically effective shunt
- Exclude a shunt-dependent heart malformation
- Exclude a persistent pulmonary hypertension (PPHT)
- Thrombopenia (<50,000 mm^{-3}), recent bleeding
- Signs of renal insufficiency (urine volume <1 $ml \cdot kg^{-1} \cdot h^{-1}$) is a contraindication for conservative treatment with indomethacin or ibuprofen

Therapy

- Conservative treatment with indomethacin or ibuprofen

Operation

- Operative closure through a left lateral thoracotomy using a Hemoclip

Postoperative care

- A chest tube for lung expansion, gain hemostasis and drain secretions

Prognosis

- Good
- Increase of the mean arterial blood pressure

16.6.2 Mediastinitis

General considerations

- Bacterial infection of the mediastinum caused mainly by esophageal or tracheal trauma (perforations, anastomotic insufficiencies, deep caustic lesions due to lye ingestion, penetrating thoracic wounds)
- In cases of esophageal or tracheal perforation mediastinal emphysema is likely to develop
- Some patients develop pleural effusions

Signs

- High fever
- Elevated heart rate, in some cases venous inflow congestion
- Respiratory distress

Preoperative work-up

- Chest X-ray (widening of the mediastinum; thoracic effusion)
- Blood cultures
- Material from the mediastinum for bacterial culture should be obtained

Indication for operation

- Always in cases of fully developed mediastinitis

Operation

- Jugular median incision
- Blunt preparation
- Lavage with povidone-iodine solution
- More than one thick chest tube to drain the anterior mediastinum
- The middle and posterior mediastinum should be drained via a lateral posterior thoracotomy using the extrapleural approach
- More than one thick chest tube should be inserted
- Anastomotic insufficiencies are repaired by an intercostal muscle flap covering the defect
- Simple suture closure of distal recent esophageal perforations should be covered by a cuff on the gastric wall (intrathoracic partial fundoplication)
- In overlooked esophageal perforations (>72 h) esophageal resection and later replacement are usually indicated
- Artificial material in inflamed tissues is contraindicated

Postoperative care

- Aggressive treatment with antibiotics
- Repeated CT examinations to control the drainage and to detect new abscess formations

Prognosis

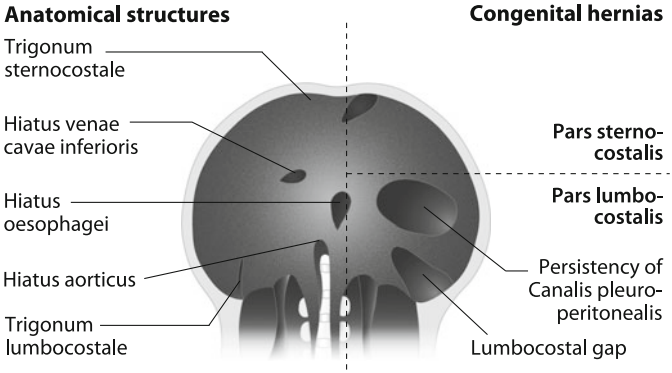
- Depends on the patient's constitution, the involved bacteria, and the distribution of the inflammation

17 Diaphragm

17.1 Diaphragmatic Hernia

General considerations

- Postero-lateral diaphragmatic defect (usually on the left side) (Fig. 17.1, Table 17.1) with communicating thoracic and abdominal spaces
- Generally accompanied by lung hypoplasia, which may be secondary to compression by the herniated viscera or a primary pulmonary defect. Both lungs are affected, particularly the one ipsilateral to the lung defect
- In left-sided hernias the stomach, the small bowel, and part of the colon are usually located in the thorax. The left lobe of the liver and the spleen may also be involved. In cases of dorsal aplasia of the diaphragm, the adrenal gland and even the kidney may be displaced into the thorax
- In right-sided hernias the liver and the small and large bowel may be located in the thorax
- Many associated malformations may be present in addition to lung hypoplasia: malrotation, cardiovascular defects, lung sequestration, and others



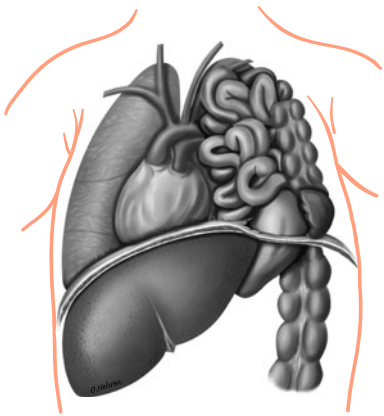


Fig. 17.1 Anatomy and pathology of the diaphragm

Table 17.1 Classification of diaphragmatic defects

Diaphragmatic eventration	▪ Hernia with sac ▪ Diaphragmatic relaxation ▪ Phrenic palsy
Diaphragmatic hernia (Bochdalek)	Lumbocostal hernia, often left sided
Hernia of Morgagni	Sternocostal hernia

Signs

- Severe dyspnea and cyanosis after birth
- Hypoventilation of the ipsilateral side
- Paradoxical movements, with displacement of the abdominal contents into the thorax during inspiration
- Displacement of the mediastinum to the contralateral side

Differential diagnosis

- Pneumothorax
- Congenital lung cysts

Antenatal diagnosis

- Prenatal diagnosis is possible upon screening ultrasonography (in these cases the mother should be referred to centers in which adequate neonatal treatment is possible)
- Ultrasonographic lung biometry
  - Lung-to-head ratio (LHR)
    LHR = lung diameter (width) (mm) × lung diameter (length) (mm)/head circumference (mm)
  - 95% survival if LHR >1.4
  - 75% mortality if LHR <1
- Prenatal detection of pulmonary hypoplasia using ultrasonographic lung biometry is possible, however the prediction of its severity is not possible

Preoperative work-up

- Chest X-ray
- Thoraco-abdominal ultrasonography
- Stabilization of respiratory function for several hours to some days is advisable before surgical treatment
- Neonatal intubation and adequate oxygenation
- Nasogastric tube for gastric emptying

- Ventilatory assistance with pressures below 20–25 cmH_2O, PEEP of 0–1 cmH_2O, frequency of 40–60 breaths per minute and *I:E* of 1:1
- Oscillatory ventilation if required
- Arterial and venous pressure monitoring
- Pulse oximetry
- Transcutaneous preductal and postductal PaO_2 monitoring
- Correction of acidosis
- ECMO, if required

Operation

- Positioning on a warm blanket with elevation of the thoraco-lumbar region
- Left subcostal or median laparotomy
- Careful hemostasis particularly when under extracorporeal membrane oxygenation (ECMO)
- Careful repositioning of the herniated viscera into the abdomen paying particular attention to the liver and spleen
- Definition and dissection of the borders of the orifice. This is particularly necessary in the posterior rim, which is usually narrow
- Primary closure, when possible, with interrupted stitches
- If primary closure is not possible, a prosthetic patch can be used. Gore-Tex® is a good prosthesis
- Revision of the abdomen and correction, if possible, of associated intraabdominal malformations
- No pleural drain is necessary, except in those cases where there is a pneumothorax. A balanced drain is advisable to prevent excessive negative pleural pressure
- The operation is illustrated in Fig. 17.2

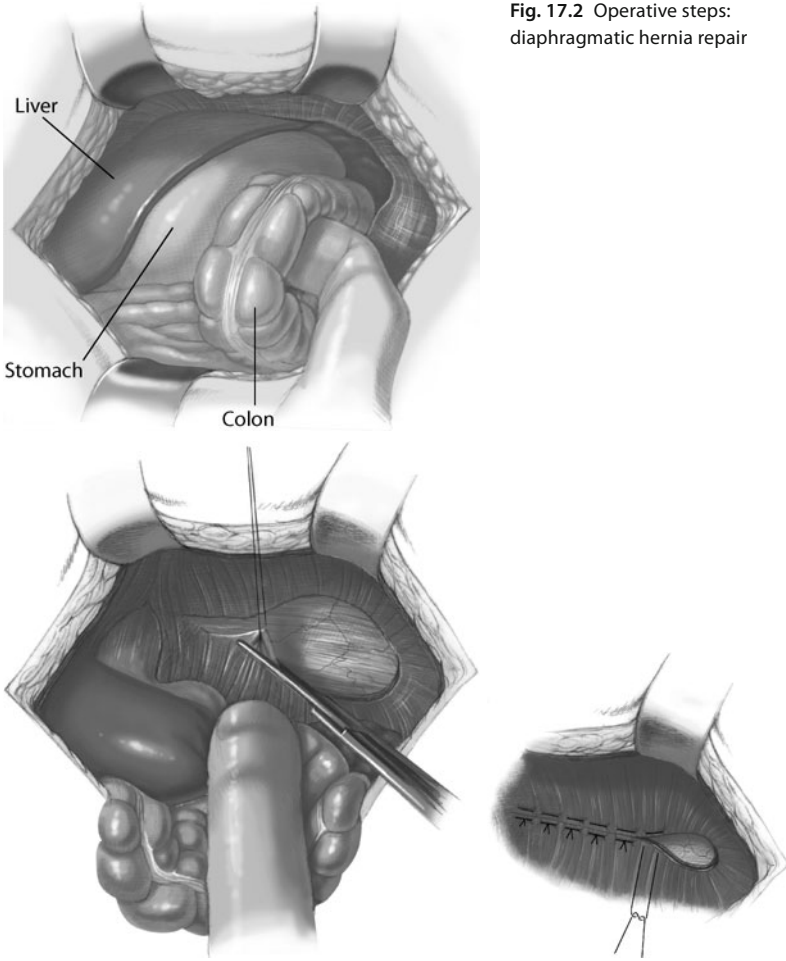


Fig. 17.2 Operative steps: diaphragmatic hernia repair

Postoperative care

- Similar to the preoperative measures taken during stabilization
- Adequate analgesia should be provided
- As soon as possible: transition to spontaneous respiration with a period of CPAP in between

Prognosis

- Mortality remains between 20% and 50%
- The prognosis is better when the signs appear 6 h or more after birth
- ECMO may modify the prognosis in worst cases

17.2 Fetoscopic Approach (FETENDO)

- Temporary fetal tracheal occlusion reverses the pulmonary hypoplasia seen in congenital diaphragmatic hernia (CDH) and provides an alternative treatment strategy for some fetuses with CDH by distending the hypoplastic lungs
- The safety and efficacy of the following methods are being tested in ongoing trials. Only those fetuses most severely affected, with liver herniated into the chest, a diagnosis before 24 weeks, and a ratio of lung to head of less than 1.4, are eligible for this treatment. Less severely affected fetuses are best managed after birth

PLUG (Plug the Lung Until it Grows)

- This fetoscopic approach uses small scopes and video equipment to place a detachable balloon by fetal bronchoscopy, without opening the uterus
- The balloon occludes the trachea and enables the lungs to grow. Although the lungs grow in size the lung function is not necessarily increased

EXIT (Ex utero Intrapartum Treatment)

- This procedure is used to reverse tracheal occlusion devices in fetuses with severe CDH who have been treated with the PLUG procedure. Carried out before the umbilical cord is severed, this procedure offers the advantage of ensuring uteroplacental gas exchange while the fetus is still receiving placental support
- For this procedure a well-trained interdisciplinary team is needed

17.3 Extracorporeal Membrane Oxygenation

General considerations

- Cases with a prenatal diagnosis of CDH should be referred to centers in which optimal care including ECMO and neonatal surgery is possible
- ECMO is performed under complete heparinization, with the consequent bleeding risks
- It consists of an veno-arterial bypass with membrane oxygenation

Operation: Steps shown in Fig. 17.3

- Position the patient with the neck hyperextended and rotated to the left
- Make a transverse incision 1 cm above the clavicle
- Dissect the carotid artery, internal jugular vein, and vagus nerve
- Open the artery first, avoiding dissection of the intima
- Introduce a Ch 10 arterial cannula for 2.5 cm towards the brachiocephalic trunk
- Introduce a Ch 12–24 venous cannula for 6 cm towards the right atrium
- Both cannulas are fixed with double ligatures and additionally sutured to the skin

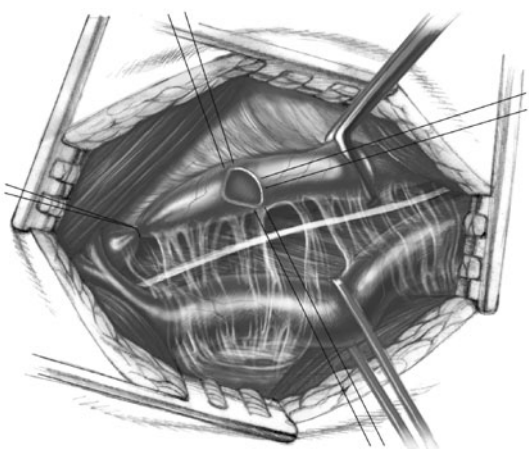


Fig. 17.3 Operative steps: vessel cannulation for ECMO

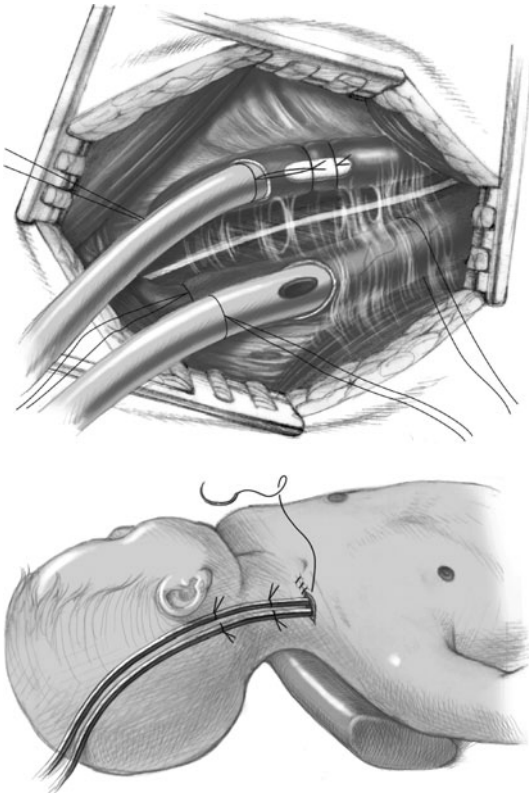


Fig. 17.3 *(continued)* Operative steps: vessel cannulation for ECMO

17.4 Diaphragmatic Eventration

General considerations

- X-ray diagnosis of a high-positioned diaphragm
- Usually found in patients who have had heart operations (phrenic nerve palsy)
- Only some patients have signs

Classification

- Diaphragmatic eventration classification is given in Table 17.2

Table 17.2 Diaphragmatic eventration classification

Diaphragmatic hernia with sac	There is no real communication between the abdominal and the thoracic spaces because of the interposition of a peritoneal sac
Diaphragmatic relaxation	The diaphragmatic tissue is hypoplastic mostly on the left side Diagnosis with the former condition is only histological
Phrenic palsy	Relaxation due to phrenic nerve palsy. Often due to traumatic birth delivery, more often right-sided

Signs

- In newborns
  - In severe cases it may be identical to a diaphragmatic hernia, but this is rare
- Beyond the newborn period
  - Respiratory difficulty and failure to thrive
  - Small abdomen
  - Hydroaeric bruits in the thorax
  - Rarely incarceration and intestinal obstruction

Preoperative work-up

- Chest X-ray
- History: possible thoracic or heart surgery (phrenic palsy)
- Diaphragmatic immobility upon fluoroscopy
- Diaphragmatic immobility upon ultrasonography
- Ultrasonographic detection of heart defects

Operation

- Subcostal laparotomy and mobilization of the liver or stomach and spleen
- Plication (Fig. 17.4) with several U-shaped sutures avoiding the phrenic nerve
- Alternative
  - Postero-lateral low thoracotomy (8th or 9th space)
  - Plication from above avoiding the phrenic nerve with pericostal knotting
  - No pleural drains left

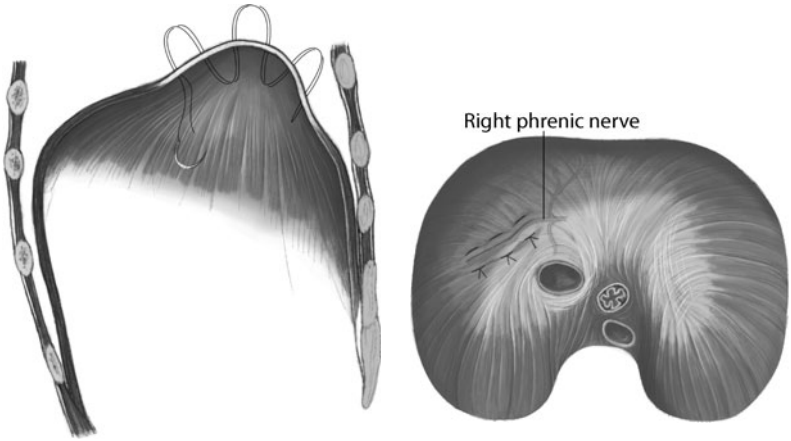


Fig. 17.4 Diaphragmatic plication

Postoperative care

- Spontaneous breathing as soon as possible

Prognosis

- Excellent for unilateral relaxations
- Bilateral ones may have a worse prognosis

17.5 Gastroesophageal Reflux/ Paraesophageal Hernia

General considerations

- Gastroesophageal reflux (GER) is a normal event but may become pathological when too severe or too frequent
- 90% of infants with GER improve spontaneously before they are 1 year old

- GER is particularly frequent after esophageal atresia, diaphragmatic hernia, anterior abdominal wall defects, and malrotation repair, or in children with brain damage of various origins who are mentally retarded
- GER may be accompanied by hiatal hernia with prolapse of the stomach in the thorax

Classification

- Gastroesophageal reflux/paraesophageal hernia classification is given in Table 17.3

Table 17.3 Classification of gastroesophageal reflux/paraesophageal hernia

Paraesophageal hernia	The gastroesophageal junction is in the abdomen but the fundus and part of the stomach pass into the mediastinum through the hiatus
Upside-down stomach	The stomach is in the mediastinum with the antrum above the fundus
Mixed forms	Several combinations

Anti-reflux mechanisms

- Anatomic
  - The lower esophageal sphincter
  - The length of the intra-abdominal esophagus
  - The angle of His (Fig. 17.5)
  - The lower esophageal mucosal rosette
  - The diaphragmatic crural sling
- Physiologic
  - The permanent tone of the sphincter's smooth muscle
  - Contraction of the diaphragmatic crural sling on inspiration
  - Esophageal peristalsis
  - Normal gastric emptying
  - Salivary and esophageal gland secretion
  - Mucosal regeneration

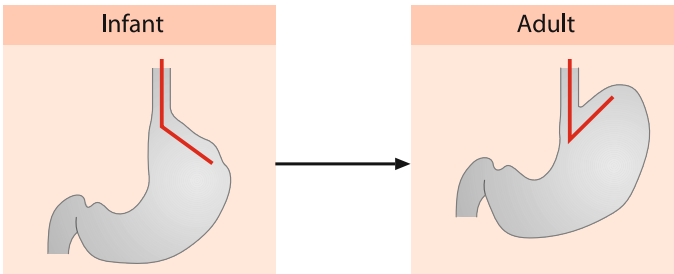


Fig. 17.5 Angle of His

Pathophysiology of GER

reversible

Reflux from the stomach → Mucosal erythema → Vulnerable mucosa

↓

Dysmotility ← Ulcerative esophagitis

↓

Barrett's esophagus

irreversible

Signs

- Vomiting
- Aspiration
- Recurrent pneumonia
- Heartburn
- Melena
- Iron deficiency anemia
- Dysphagia
- Failure to thrive
- Malnutrition
- Stridor

Investigations

- Barium meal may be useful for GER detection and is definitely useful for identifying stenosis, malrotation or delayed gastric emptying
- Scintigraphy is helpful for assessing esophageal clearance and/or aspiration and particularly for evaluating gastric emptying
- Ultrasonography, including color Doppler, is increasingly used for reflux detection and assessment of the pylorus
- 24-h pH-metry is the best way of quantifying reflux (pH <4 is pathologic if maintained for more than 4% of the time but also if long episodes occur, particularly during the night)
- Esophagoscopy and biopsy show esophagitis
  - Grade I – mucosal erythema
  - Grade II – contact bleeding
  - Grade III – ulceration
  - Grade IV– stricture
- Esophageal manometry (absence of pressure at the lower sphincter or non-regulatory relaxations) is a complicated procedure used only to investigate particular conditions
- Detection of occult blood in stool is sometimes helpful
- Sideremia

Conservative treatment

- Upright positioning at 30°
- Frequent and small-volume feeds
- Antacids (adsorbent)
- H_2-receptor blockers (cimetidine, ranitidine)
- Prokinetics (domperidone, metoclopramide)
- Avoidance of theophylline, prostaglandins, dopamine, diazepam, anticholinergics, antiadrenergics, and chocolate

Indication for operation

- Esophageal stenosis or Barrett esophagus
- Failure of conservative treatment after a trial of several months
- Anatomic anomalies (paraesophageal hernia)

- Previous operations for malformations such as esophageal atresia, CDH, and malrotation with symptomatic GER
- Mentally retarded children with symptomatic GER
- Infants with refractory failure to thrive and GER
- GER-associated respiratory tract disease with long episodes of reflux during the night detected upon pH-metry

Operation (Nissen fundoplication)

- Median supraumbilical laparotomy or laparoscopic approach (Fig. 17.6; the same steps are required for both operations)
- Mobilization of the terminal esophagus and the hiatus (Fig. 17.7)
- Mobilization of the fundus by section of two or three short vessels
- Insertion of a very thick catheter or dilatator into the esophagus
- Closure of the hiatus with two to three non-absorbable sutures
- Passage of the fundus behind the esophagus and creation of a Nissen wrap secured with three non-absorbable stitches that must attach substantially to the terminal esophagus
- The hiatal repair and the wrap should be loose enough to avoid dysphagia and gas bloat
- Gastrostomy may also be performed when operating on mentally retarded children

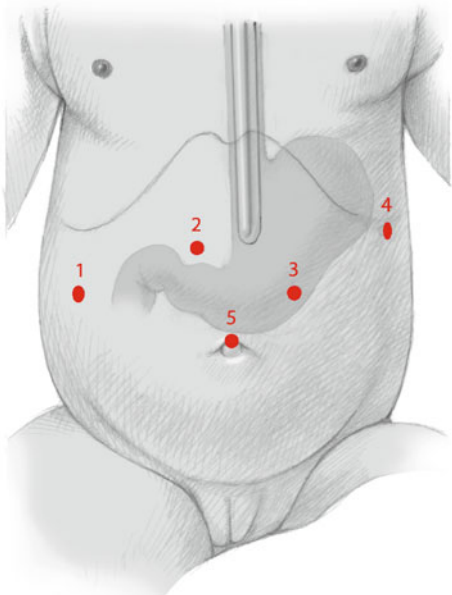


Fig. 17.6 Trocar position for laparoscopic fundoplication

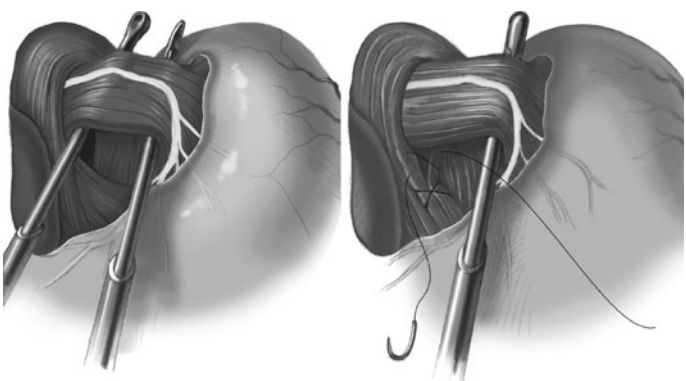


Fig. 17.7 Operative steps for Nissen fundoplication

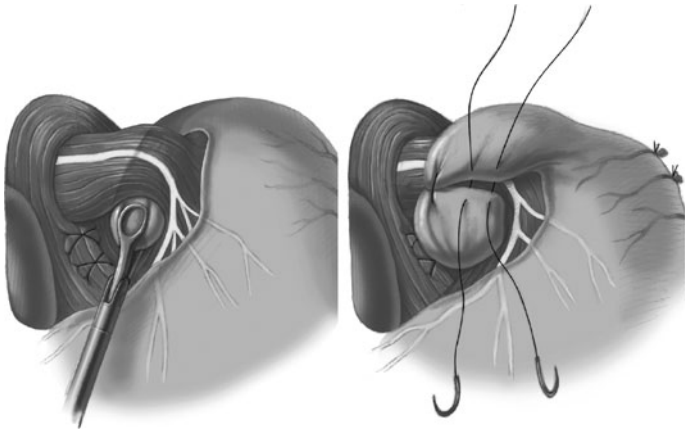


Fig. 17.7 *(continued)* Operative steps for Nissen fundoplication

Operation (Semifundoplication)

- The first four steps are the same as for a Nissen fundoplication
- Closure of the hiatus with two to three non-absorbable sutures
- Fixation of the esophagus to the hiatus in three quadrants (right, left, and ventrally)
- Suture of the fundus to the hiatus on the right side of the esophagus
- Mobilization of the fundus in order to cover the esophagus ventrally
- Suturing of the fundus and the right side of the esophagus
- Suturing of the fundus to the diaphragm with several stitches on the left

Postoperative care

- Nasogastric tube until recovery of transit
- Parenteral feeding from 2nd day until feeds are started
- Small and frequent feeds at first
- Discharge when ambulatory and feeding
- Soft diet for 2 weeks

Complications

- Recurrent GER after wrap failure
- Ascent of the wrap into the thorax due to failure of crural repair
- Stenosis of the wrap
- Gas bloat syndrome (usually temporary)
- Dumping syndrome

Prognosis

- Non-operative cure in about 95% of cases presenting in infancy
- Operative success in about 90% of cases except in patients who have had previous operations for esophageal atresia and in brain-damaged individuals for whom the recurrence rate is 25%

17.6 Traumatic Diaphragm Rupture

General information

- Occurs after blunt trauma with high intraabdominal pressure
- Left-sided in 70% of cases
- The stomach may prolapse into the thorax; the colon, small bowel, spleen or liver may also be involved
- Associated injuries of the liver, spleen, and intestine

Signs

- Shock
- Dyspnea
- Cyanosis
- Pain

Preoperative diagnostics

- Chest and abdominal X-ray
- Ultrasonography
- CT scan

Operation

- Abdominal approach
- Search for associated visceral lesions
- Primary repair of the diaphragm
- If spleen lesions, splenorrhaphy when possible
- Underwater chest tube

Postoperative care

- Withdraw the pleural tube as soon as possible
- Analgesia
- Monitoring for at least 24 h
- Discharge thereafter as soon as possible

Prognosis

- Good

18 Esophagus

18.1 Esophageal Stenosis

General considerations

- Esophageal stenosis can be congenital or acquired
- Congenital esophageal stenosis is either related to abnormal differentiation during the development of the esophagus and the trachea or is due to compression from neighboring structures
- Corrosive esophageal stenosis is common in young children who put everything into their mouth
- Postoperative stenosis is also common after surgical procedures for congenital anomalies

Classification

- The classification of esophageal stenosis is outlined in Table 18.1; the differential diagnoses are shown in Fig. 18.1

Table 18.1 Classification of esophageal stenosis

Congenital			
Intrinsic		**Extrinsic**	
Segmental fibromuscular hypertrophy	Segmental hypertrophy of the muscular and submucosal layers affecting mainly the distal third of the esophagus	Abnormally located pulmonary artery	External compression by the pulmonary artery affecting mainly the upper third of the esophagus
Ectopic tracheobronchial remnants	Tracheobronchial remnants sequestered into the esophageal wall affecting mainly the middle third of the esophagus	Double aortic arch	External compression by the pulmonary artery affecting mainly the upper third of the esophagus
Membranous web	Abnormal folds in the esophageal mucosa forming a diaphragm affecting mainly the middle third of the esophagus		
Achalasia	Absence of normal esophageal peristalsis with failure of complete relaxation of the lower sphincter		
		Acquired	
		Postoperative	Mainly after esophageal atresia repair
		Corrosive	
		Peptic stenosis	Residual stenosis after gastroesophageal reflux mainly affecting the lower third of the esophagus
		Barrett esophagus	Transformation of the esophageal mucosa with stenosis; pre-cancerous
		Erosion	Accidental ingestion of corrosive substances resulting in extreme morbidity, mostly in children <5 years old

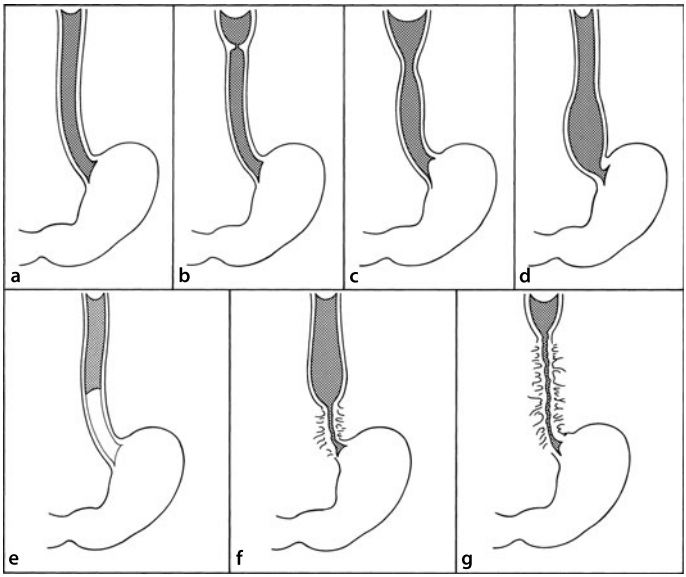


Fig. 18.1a–g Differential diagnoses of esophageal stenosis. **a** Normal finding, **b** membrane stenosis, **c** hourglass stenosis, **d** achalasia, **e** Barrett's esophagus, **f** peptic stenosis after reflux, **g** caustic stenosis

Signs

- Rarely during the neonatal period. Progressive dysphagia and vomiting of undigested food after introduction of semisolid or solid food at about the age of 6–7 months
- Accidental ingestion of a foreign body causing dysphagia can lead to the diagnosis
- Failure to thrive
- Aspiration
- Respiratory signs such as wheezing and stridor

Investigations

- The goal of diagnostic examinations is to recognize the correct etiological factor for this stricture
- Contrast esophagogram
- Esophagoscopy with biopsy (mostly normal mucosa overlying the stenosis)
- pH-metry
- Echocardiography in some cases
- See also below for each condition

Therapy

- Initially managed by hydrostatic or pneumatic balloon dilatation, which have a radial expanding effect (see also section 18.6)

Operation

- Resistant stricture requires resection and reanastomosis
- Esophageal replacement with colon or stomach is also possible
- See also methods in Table 18.2 for each condition

Table 18.2 Esophageal anomalies and their surgical repair

Anomaly	Operation
Segmental fibromuscular hypertrophy	Esophageal segmental resection and reanastomosis
Ectopic tracheobronchial remnants	Remnant resection and esophageal closure
Membranous web	Balloon dilatation or laser coagulation or resection
Peptic stenosis	Longitudinal incision with transverse closure Esophageal segmental resection and reanastomosis
Vessel anomalies	Vessel ring resection and vessel reanastomosis

Prognosis

- Depends on the situation and the underlying disease

18.2 Corrosive Esophagitis

General considerations

- Corrosive damage depends on the type, amount, concentration, and pH of the chemical agent involved
- 90% follow ingestion of strong alkali found in household detergents
- 10% are related to acid ingestion (coagulation necrosis)
- Liquid alkaline substances cause deep and penetrating liquefacient necrosis in the esophagus (coagulation necrosis)
- In 50% of cases the stomach is also affected
- Mineral acids produce coagulation necrosis usually causing superficial damage. However, ingestion of highly concentrated acid produces similar severe damage to the esophageal mucosa in approximately 40% of cases

Classification

- Based on endoscopic findings the esophageal injury is classified in four degrees (Table 18.3)
- However, the initial endoscopic distinctions are not always precise and predictive
- When muscular layers are involved, esophageal injury results in grade III and IV burns with early and late consequences defined as a corrosive disease

Table 18.3 Esophageal injury classification

Grade		Endoscopic findings
I		Erythema and edema
II	a	Erosions, superficial ulceration, hemorrhagic exudation
	b	Circumferential lesions
III		Multiple ulcers and superficial necrosis, overlying dense fibrin
IV		Deep necrosis with perforation

Phases of the corrosive disease (Table 18.4)

- Edema and deep ulceration
- Vessel thromboses
- Development of necrotic tissue
- Bacterial invasion

Table 18.4 Phases of esophageal injury

Phase	Anatomic correlate	Time
Acute phase	Local inflammation with infiltration by polynuclear cells	Immediately
Subacute phase	Demarcation and sloughing of the necrotic tissue and formation of granulation tissue Fibrotic proliferation and deposition of collagen	First week
Chronic phase	Scarring process with formation of esophageal stricture	>3 weeks

Signs

- Possible external perioral signs
- Signs ranging from minimal ones to manifest shock
- Salivation, spitting
- Coughing
- Painful swallowing
- Chest pain
- Dysphagia
- Respiratory distress
- Epigastric pain (grades III, IV)
- Retching (grades III, IV)
- Hematemesis (grades III, IV)

Investigations

- Diagnostic esophagogastroscopy within the first 48 h (evaluation of the extent and degree of the burn or exclude esophageal injury)
- When esophagogastroscopy is not possible, diagnostic laparotomy and revision of stomach and cardia should be performed

Therapy

- Treat shock
- Nasogastric tube
- No stomach irrigation
- No emetic medication
- Management is outlined in Table 18.5

Table 18.5 Management of corrosive esophagitis

Grade	Control endoscopy	Findings	Therapy	Discharge
I	3–4 days	Restitution	Symptomatic	1 week
IIa	3–4 days	If no evidence of stenosis	Symptomatic	2 weeks
IIb	3–4 days 3 weeks	If esophageal stenosis	Prograde bougienage every 7–10 days for 2–6 months	3 weeks
III	3–4 days 3 weeks	Esophageal stenosis	Parenteral nutrition, gastrostomy, retrograde dilatation	3–4 weeks
IV	3–4 days 3 weeks	Secondary gastro-esophageal reflux due to fibrotic shortening of the esophagus	Parenteral nutrition, gastrostomy, retrograde dilatation, antireflux surgery, esophagectomy	3–5 weeks

Operation

- If regular esophageal dilatation does not lead to the resolution of a stricture within a year, esophageal replacement is required
- Isoperistaltic left colon transposition, via a retrosternal or a transhiatal route, with one-stage esophagectomy is the method of choice
- Gastric or gastric tube interposition is another option

Complications

- Scarring of the esophagus is the most common (ca. 30%) sequela of the corrosive injury
- Aspiration pneumonia and sepsis are serious early complications (within 72 h of injury)
- Esophageal or gastric perforation followed by mediastinitis and peritonitis requiring surgery
- Delayed longitudinal scarring and shortening of the esophagus
- Reflux esophagitis

Prognosis

- Depends on the grade and complications
- Long therapy

18.3 Achalasia

General considerations

- Motility disorder of the distal esophagus
- Absence of normal esophageal peristalsis with failure of complete relaxation of the lower sphincter
- Reduced neuropeptides in the esophageal muscle layers suspected
- Decrease or loss of myenteric ganglion cells
- Fewer than 5% of all cases present before the age of 15

Signs

- Dysphagia
- Vomiting and regurgitation of undigested food
- Weight loss
- Failure to thrive
- Retrosternal discomfort
- Pulmonary problems due to microaspirations

Investigations

- Chest X-ray with air/fluid level in the esophagus
- Contrast swallow with the typical rat tail deformity (dilated esophagus and a smooth tapering of the esophagogastric junction)
- Endoscopy (retained food in the esophagus)
- Esophageal manometry with pressures >45 mmHg and uncoordinated peristalsis (gold standard)

Therapy

- Calcium channel blockers
- Intrasphincteric injection of botulinum toxin
- Dilatation
- Operation if conservative therapy fails

Preoperative work-up

- Patient on clear fluids for 2 days prior to operation
- Endoscopy prior to operation to check that the esophagus is completely empty
- Large feeding tube in the stomach
- Antibiotic prophylaxis

Operation

- Heller's esophagocardiomyotomy (Fig. 18.2)
- Via the abdomen or the thorax as an open procedure or by using a minimally invasive approach
- Mobilization of the gastric fundus and the distal esophagus
- The abdominal esophagus is freed by blunt dissection into the posterior mediastinum
- Identification of the anterior vagus
- With simultaneous gastroscopy, dissect the esophagus muscles 4 cm above and 2 cm below the stenosis into the gastric wall
- A concomitant fundoplication is recommended as a posterior Toupet procedure or as an anterior Thal procedure (see also section 17.5)

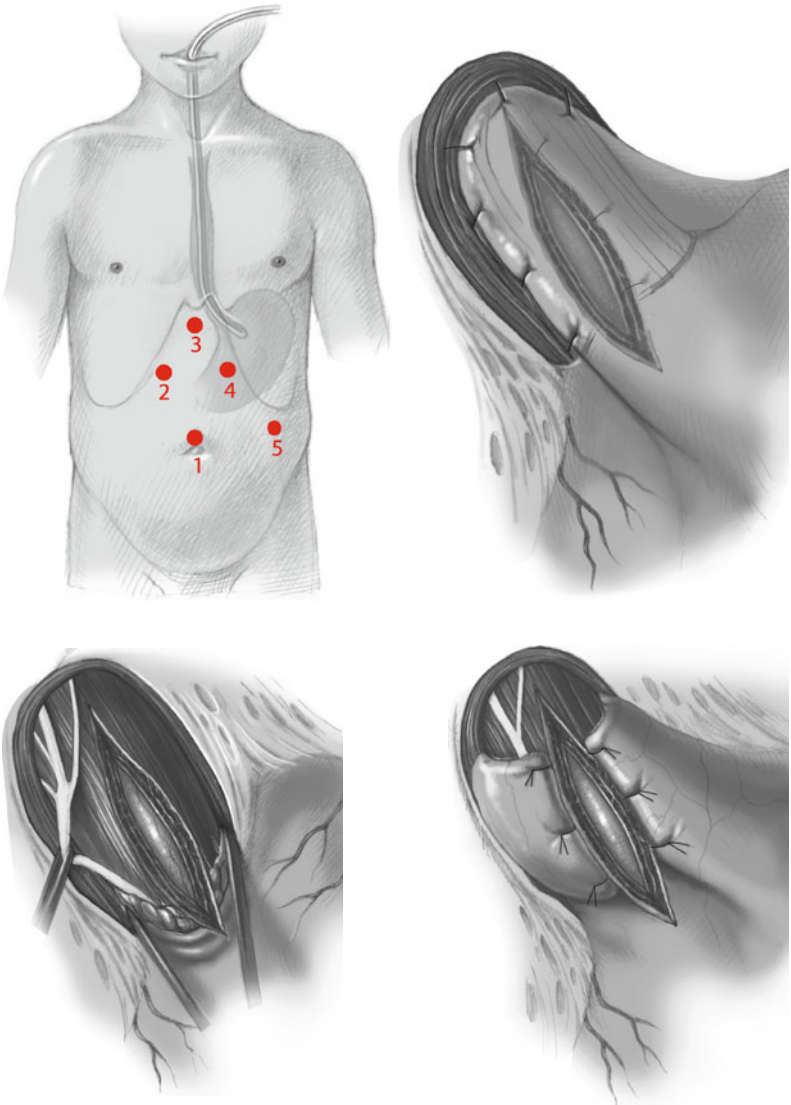


Fig. 18.2 Trocar position and operative steps

Postoperative care

- Nasogastric tube for 2–3 days
- Antibiotics for 24 h

Prognosis

- About 50% recur
- Good if no recurrence

18.4 Esophageal Atresia

General considerations

- Interruption in the continuity of the esophagus with or without tracheal fistula
- Esophageal atresia is related to abnormal differentiation of the gastrointestinal and respiratory tracts
- The existence of familial cases with affected offspring and siblings implies the influence of genetic factors
- The incidence is about 1:3000 births with a slight male predominance
- Other associated malformations occur in about 50% of cases
- It is presumed to be part of a broad spectrum of anomalies associated with syndromes

Classification

- Different forms
- The most common form is the blind upper pouch with a tracheo-esophageal fistula
- Type I–IIIc described by Vogt
- Type A–E described by Gross
- Classification is detailed in Table 18.6 and Fig. 18.3

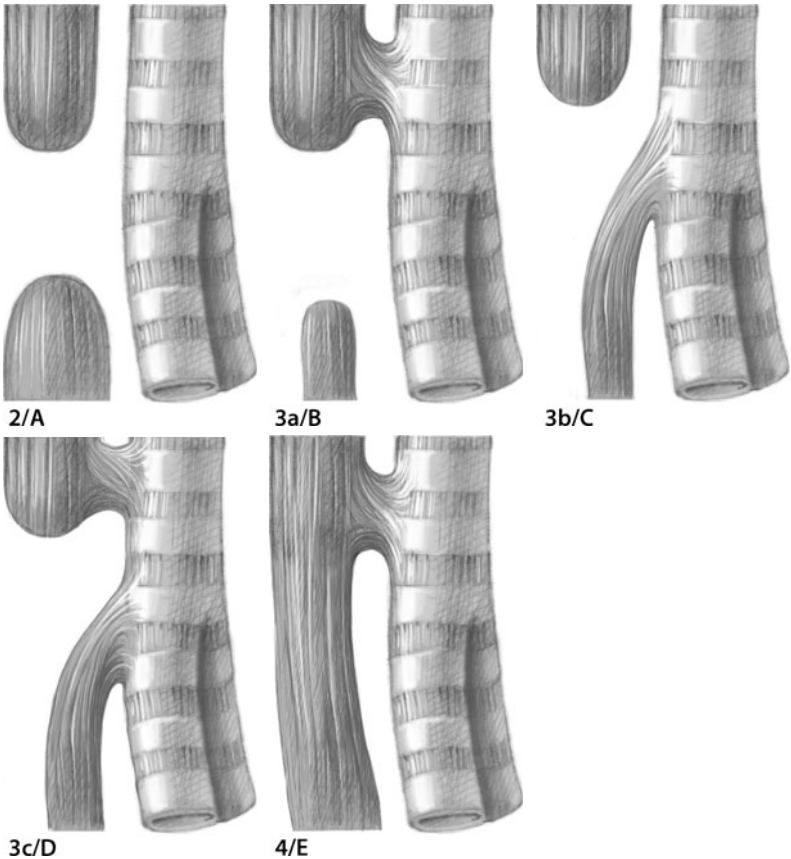


Fig. 18.3 Esophageal atresia classification (Vogt/Gross)

Table 18.6 Classification of esophageal atresia. Types I–IIIc, Vogt; types A–E, Gross

I		II		IIIa		IIIb		IIIc		H-fistula
		A		B		C		D		E
Proximal	Distal	Proximal	Distal	Proximal	Distal	Proximal	Distal	Proximal	Distal	
Aplasia	Aplasia	Blind	Blind	Fistula	Blind	Blind	Fistula	Fistula	Fistula	
Rare		6%–8%		2%		85%		3%		3%–4%

Table 18.7 Treatment approaches for esophageal atresia

Form	Proximal esophagus	Distal esophagus	First option	Second option
A	Aspiration has to be avoided	Feeding should be possible	Primary esophago-esophagostomy	
B	Fistula closure is necessary to avoid aspiration	Aspiration has to be avoided	Primary esophago-esophagostomy	Gastrostomy and cervical esophagostomy
C	Aspiration has to be avoided Feeding should be possible	Feeding should be possible Fistula closure is necessary to avoid aspiration	Primary esophago-esophagostomy	Gastrostomy Cervical esophagostomy
D	Feeding should be possible Fistula closure is necessary to avoid aspiration	Feeding should be possible Fistula closure is necessary to avoid aspiration	Primary esophago-esophagostomy	Gastrostomy Cervical esophagostomy
A/B/C/D with long gap >6 vertebral bodies				
	First option		Second option	
	Gastrostomy Cervical esophagostomy Longitudinal stretching		Gastric transposition or colonic esophagus replacement	

Associated syndromes

- At least 15 different syndromes include esophageal atresia in their pattern
- VATER or VACTERL are probably the best known:
  - VATER (Vertebral–Anal–Tracheal–Esophageal–Renal)
  - VACTERL (Vertebral–Anal–Cardiac–Tracheal–Esophageal–Renal–Limp)

Associated malformations

- Trisomy 21
- Duodenal or lower intestinal atresia
- Diaphragmatic hernia
- Skeletal anomalies (vertebral deformations)
- Cardiac malformations
- Ano-rectal malformations
- Tracheomalacia

Signs

- Polyhydramnios
- Fluid shifting in the upper pouch; detected at antenatal ultrasonography
- Postnatal drooling of saliva
- Cyanotic attacks, dyspnea
- Impossible to pass a feeding tube into the stomach
- Aspiration danger

Investigations

- Plain X-ray with a feeding tube (Charr 12–14) in the upper pouch
- Air in the stomach and intestine provides evidence of a lower tracheo-esophageal fistula
- Use of contrast medium is rarely needed and should be restricted to use by experienced pediatric radiologists
- Abdominal ultrasonography
- Cardiologic assessment including echocardiography
- Facultative endoscopy and bronchoscopy
- Appropriate investigations to recognize suspected additional malformations

- Exclusion of a right-sided aortic arch
- Definition of the distance between the upper pouch and the distal esophagus part

Preoperative work-up

- Continuous aspiration of the upper pouch through the feeding tube
- The baby is nursed in the intensive care unit (ICU)
- Immediate surgery is rarely required
- Endotracheal tube beyond a possible distal tracheo-esophageal fistula

Therapeutic strategy

- The main aim of the operation is primary treatment using the patient's esophagus
- In order to achieve this task it is important to know the distance between the upper pouch and the distal end of the esophagus
- An open procedure with extrapleural preparation is standard, however thoracoscopic repair is gaining in popularity
- Table 18.7 gives the different approaches used in different situations

Standard operation

- Right latero-dorsal thoracotomy or vertical incision in the midaxillary line (better cosmetic result)
- Mobilization of the latissimus dorsi muscle taking care to avoid damaging the thoracodorsal nerve
- Mobilization of the serratus anterior muscle along its origin
- Division of the intercostal muscles along the border of the fifth rip
- The parietal pleura is swept away from the thoracic wall taking care not to open it
- Preparation of the azygos vein, which is dissected
- The distal esophago-tracheal fistula is freed from surrounding tissue
- Traction sutures are placed at the tracheal and esophageal ends
- The esophageal fistula is divided and simultaneously closed, avoiding any blood in the trachea
- A feeding tube is inserted in the upper pouch so that it is stretched

- The upper pouch is freed from surrounding tissue and dissected in order to pull it down to the lower esophagus
- The upper pouch is opened at its lowermost point by a horizontal incision resulting in a fish-mouth-shaped aperture
- The sizes of the lumina of the two esophageal parts are always different: the proximal segment is dilated and the distal segment narrow
- The goal is a tension-free end-to-end anastomosis
- A feeding tube is passed through the anastomosis
- The operative steps are illustrated in Fig. 18.4

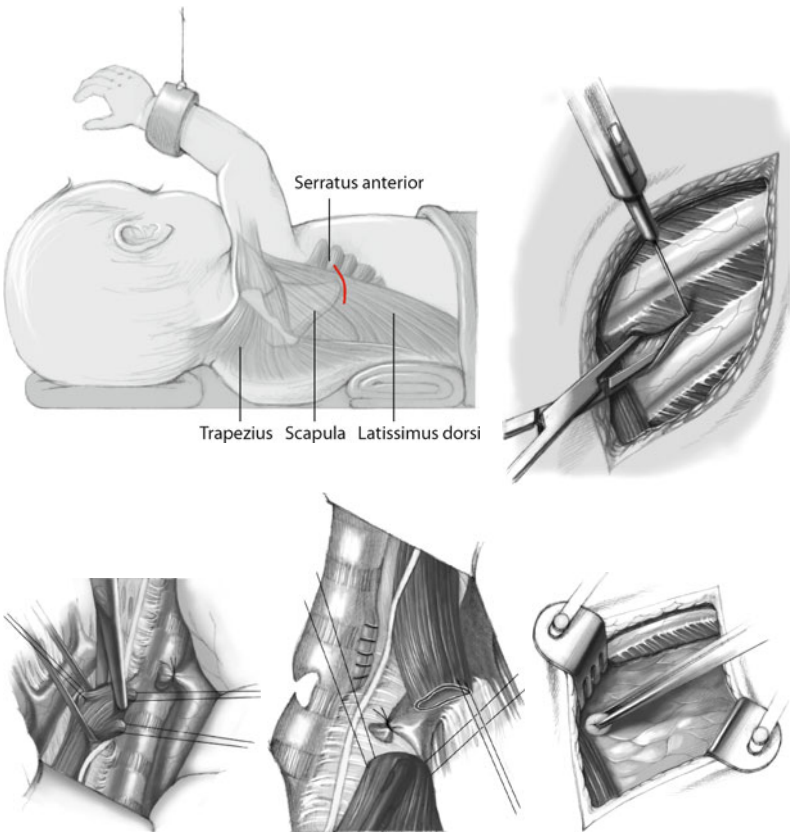


Fig. 18.4 Operative steps: the standard repair of esophageal atresia

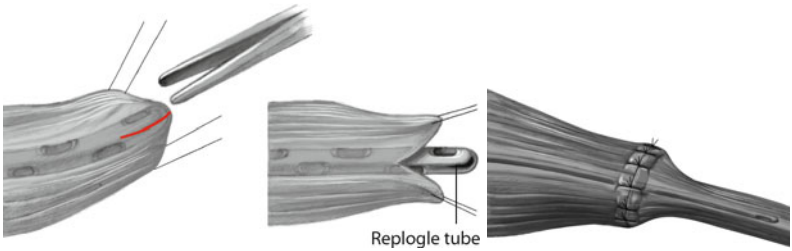


Fig. 18.4 (*continued*) Operative steps: the standard repair of esophageal atresia

Operation: circular myotomy

- The aim is to gain length if, despite mobilization of the upper pouch up to the thoracic inlet, the esophagus endings do not meet (Fig. 18.5)
- A balloon is inserted in the upper pouch
- The muscular layer is divided in a circular or in a spiral fashion
- The anastomosis can be performed after opening the lowermost point

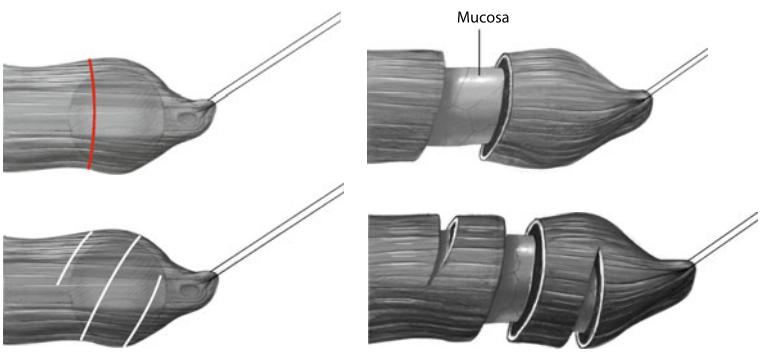


Fig. 18.5 Circular myotomy

Operation: mucosal-muscular flap

- The aim is to gain length if, despite mobilization of the upper pouch up to the thoracic inlet, the esophagus endings do not meet (Fig. 18.6)
- A flap is cut into the upper dilated pouch and turned through 90°
- The dorsal wall is sutured
- A tube is created with the rest of the flap

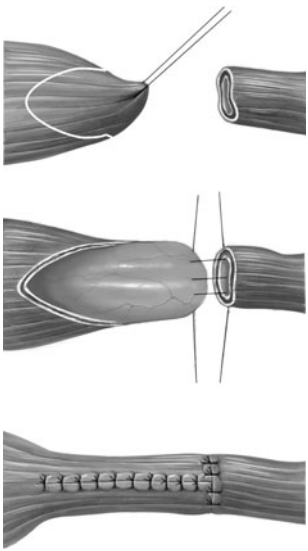


Fig. 18.6 Mucosal muscular flap

Postoperative care

- Intensive care therapy
- Extubation only when a reintubation is unlikely
- Antibiotic therapy
- Nasogastric tube until X-ray control
- Esophagogram 10 days postoperatively with contrast medium
- Feeding according to progress

Complications

- Early complications
  - Pulmonary distress due to preoperative aspiration
  - Anastomotic insufficiency needing revision in the first 2 days
  - Mediastinitis after anastomotic insufficiency with drainage
- Late complications
  - Esophago-tracheal fistula
  - Anastomotic strictures needing bougienage
  - Tracheomalacia
  - Gastroesophageal reflux

Prognosis

- Depends on the form of esophageal atresia and the course of treatment
- Waterson prognostic classification is given in Table 18.8

Table 18.8 Waterson prognostic classification of esophageal atresia

Group	Birth weight	Additional malformation	Survival (%)
I	>1500 g	No cardiac malformation	97
II	<1500 g	Or cardiac malformation	59
III	<1500 g	With cardiac malformation	22

Special Surgical Procedures for Long Gap

Operation: esophagostomy

- The neck is hyperextended and the head is turned to the right
- A transverse incision is made about 1 cm over the clavicula
- Division of the sternal origin of the sternocleidomastoid muscle
- Dissection of the esophagus and identification of the trachea
- The esophagus is freed from surrounding tissue taking care not to injure the recurrent laryngeal nerve
- An esophagostoma is created on the skin through the incision

Operation: gastrostomy

See Chap. 21.2

Operation: longitudinal stretching

- A 10-F tube is passed through the gastrostomy as far as the distal esophagus (Fig. 18.7a)
- A curved metal sound is inserted into the tube
- Under anesthesia a similar tube with a sound is introduced into the upper pouch
- Both probes are pushed towards each other under fluoroscopic control (Fig. 18.7b)
- The procedure is performed twice daily for 3–5 min under mild sedation and analgesia
- Distinct overlapping of the segments enables an end-to-end esophagoesophagostomy to be made after about 5–6 weeks

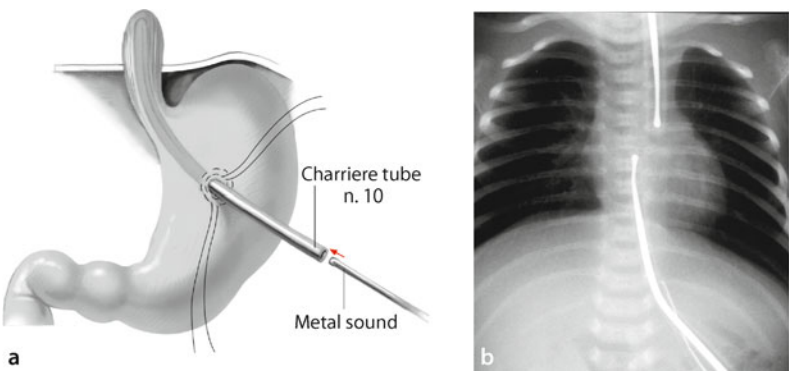


Fig. 18.7a,b Longitudinal stretching

Operation: gastric transposition

- Elliptical incision around the cervical esophagostomy
- Mobilization of the upper esophagus
- Upper midline incision
- The stomach is exposed and the gastrostomy is closed
- The greater and lesser curvatures of the stomach are mobilized preserving the integrity of the right gastroepiploic and right gastric arches
- The distal esophagus stump is resected
- A pyloroplasty is performed
- The fundus is marked with two different-colored sutures in order to avoid rotation
- A mediastinal tunnel directly anterior to the prevertebral fascia is formed by blunt, gentle digital dissection from the abdomen and the cervical incision
- The stomach is pulled through the mediastinal tunnel to the cervical incision
- The upper esophagus is anastomosed with the stomach fundus
- The surgical steps are illustrated in Fig. 18.8

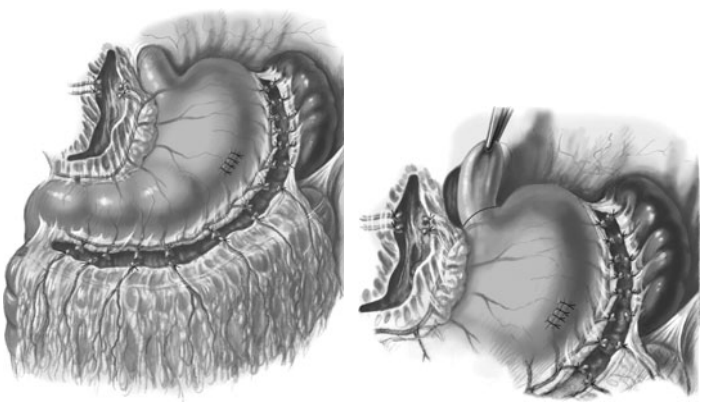


Fig. 18.8 Operative steps: gastric transposition

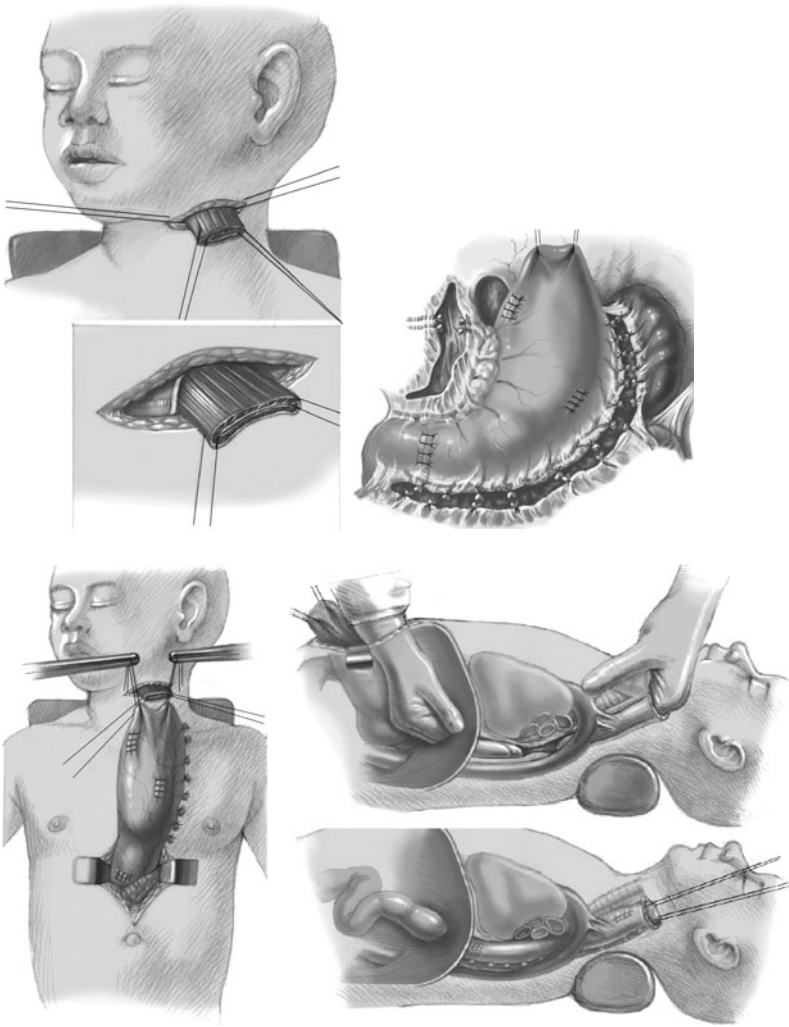


Fig. 18.8 *(continued)* Operative steps: gastric transposition

Operation: colonic esophagus replacement

- Left transverse supraclavicular incision
- Dissection of the esophagus
- Midline abdominal incision
- Mobilization of the colon
- The blood supply to the middle colic vessels is clamped by bulldog clips and the vascularization of the colon is controlled
- The esophago-gastric junction is exposed and the esophagus resected
- The transverse colon is dissected and stretched to re-evaluatate whether the vascularization is sufficient
- A mediastinal tunnel directly anterior to the prevertebral fascia is formed by blunt, gentle digital dissection from the abdomen and the cervical incision
- The colon is pulled through the mediastinal tunnel to the cervical incision
- The upper esophagus is anastomosed with the colon in an isoperistaltic manner
- The gastro-colic anastomosis is performed in two layers
- A 270° stomach anti-reflux wrap is performed
- Additional pyloroplasty
- The surgical steps are shown in Fig. 18.9

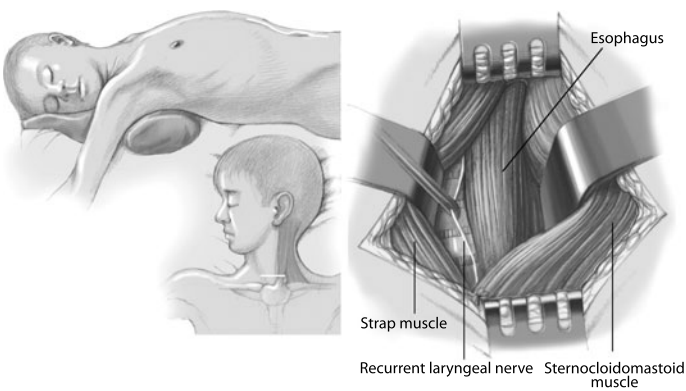


Fig. 18.9 Operative steps: esophageal replacement with colon

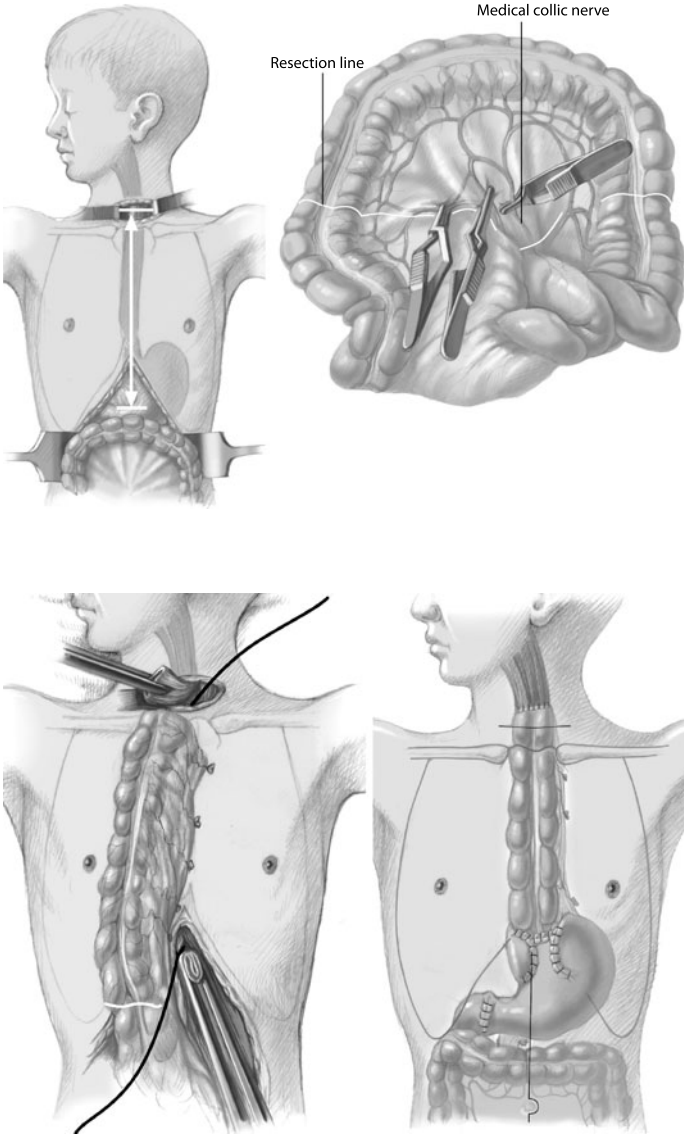


Fig. 18.9 *(continued)* Operative steps: esophageal replacement with colon

18.5 Esophagotracheal H-Fistulas

General considerations

- The H-fistulas are mostly at the level of the second thoracic vertebra
- The fistula is mostly orientated trachea-up to esophagus-down

Signs

- Delayed presentation
- Choking during feedings
- Cyanotic spells
- Drum-like extension of the abdomen due to air passing directly to the stomach during breathing
- Dyspnea
- Recurrent pneumonias due to aspirations

Investigations

- Tracheoscopy
- Esophagoscopy
- Video esophagography with water-soluble contrast medium

Operation

- Tracheoscopic placement of a feeding tube through the fistula
- Hyperextended neck turned to the left
- Incision in a suitable skin crease about 1 cm above the clavicula
- The medial border of the sternocleidomastoid muscle is retracted
- Dissection of the tissues and exposure of the trachea and the esophagus
- Careful development of the plane between trachea and esophagus
- Identification of the fistula by palpating the feeding tube
- Stay sutures are placed on the fistula
- The fistula is divided and the trachea is closed after retracting the feeding tube
- The esophagus is then closed after the anesthetist has placed a nasogastric tube
- A small muscle flap may be placed in the plane between the trachea and the esophagus

- Alternatively, but not necessary, a small piece of Gore-Tex® is used
- The incision is closed
- The operative steps are illustrated in Fig. 18.10

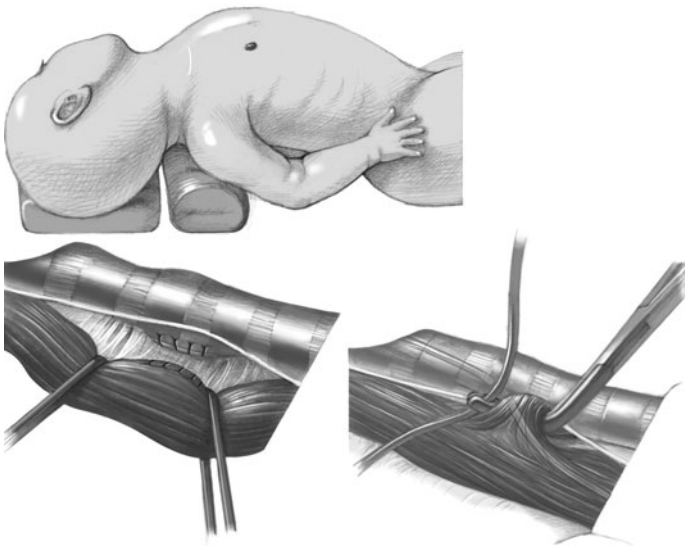


Fig. 18.10 Operative steps: esophagotracheal H-fistula repair

Postoperative care

- Intensive care
- Intubation for at least 2 days
- Antibiotic therapy
- X-ray with contrast on day 5
- Feeding tube for 5 days
- Feeding thereafter

Prognosis

- Very good

18.6 Esophageal Dilatation

General considerations

- The aim of a bougienage is dilatation of a stenosis from the inside of the esophagus
- Dilatations are performed under general anesthesia or whilst the patient is sedated
- Dilatations must be performed gently as perforation is always possible
- Dilatation therapy requires more than one session; sometimes sessions every other day

Methods

- Bougienage with Rehbein bougies (Fig. 18.11)
- Pneumatic bougienage (Fig. 18.12)

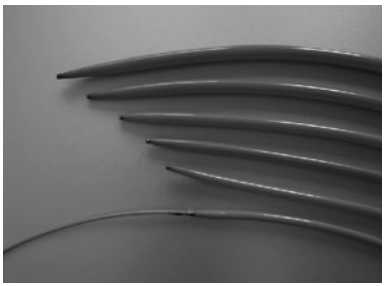


Fig. 18.11 Rehbein bougies

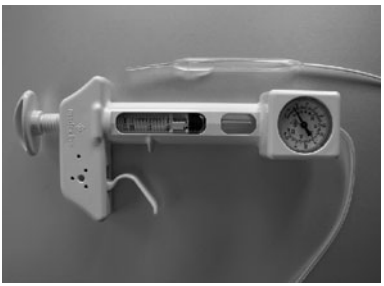


Fig. 18.12 Pneumatic balloon

18.7 Esophageal Varices

General considerations

- Dilatation of the physiological portocaval system
- Induced by portal hypertension due to extra- or intra-hepatic block
- Extra-hepatic congenital malformations are the main reasons (portal vein obliteration, valves)

- Catheterization of the umbilical vein directly after birth may lead to portal vein obliteration
- Intra-hepatic congenital malformations with biliary atresia

Classification

- Child–Pugh classification (Table 18.9)

Table 18.9 Child–Pugh classification of liver cirrhosis

	1 point	2 points	3 points
Prothrombin time (seconds prolonged) (%)	<4	4–6	>6
Encephalopathy	No	Grade I–II	Grade III–IV
Ascites ($g \cdot day^{-1}$)	None	Conservative therapy	Therapy resistant
Serum albumin ($g \cdot l^{-1}$)	>3.5	3.5–2.8	<2.8
Serum bilirubin ($mg \cdot l^{-1}$)	<2.0	2.0–3.0	>3.0
Child A = 5–6 points; Child B = 7–9 points; Child C = 10–15 points			

Signs

- Massive bleeding from the esophagus
- Periumbilical caput medusae
- Hemorrhoids
- Splenomegaly
- Anemia and thrombocytopenia
- Dilated abdomen
- Ascites

Preoperative work-up

- Blood count and coagulation status
- Liver function tests
- Esophagogram
- Esophagoscopy
- Ultrasonography of the liver
- Doppler ultrasonography of the portal vein
- Angio-MRI with imaging of the mesenteric artery

Therapy

- Blocking of the esophagus with an inflatable probe (Sengstaken probe)
- Irrigation of the stomach and the esophagus with cold 0.9% saline solution taking care that the patient does not aspirate
- Esophagoscopy with sclerozation of the esophageal varices

Operation

- The aim of the operation is discontinuation of the venous flow at the gastro-esophageal junction
- Different methods of shunt operations are propagated (Fig. 18.13)
- In cases of severe liver cirrhosis a liver transplantation is indicated

Prognosis

- Depends on the grade of liver cirrhosis
- The 5-year survival rate after shunt operation is about 50%

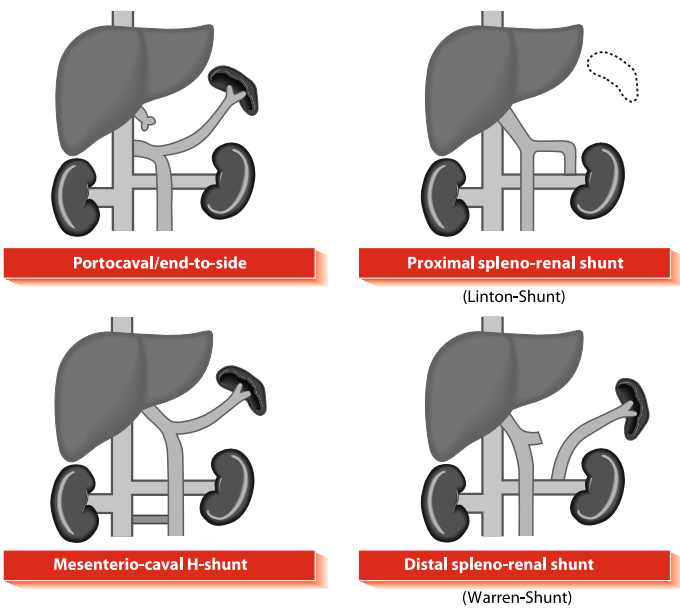


Fig. 18.13 Shunt operations

19 Abdominal Wall

19.1 Umbilical Hernia

General considerations

- Maldevelopment in the closure of the transit point of the umbilical vessels
- Approximately 20% of all full-term neonates have an umbilical hernia, as opposed to 70%–80% of premature neonates
- The hernia is a peritoneal sac, adherent to the umbilical skin
- In about 80% of cases spontaneous regression of the hernia will occur during the first 2 years of life
- Operative treatment is always indicated in cases where there is a very large umbilical hernia (hernia ring >2 cm in diameter) and where there is persistence beyond the second year of life
- Dressings and hernia trusses are obsolete; their only effect is to injure the skin

Preoperative work-up

- Normal preoperative measures
- The procedure may be done as an outpatient procedure in children older than 6 months

Operation

- The operative steps are illustrated in Fig. 19.1
- Infraumbilical semicircular incision
- Preparation of the fascia caudal to the hernia sac

- Isolation and division of the sac
- Closure of the sac with interrupted sutures
- Burying of the first row of sutures with a second (leave the middle suture long)
- Inversion of the skin and fixation of the deepest point with the suture left long
- Intradermal closure of the skin incision

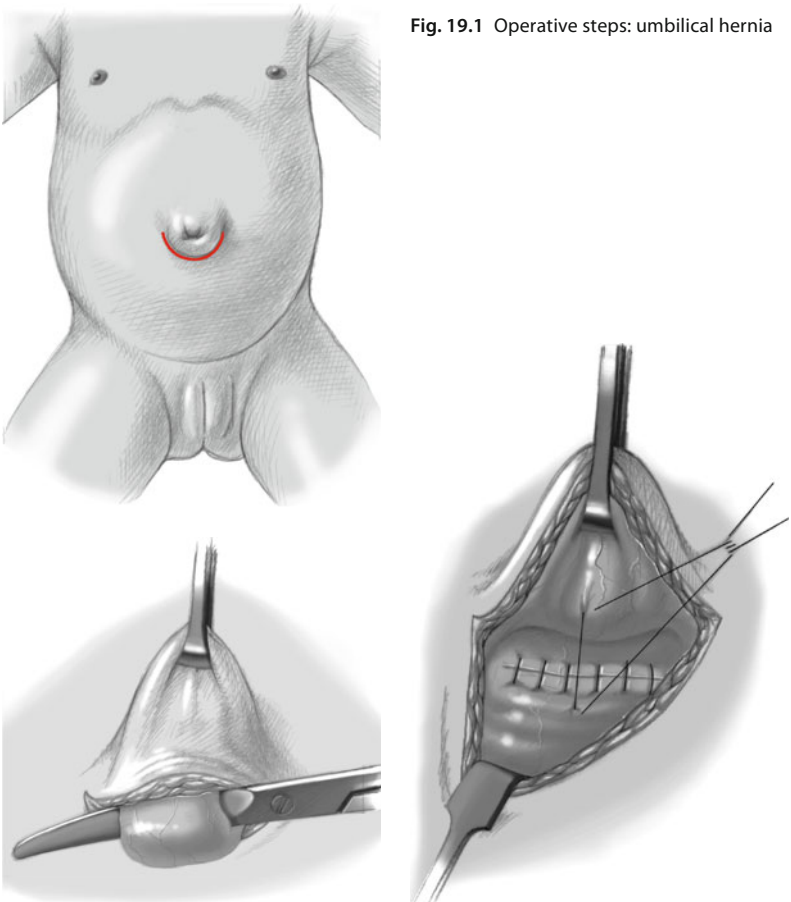


Fig. 19.1 Operative steps: umbilical hernia

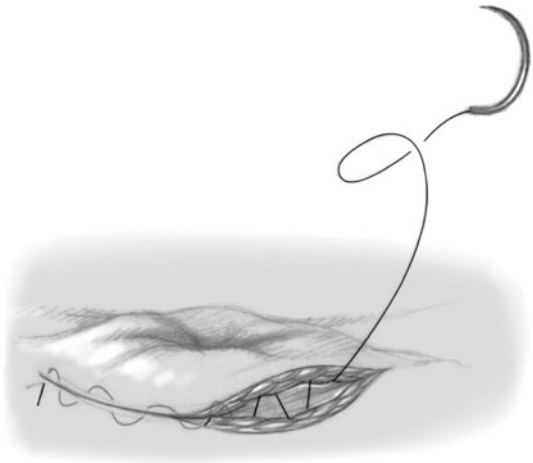


Fig. 19.1 *(continued)* Operative steps: umbilical hernia

Postoperative care

- Suture removal on the 6th postoperative day

19.2 Supraumbilical Hernia

General considerations

- Malformation along the linea alba
- Epigastric hernias are frequent around the center of the epigastrium
- Fat from the falciform ligament may prolapse into the hernia
- Supraumbilical hernias occur directly above the navel
- Omentum may prolapse in the hernia

Signs

- Pain is the most frequent sign, caused by agglutination of fat or omentum

Preoperative work-up

- Marking of the fascial defect preoperatively, with the patient lying supine and the upper part of their body lifted

Operation

- The operative steps are illustrated in Fig. 19.2
- Transverse incision over the marked defect
- Isolation and opening of the hernia
- Resection of the prolapsed omentum or fat is advised
- Closure of the fascia
- Intradermal closure of the skin

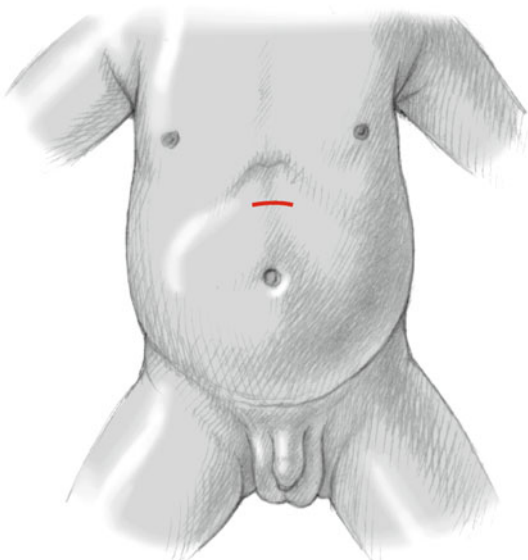
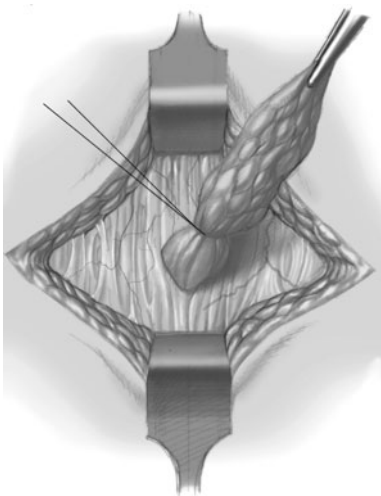


Fig. 19.2 Operative steps: supraumbilical hernia



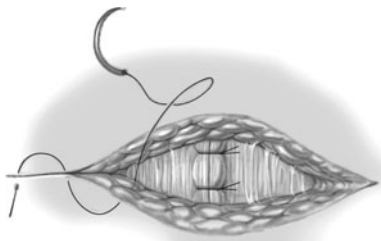


Fig. 19.2 Operative steps: supraumbilical hernia

Postoperative care

- Suture removal on the 6th postoperative day

19.3 Omphaloenteric Duct

General considerations

- Connection between the yolk sac and the umbilical loop of the embryonic gut
- The regression of this duct has various courses (Fig. 19.3), leading to the following anatomical variants:
  - Meckel's diverticulum – the duct as a protrusion from the gut (see Chap. 23)
  - Omphaloenteric duct with cyst formation and partial obliteration
  - Persistent omphaloenteric duct – connection between the gut and the navel

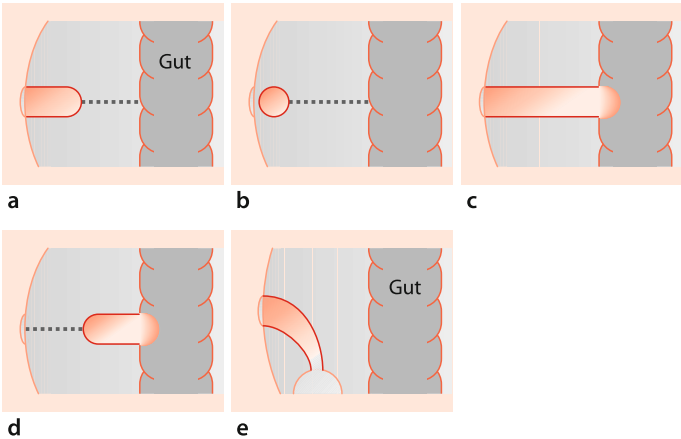


Fig. 19.3a–e Possible regression anomalies of the omphaloenteric duct. **a** umbilical fistula, **b** umbilical cyst, **c** persistent omphaloenteric duct, **d** Meckel diverticulum, **e** urachus

Signs

- Wet navel
- Hemorrhage
- Intussusceptions
- Intestinal obstruction

Preoperative work-up

- Ultrasonography
- Plain abdominal X-ray
- Probing of the fistula
- Contrast imaging of the fistula
- Isotope scintigraphy

Operation

- The operative steps are illustrated in Fig. 19.4
- Resection of Meckel's diverticulum

- Circular umbilical incision, if necessary with lateral extension
- Mobilization of the duct
- Opening of the peritoneum and exposure of the connection to the ileum
- Resection of the duct and closure with suture or clips
- Reconstruction of the umbilicus

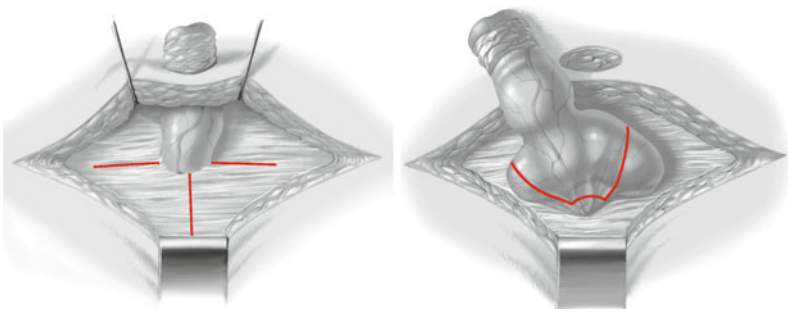


Fig. 19.4 Operative steps: omphaloenteric duct

Postoperative care

- Suture removal on the 6th postoperative day

19.4 Urachal Cysts/Urachal Fistulas

General considerations

- Connection between the umbilicus and the bladder dome (Fig. 19.5)
- If the connection does not obliterate a fistula results, either totally patent or partially obliterated
- Obliteration of both ends will lead to the formation of a cyst in the region between the umbilicus and the bladder
- May be combined with prune belly syndrome or posterior urethral valves
- Spontaneous regression may occur within the first few months of life

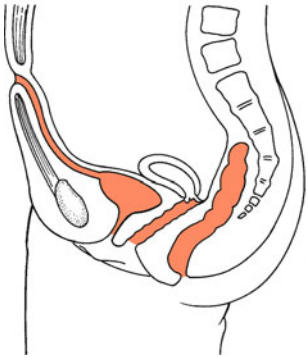


Fig. 19.5 Urachal fistula

Signs

- Wet navel
- Small, secreting opening at the base of the navel
- Secondary infection
- Urinary tract infections
- Tumor in the lower abdomen by the urachal cyst
- Pyourachus by infected urachal cyst

Preoperative work-up

- Probing of the fistula
- Radiographic imaging with water-soluble contrast medium
- Ultrasonography
- Voiding cystourethrography
- Local disinfectant or possibly targeted antibiotics until the region is free from inflammation

Operation

- Urachal fistula
  - Semicircular umbilical incision
  - Extraperitoneal exposure of the fistula
  - Circumcision of the fistula at the umbilicus
  - Resection of the fistula down to its entry into the peritoneum

- Urachal cyst
  - Lower abdominal transverse incision
  - If possible, total extraperitoneal excision of the cyst
  - Closure at the bladder dome with over-sewing of the bladder wall

Postoperative care

- In cases of infected fistulae and cysts antibiotic treatment should be continued after the operation
- Removal of sutures on the 6th postoperative day

19.5 Inguinal Hernia

General considerations

- In children almost always indirect hernias; at the testicular descent the internal inguinal ring (the vaginal process) remains patent and a connection between the peritoneal and scrotal cavities persists
- Inguinal hernia is more frequent in boys than in girls (ratio 9:1)
- Inguinal hernia is more frequent in prematures
- About 60% of inguinal hernias are right-sided, as the right testis descends later than the left
- About 25% of inguinal hernias are left-sided
- About 15% of inguinal hernias are bilateral

Signs

- Inguinal swelling when the patient cries
- Inguinal asymmetry
- Palpation of a hernia ring and reduction of gut
- "Silk sign." One has the feeling that the palpating finger at the vaginal process slides over a silk cloth
- Variations in the size of the scrotum caused by fluid collection through the connection

- Intestinal obstruction; signs caused by trapping of gastric contents
  - Tenderness of the swelling
  - Local redness (after repeated attempts at reduction)
  - Vomiting
  - Fever

Mothers are the best observers!
Rather believe in the mother than in your own hands.

Complications

- Incarceration, most often a loop of small intestine
- Weaning (milk → solid diet) often precipitates the inguinal hernia in those who are predisposed
- Frequently occur after a change of diet (the consistence of feces changes)
- Danger of intestinal wall necrosis
- Danger of decreased testicular circulation through spermatic cord compression
- In girls the ovary is endangered
- Incarceration rate:
  - Prematures in first trimester: 50%
  - Mature neonates <1 year: 30%
  - Mature neonates >1 year: 15%

Classification

- Inguinal hernia classification is given in Table 19.1 and Fig. 19.6

Table 19.1 Inguinal hernia classification

Medial hernia	Medial of the epigastric vessels	Directly through the outer inguinal ring	Direct hernia
Lateral hernia	Lateral of the epigastric vessels	Indirectly through the inguinal canal	Indirect hernia
Femoral hernia	Medial or lateral of the epigastric vessels	Through the femoral hiatus	

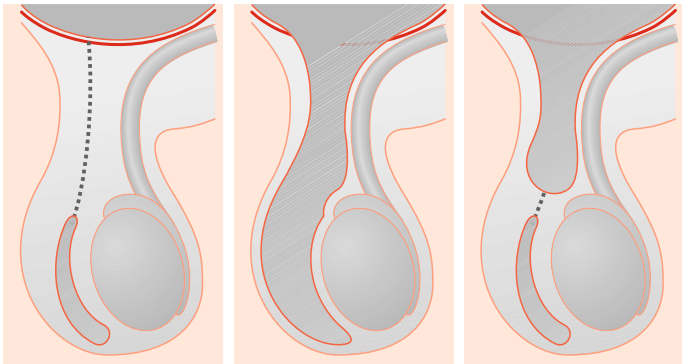


Fig. 19.6a–c Classification of inguinal hernias. **a** Physiological obliteration, **b** scrotal hernia, **c** inguinal hernia

Operation

- The operative steps are illustrated in Fig. 19.7
- Reduction attempt at the time of diagnosis – except when there is incarceration with signs of intestinal ischemia – under sufficient analgesia. Take your time when performing the reduction
- Constant compression of the swelling from the caudal end with simultaneous pressure over the external inguinal ring from the cranial side
- For emptying, compression of the intestinal loop
- Operation only on the side of the documented hernia
- Bilateral operation when the operation on the first side was uneventful
- Cleaning of the lower abdomen, the inguinal region, and the scrotum
- Incision in the lower abdominal fold
- Exposure of the external inguinal ring
- Opening of the fascia, isolation of the sac and the spermatic cord structures
- Clamping of the hernia sac after opening the cremaster muscle and separation from the deferent duct and the cord vessels
- Preparation of the sac to the base at the internal inguinal annulus

- Placement of a transfixing suture at the base of the sac and burying of the stump under the external muscle
- Closure of the fascia
- Closure of the skin with an intradermal suture
- The operation in girls is similar, with the difference being that the inguinal canal is completely closed

Fig. 19.7 Operative steps: inguinal hernia

Postoperative care

- Except for babies younger than 1 year the patient can be discharged on the day of operation
- Analgesia prescribed individually
- Suture removal on the 6th postoperative day

Complications

- Injury to the deferent duct or vessels
- Immediate microsurgical reconstruction
- Recurrence by incomplete closure of the sac or unnoticed peritoneal tear
- Testicular atrophy by narrowing of the vessels
- Hydrocele testis, when the sac around the testis has not been left open
- Wound infection, especially after incarceration

19.6 Femoral Hernia

General considerations

- Very rare in children
- Mistaken for enlarged inguinal lymph nodes

Signs

- Bulge somewhat below the inguinal region
- Pain on walking

Operation

- Exposure of the external inguinal ring and the inguinal ligament
- Preparation of the inguinal ligament to the lacuna vasorum
- Exposure of the hernia sac without opening it
- Reduction of the sac and suture of the pectineal ligament to the inguinal ligament without narrowing the femoral vein
- Closure as performed in inguinal hernia
- The operative steps are illustrated in Fig. 19.8

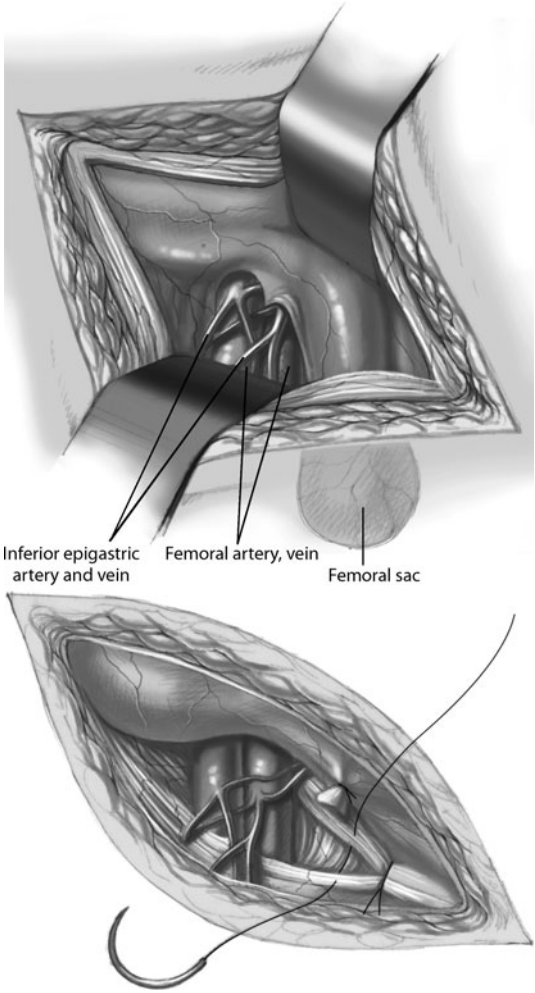


Fig. 19.8 Operative steps: femoral hernia

19.7 Other Types of Hernias

Definitions

- Other types of hernia are defined in Table 19.2

Table 19.2 Hernia definitions

Inguinal hernia	Direct, acquired, presentation medial to the epigastric vessels Indirect, congenital, presentation lateral to the epigastric vessels
Femoral hernia	Below the inguinal ligament, mostly medial to the femoral vessels
Incisional hernia	Result of muscle weakness after an operation
Paraumbilical hernia	A navel of normal size lies beside the hernia swelling
Spieghel's hernia	In the linea semilunaris lateral to the abdominal rectus muscle
Rectal diastasis	Congenital or acquired diastasis of the rectus muscle margins
Obturator hernia	Along the obturator vessels through the obturator foramen
Ischiadic hernia	Through the ischiadic foramen
Perineal hernia	Through the vesicouterine or rectouterine fossa
Superior lumbar hernia	Through the upper lumbar triangle, 12th rib/erector spinae muscle
Inferior lumbar (Petit's hernia)	Above the iliac crest and lateral to the latissimus dorsi muscle
Littre–Richter hernia	Incarceration of part of the intestinal wall

19.8 Hydrocele

General considerations

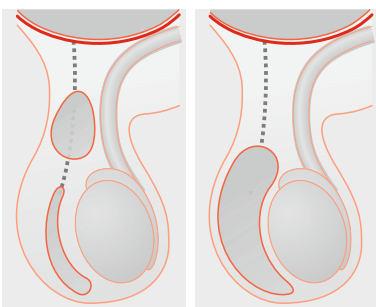
- Hydroceles are fluid collections in a cavity lined with peritoneum, occurring because the vaginal process has remained patent
- Very often accompanied by an indirect inguinal hernia
- Frequently present in neonates, with spontaneous regression during the first 6 months of life
- Extremely large hydroceles may cause pressure injury to the testis
- Differential diagnosis: testicular tumor or cyst

Classification

- Hydrocele classification is given in Table 19.3 and Fig. 19.9

Table 19.3 Hydrocele classification

Hydrocele testis	Isolated hydrocele of the testis without/with connection to the abdominal cavity
Hydrocele funiculi spermatici	Isolated hydrocele of the spermatic cord without/with connection to the abdominal cavity
Hydrocele testis et funiculi	Combination of the above-mentioned types
Symptomatic hydrocele	Fluid collection in the scrotum after incarceration, testicular torsion, epididymitis, etc.
Pyocele	Infected hydrocele with putrid content



a b

Fig. 19.9a,b Forms of hydrocele. **a** Funicular hydrocele, **b** testicular hydrocele

Signs

- Swelling in the scrotum, varying in volume. Mostly larger in the evenings than in the mornings
- The scrotal skin folds may be obliterated by large hydroceles

Preoperative work-up

- It is possible to grasp totally around the upper pole of the hydrocele testis
- The hydrocele funiculi spermatici can be palpated as an isolated cyst between the inguinal region and the testis

- Transillumination (not certain in babies, where also the intestines can be transilluminated)
- Ultrasonography

Operation

- Indicated when there is no tendency to spontaneous regression
- Operation after the 6th month of life for remaining hydrocele
- Operative technique as described above for an inguinal hernia
- For recurrent hydrocele
  - Scrotal incision
  - Exposure and broad opening of the hydrocele
  - Reversal of the hydrocele wall around the cord structures
  - Adaptation of the wall with sutures, so that the peritoneum is turned outwards (avoidance of agglutination and renewed recurrence)

Postoperative care

- This is the same as explained above for inguinal hernia repair

Prognosis

- Frequency of recurrence after correct management is <1%

19.9 Omphalocele (Exomphalos)

General considerations

- Fetal maldevelopment with failure of the usual physiological reduction to the abdomen of the gut
- Distinguish between hernia of the cord (defect <4 cm, containing only midgut) and omphalocele (large defect, 5–12 cm, containing also liver and other viscera)
- No skin cover, the wall of the omphalocele consists of amnion externally and peritoneum internally. The umbilical cord is an extension of the omphalocele wall

- Associated anomalies are very frequent with omphalocele (50%–60%)
  - Intestinal rotation anomalies
  - Cardiac malformations
  - Renal malformations
  - Trisomies
  - Syndromes (e.g., Cantrell pentalogy)
- Associated anomalies are rare with hernia of the cord, and the primary closure is mostly easy

Management

- Referral to tertiary center as soon as the diagnosis is set antenatally, especially if there is suspicion of multiple anomalies
- The pediatric surgeon must keep the parents well informed
- Follow-up of pregnancy can be performed in a local hospital, but preferably in a specialist center
- Reference to the tertiary center before birth
- Complications caused by rupture of the mostly thin omphalocele sac
- The best incubator is the mother's womb

Preoperative work-up

- Nasogastric tube with continuous drainage
- Sterile dressing with moist towels (no wet towels – cooling!)
- Protection with plastic foliage (prevents desiccation)
- Lateral position with support provided for the prolapsed viscera
- Intravenous access for substitution of fluid losses
- Transport in heated incubator
- Rectal irrigation with warm sterile 0.9% NaCl solution (empty gut = smaller omphalocele)

Investigations

- Abdominal and chest X-ray
- Cardiac investigation (ECG, cerebral ultrasonography)
- Blood count, blood gases, electrolyte and fluid status

Conservative therapy

- Eschar formation
- The omphalocele sac is painted daily with substances inducing eschar formation until the skin covers the omphalocele
- Long therapy
- Danger of rupture and infection
- Closure where there is a hernia covered with skin
- Definitive operation on the cicatricial hernia after the child is about 2 years old

Silo with omphalocele sac

- The sac is covered with sterile, moistened dressings followed by circular dressing with gentle pressure
- The dressed sac should be hung up above the child so that the viscera drop into the abdomen (Fig. 19.10)
- Daily dressing change with gentle kneading of the sac to release minor adhesions into the sac
- The empty sac is twisted and ligated with umbilical ties
- Once the viscera are reduced as much as possible a definitive operation (see below) is performed

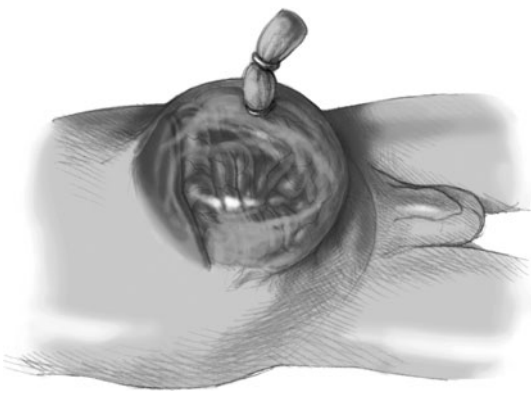


Fig. 19.10 Silo with omphalocele sac

Operation: primary closure

- For hernia of the cord and small and medium-sized omphaloceles
- Excision of the sac along the skin border
- Ligature of the umbilical structures
- Revision of the intestine (atresias, malrotation, persistent omphaloenteric duct)
- Stretching of the skin and mobilization of the rectus muscle borders
- Closure of all layers
- The operative steps are illustrated in Fig. 19.11

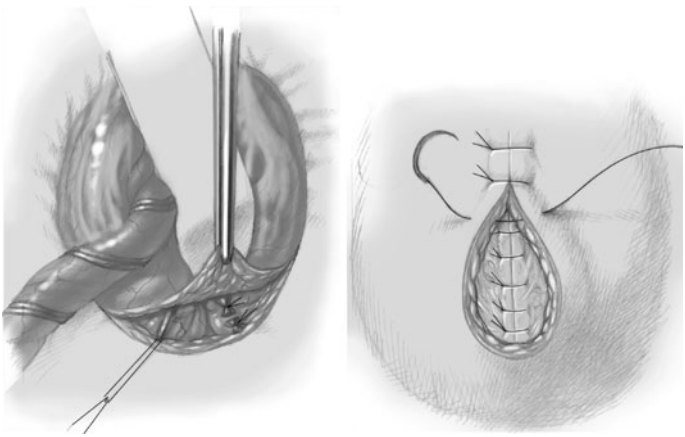


Fig. 19.11 Operative steps: omphalocele

Operation: Schuster plasty

- Preparation as above
- Suture of a silastic foil to the skin and muscle borders
- Closure of the borders of the foil in the shape of a chimney
- Subsequent reduction of the viscera over 7–14 days
- The operative steps are illustrated in Fig. 19.12

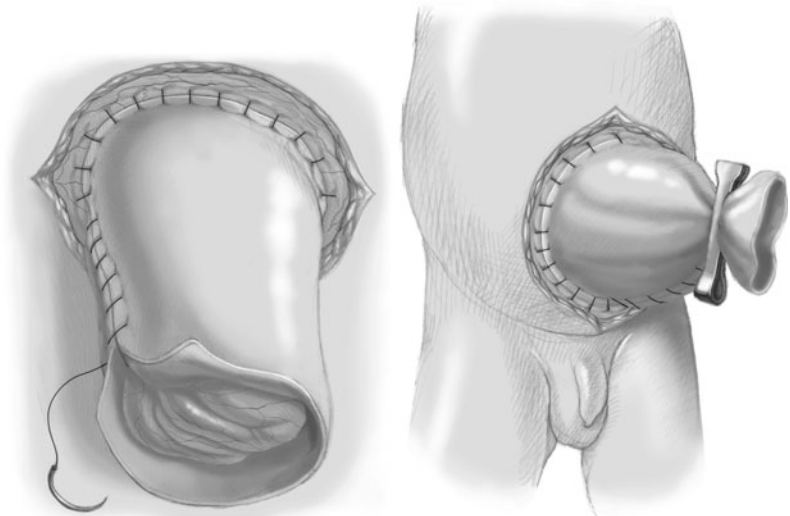


Fig. 19.12 Operative steps: Schuster plasty

Operation: Amnion-vicryl net as peritoneal substitute (editor's method)

- See Sect. 19.10

Postoperative care

- Intravenous antibiotic therapy
- Ventilatory assistance is necessary in the first few days for adaptation to the new, increased intra-abdominal pressure
- Early stimulation of intestinal movements
- Oral feeding depending on intestinal motility

Prognosis

- Strongly dependent on associated anomalies and gestational age

19.10 Laparoschisis (Gastroschisis)

General considerations

- Median abdominal wall cleft
- Midgut and occasionally other viscera prolapse through the defect with no membrane cover
- The umbilical cord arises to the left of the defect, separated by a narrow skin bridge
- Bowel walls may be thickened and fibrin-covered from the permanent exposure to amniotic fluid
- Combination with intestinal stenosis and atresias (about 14%)
- Associated cardiac and renal anomalies are rare

Management

- Referral to tertiary center as soon as the diagnosis is set antenatally
- The pediatric surgeon must keep the parents informed
- Follow-up of pregnancy can be performed in a local hospital, but preferably in a specialist center
- Refer to the tertiary center before birth
- Risk of infection through exposure of the abdominal contents
- Most of the children with laparoschisis are born prematurely (body weight 2000–2500 g)

Preoperative work-up

- Emergency, absolute indication for surgery
- Transport and investigations identical to those for an omphalocele
- Bacteriological smear from bowel wall
- Most children are hypovolemic (give fluid infusion until normovolemic)
- Broad-spectrum antibiotic therapy started preoperatively
- Rectal irrigation with heated 0.9% NaCl solution (sterile conditions); bowel emptied = decrease in size

Placental preservation with amnion as peritoneal substitute (editor's method)

- Upon delivery, the placenta is packed in sterile towels and taken to the-ater with the baby
- The amniotic membranes are separated from the placenta at a separate sterile table and placed in a receptacle with 0.9% NaCl solution
- The amniotic membranes have two surfaces, a shiny one facing the fetus and a raw one facing the uterus
- The membrane is spread out on a sterile drape with the shiny surface down and the raw surface is brushed off with a moist sterile towel. Rem-nants of decidua and chorion are removed in this way
- The remaining amnion is passed to the operating table in a receptacle containing antibiotic solution

Operation

- The operative steps are illustrated in Fig. 19.13
- Immediate reduction without anesthesia may sometimes be possible when the intestines are not thickened and fibrin-covered
- Operation carried out with the patient placed on a warming mattress
- Shorten the umbilical cord
- Inspect the intestine in order not to miss atresias
- Make a cranial midline skin incision
- Expose the muscle borders and the fascia
- Stretch the abdominal wall
- Reduce the gut into the abdomen; monitor the intra-abdominal pressure and do not allow it to exceed 20 cmH_2O
- Primary closure without tension whenever possible

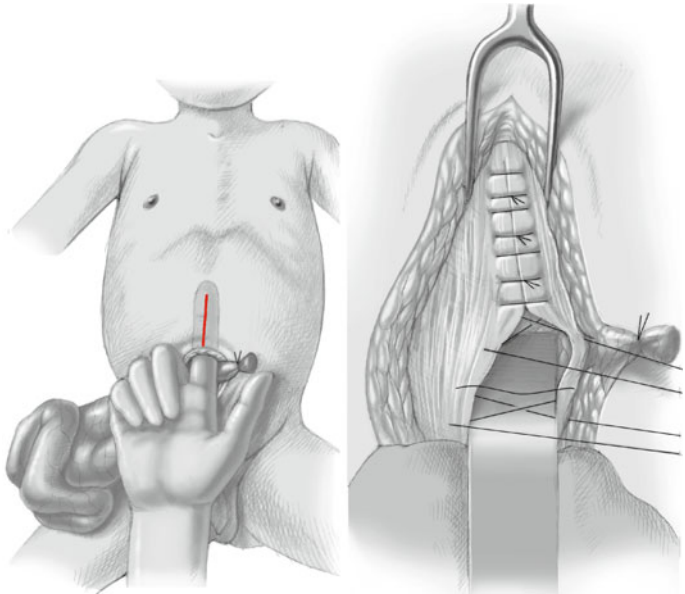


Fig. 19.13 Operative steps: laparoschisis

Operation: Amnion-Vicryl net as peritoneal substitute

- When primary closure is not possible, use amnion as a peritoneal substitute
- The previously prepared amnion (see above) is spread out under the muscle borders with the shiny surface facing the abdominal cavity. Fixation is unnecessary
- The operative steps are illustrated in Fig. 19.14

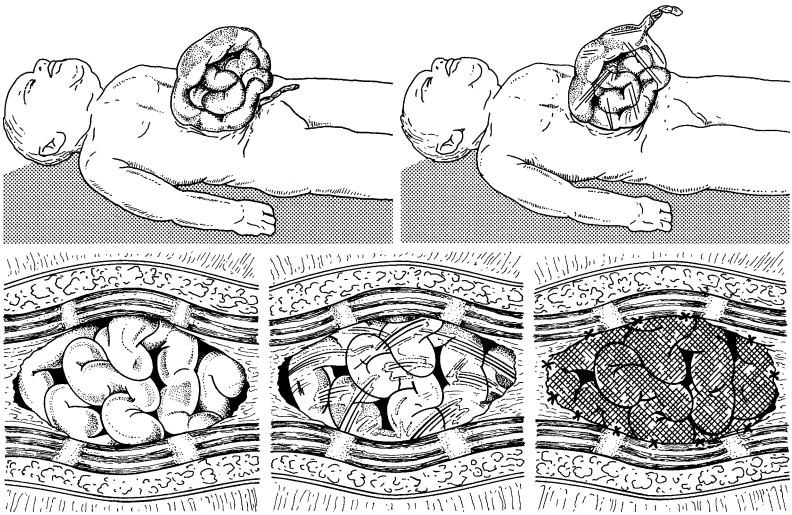


Fig. 19.14 Operative steps: Amnion-Vicryl as peritoneal substitute

Operation: reconstruction of the abdominal wall

- The Vicryl net is placed on the raw surface of the amnion along the muscle borders
- The skin can now be closed over the Vicryl net after mobilization
- In a second operation in the second year of life the cicatricial hernia is revised
- In almost every case a total reconstruction of all layers of the abdominal wall is possible, as the abdominal cavity is now large enough to accommodate the viscera
- With the method used, no agglutination between the abdominal viscera and the abdominal wall occurs, because of the presence of the amnion. The Vicryl net is absorbed and transformed into a firm fibrous plate

Postoperative care

- Systemic antibiotic prophylaxis
- Ventilatory assistance is necessary in the first few days for adaptation to the increased intra-abdominal pressure
- Early stimulation of intestinal motility
- Oral feeding depending on intestinal motility
- Rectal enemas

Prognosis

- In general, more than 90% are cured in centers with sufficient experience
- Long-term parenteral nutrition increases the danger of sepsis
- Low morbidity

20 Abdomen

20.1 Principles of Abdominal Surgery

- Every operation should have a benefit for the patient
- The benefit might be diagnostic or therapeutic
- This benefit has to be achieved with the surgical procedure that provides maximal benefit with minimal risk
- Mutilating operations should be always avoided

20.2 Organ Anatomy

- Stomach (Figs. 20.1, 20.2)
- Small bowel (Fig. 20.3)
- Colon (Fig. 20.4)
- Rectum (Fig. 20.5)
- Liver (Fig. 20.6)
- Pancreas (Fig. 20.7)
- Adrenals (Fig. 20.8)

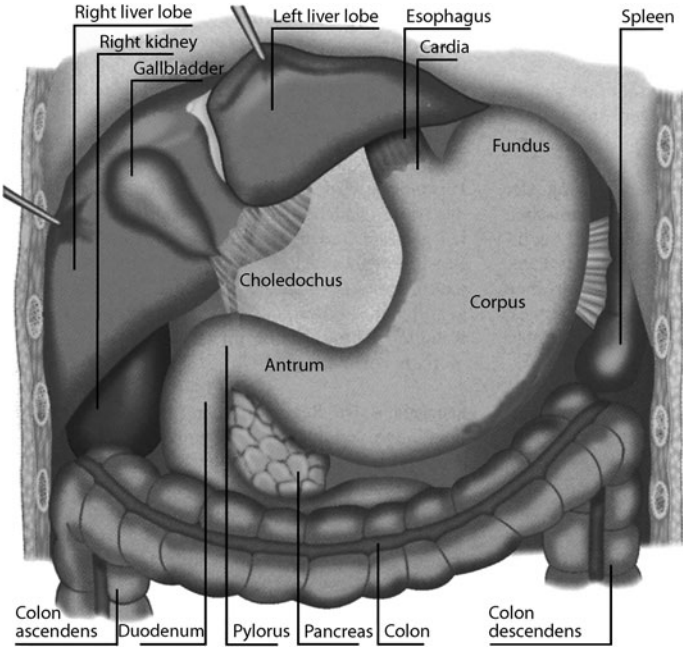


Fig. 20.1 Topographic anatomy of the stomach

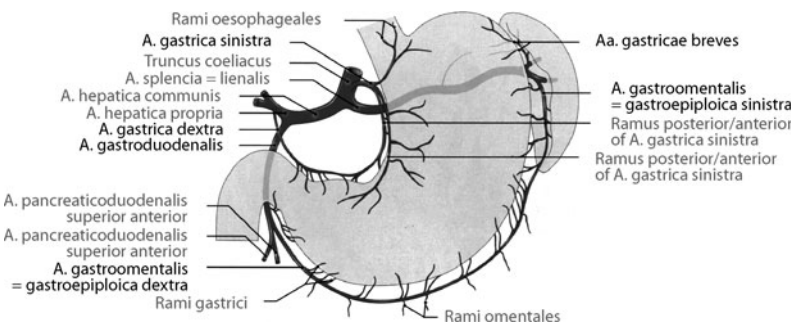


Fig. 20.2 Vascular anatomy of the stomach, spleen, liver, and duodenum

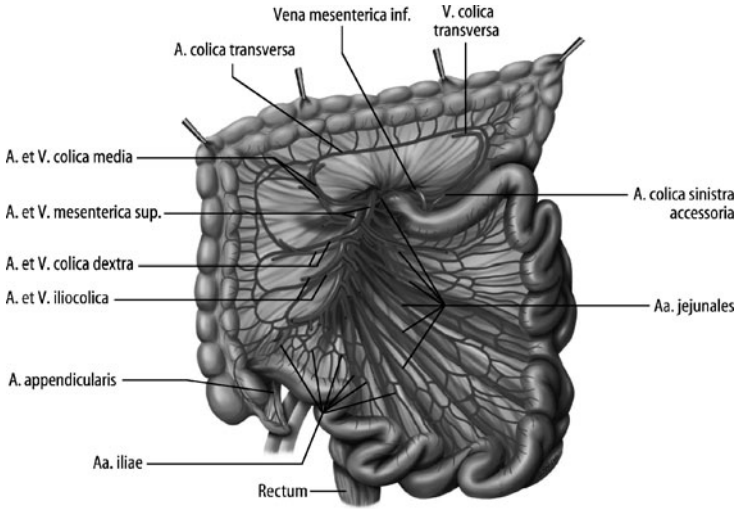


Fig. 20.3 Topographic and vascular anatomy of the small bowel

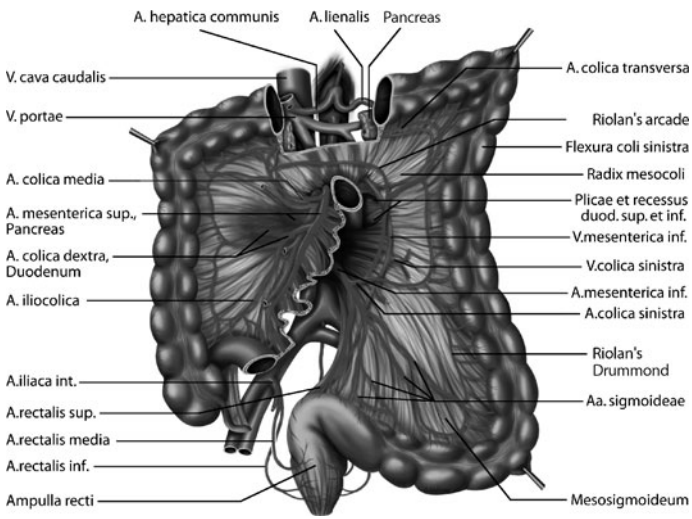


Fig. 20.4 Topographic and vascular anatomy of the colon

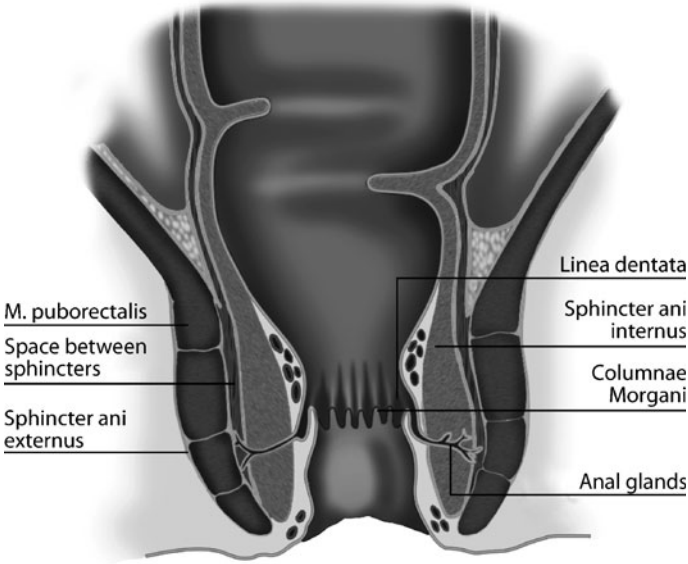


Fig. 20.5 Topographic anatomy of the rectum

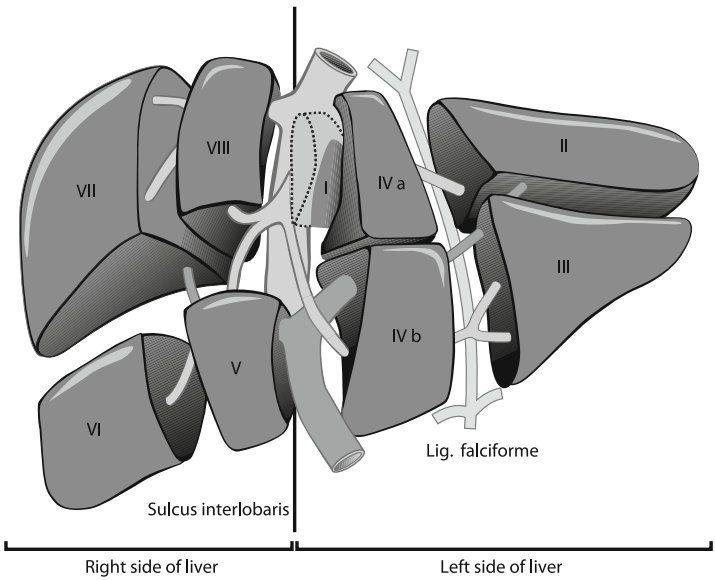


Fig. 20.6 Segmental arrangement of the liver

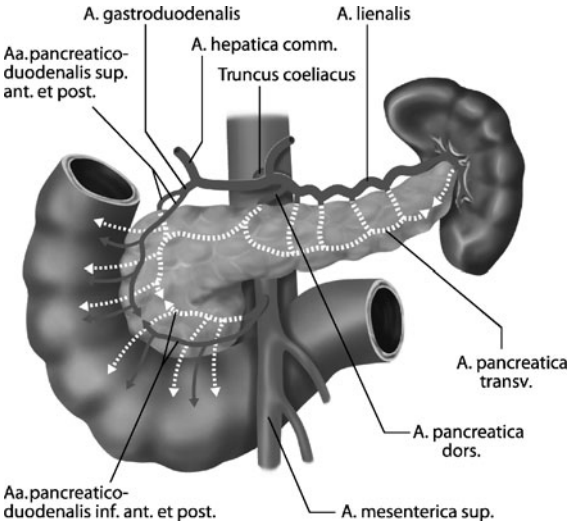


Fig. 20.7 Topographic and vascular anatomy of the pancreas

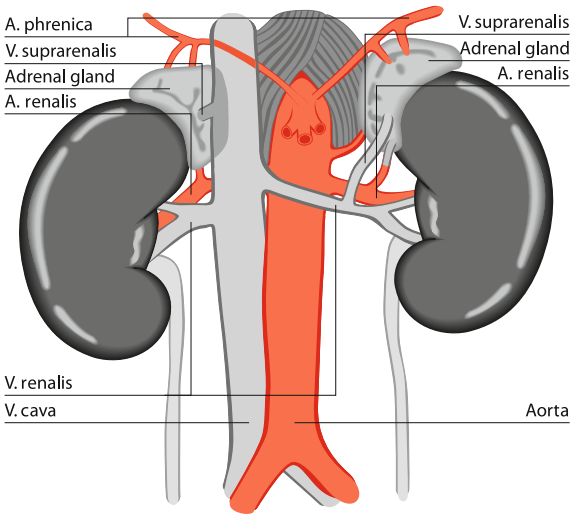


Fig. 20.8 Topographic and vascular anatomy of the adrenals

20.3 Anatomical Abdominal Particularities in Children

Age-related differences in anatomy

- Age-related differences in anatomy are listed in Table 20.1 and illustrated in Fig. 20.9

Table 20.1 Age-related differences in anatomy

	Newborn	Older child/adult
Form	Square	Rectangle
Costal arch	Wide angle	Acute angle
Rectus muscle	Wide, more lateral	Narrow more centralized
Liver	Large, below the epigastrium	Below the costal arch
Umbilicus	Closer to the symphysis	Distant to the symphysis
Bladder	Over the symphysis	Behind the symphysis

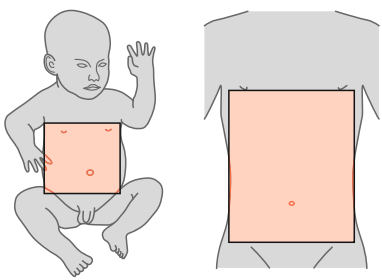


Fig. 20.9 Differences in the abdomen of infants and adults

20.4 Abdominal Incisions

- Types of abdominal incision and their advantages and disadvantages are described in Table 20.2

Table 20.2 Types of abdominal incision, their advantages and disadvantages

Incisions	Definition	Advantages	Disadvantages
Midline	Incision in the midline incising the skin on the right side of the umbilicus	Good overview of the abdominal cavity	Less good cosmetic result
Transverse	Transverse incision over the umbilicus with dissection of the muscles	▪ Good overview of the abdominal cavity and the diaphragm ▪ Extension, good possible ▪ Good cosmetic result	Dissection of the muscles
Costal margin	Incision about 2–3 cm parallel to the costal arch	▪ Good overview of the abdominal cavity and the diaphragm ▪ Extension, good possible	Less good cosmetic result
Pararectal	Pararectal incision on the right side going directly beside the rectus muscle	▪ Good extension possibility ▪ No muscle dissection	Less good cosmetic result
Gridiron	Transverse incision over the groin	Good cosmetic result	Muscle dissection necessary Extension, less possible
Pfannenstiel	Transverse incision over the symphysis	▪ Good overview of the pelvic organs ▪ Good cosmetic result	Extension, less possible

20.5 Common Bowel Surgical Techniques

- Small bowel anastomosis
  - End-to-end ileo-ileostomy, side-to-side ileo-ileostomy, and ileo-ascendostomy (Fig. 20.10)
  - Diversion anastomoses: Y-Roux, and omega with a side-to-side anastomosis (Fig. 20.11)
  - Stricturotomy (Fig. 20.12)
- Colon
  - Right hemicolectomy (Fig. 20.13)
  - Left hemicolectomy (Fig. 20.14)

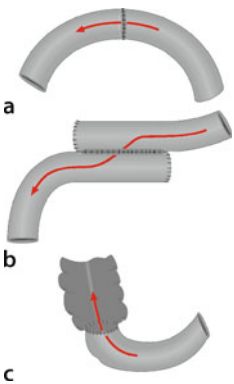


Fig. 20.10a–c Anastomoses techniques. **a** End-to-end ileo-ileostomy, **b** side-to-side ileo-ileostomy, **c** ileo-ascendostomy

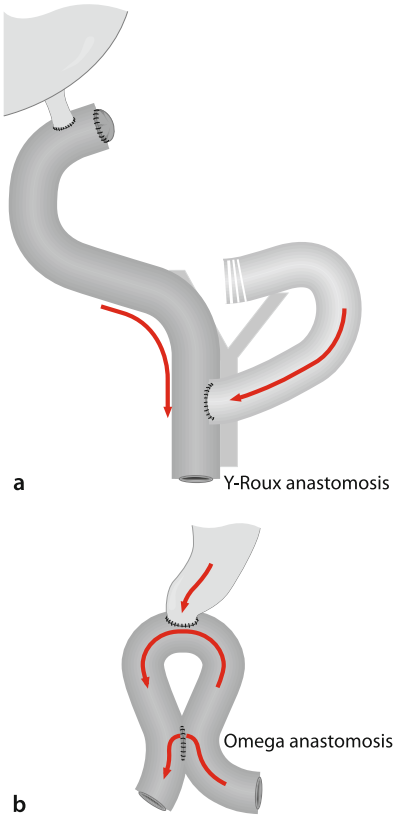


Fig. 20.11a,b Diversion anastomoses. **a** Y-Roux anastomosis, **b** omega anastomosis with a side-to-side anastomosis

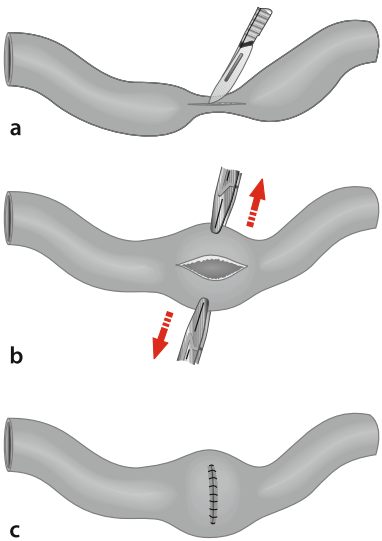


Fig. 20.12a–c Principle of stricturotomy

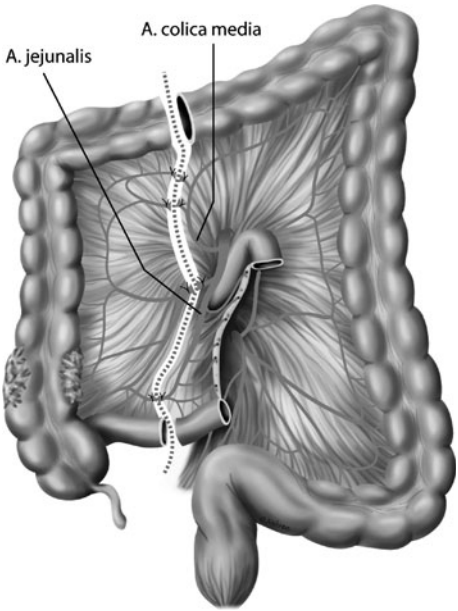


Fig. 20.13 Right hemicolectomy

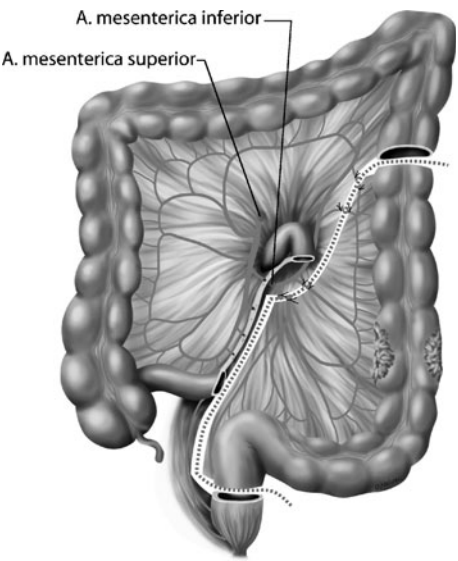


Fig. 20.14 Left hemicolectomy

21 Stomach and Duodenum

21.1 Infantile Hypertrophic Pyloric Stenosis

General considerations

- Hypertrophy of the pyloric muscle causes gastric outlet obstruction
- Incidence 1:250 live births, most common in Caucasian populations
- 4:1 male-to-female ratio
- Family history often positive, particularly sons born to affected mothers, indicating a polygenic pattern of inheritance
- Etiology largely unknown

Signs

- History
  - Typically a full-term baby, 3–6 weeks old
  - Nonbilious projectile vomiting soon after feeds
  - Rarely: failure to thrive, constipation, seizures
- Physical signs
  - Pyloric "tumor" in right upper quadrant
  - Visible peristalsis
  - Dehydration (late presentation)
- Metabolic abnormality
  - Hypochloremic hypokalemic metabolic alkalosis, paradoxical aciduria due to renal conservation of Na^+ leading to loss of H^+
  - CO_2 increase due to the respiratory compensation

Preoperative work-up

- Investigations
  - Full blood count, urea and electrolytes, blood gas

- Ultrasonography if test feed equivocal
- Diagnostic criteria: muscle thickness >4 mm, canal length >16 mm
- Upper GI endoscopy; if test feed and ultrasonography both equivocal ("swollen" pylorus visualized, differential diagnosis neonatal gastritis)
- Contrast study historical use only

- Management
  - Operation never an emergency
  - Resuscitation: airway, breathing, circulation (ABC)
  - Nasogastric tube, gastric washouts (0.9% saline)
  - Correct dehydration using glucose-electrolyte solution 0.45% saline, 10% dextrose, 20 $mmol \cdot l^{-1}$ K^+
  - 120–150 $ml \cdot kg^{-1} \cdot day^{-1}$
  - Monitor capillary refill, pulse, urine output, blood gases and electrolytes

Conservative therapy

- Atropine has been employed
- Although recent reports indicate benefits in some cases, surgery offers the best early definitive treatment

Operation: preparation

- Elective, following correction of metabolic derangements
- Preparation
  - Decompress stomach (nasogastric (NG) tube) prior to induction
  - General anesthesia with endotracheal intubation
  - Position: supine
  - Skin preparation with chlorhexidine solution, especially as the umbilicus can be a source of sepsis from *Staphylococcus aureus* or *S. epidermidis*

Operation: umbilical pyloromyotomy

- The operative steps are shown in Fig. 21.1
- Umbilical pyloromyotomy is increasingly utilized in modern surgical units
- A curvilinear incision is made in the superior umbilical fold

- The skin is undermined to expose the linea alba
- The abdominal cavity is opened vertically via the linea
- The stomach is grasped gently with forceps and a moist swab and delivered into the wound, exposing the pyloric tumor
- A serosal incision is made from the antrum to the prepyloric vein, along the anterior surface of the pylorus
- The muscle fibers are split with a Denis Browne divulsor, curved artery forceps or the blunt end of a scalpel handle, thereby exposing the bulging pyloric mucosa. Beware of injury to the duodenal fornix – distal aspect of myotomy
- Mucosal perforation can be detected by injecting 20–40 ml of air into the NG tube and noting the integrity of the myotomy (bile-stained fluid or air bubbles indicate injury). Any injury should be repaired immediately with fine absorbable sutures (6-0 PDS)
- The linea is closed in layers, using 4-0 Vicryl®. The umbilical skin is closed with 5-0 subcuticular Vicryl® sutures
- Cosmesis is excellent

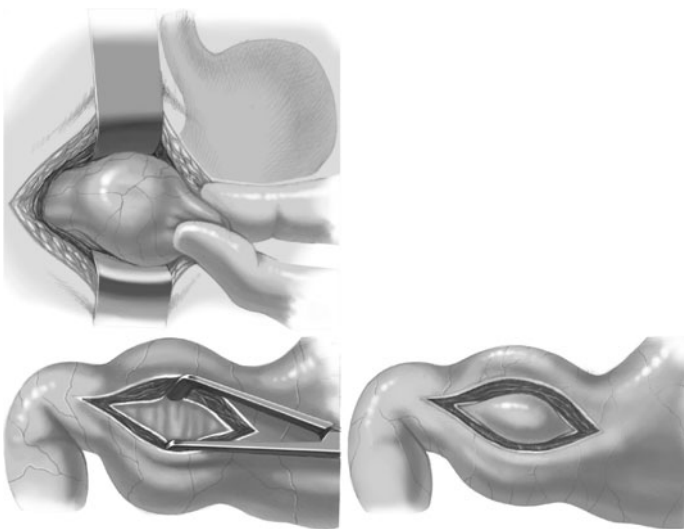


Fig. 21.1 Operative steps: hypertrophic pyloric stenosis

Operation: classical pyloromyotomy

- A transverse incision is made in the right upper quadrant
- The muscles are divided by electrocautery in the line of the incision
- The peritoneum is opened
- Operation proceeds as above, abdomen closed in layers with absorbable sutures

Operation: laparoscopic pyloromyotomy

- Available in centers that specialize in laparoscopy
- Technique as in classical pyloromyotomy, but carried out laparoscopically
- At the end of the operation, the stomach is inflated with air while the operation site is viewed constantly in order to see any mucosal breaches, which can be repaired laparoscopically
- Cosmesis is excellent and similar to that for umbilical pyloromyotomy

Postoperative care

- Many regimens have been described for feeding
- Early introduction of graded full-feeds as tolerated (warn parents about postoperative vomiting, usually resolves in 48 h)
- Persistent vomiting (severe gastro-esophageal reflux or incomplete myotomy)
- Trial with feed thickeners

Complications

- Wound infection and dehiscence (rare in specialist centers)
- Persistent vomiting (see above)
  - Incomplete myotomy: wait 10 days before reoperation
  - Ultrasonography and contrast studies are not useful as interpretation is difficult
- Mucosal perforation (1%–4%)
- Incisional hernia (rare)
- Mortality should be 0% in specialized modern units. Deaths have been reported in babies with multiple anomalies and severe coexistent medical conditions

Prognosis

- Excellent – an operation that has stood the test of time! (Ramstedt's pyloromyotomy was first described in 1911)

21.2 Gastrostomy

General considerations

- Gastrostomies are important in the management of children with surgical and nonsurgical conditions
- The main indication is the necessity for long-term feeding and constant gastric decompression, avoiding a nasogastric probe
- This is the case in the following diseases:
  - Long-gap esophageal atresia
  - Late operation in esophageal atresia
  - Esophageal stenosis
  - Duodenal atresia
  - Small bowel splinting
  - Swallowing problems experienced by neurologically impaired children
- Two main techniques are applied:
  - Open gastrostomy
  - Percutaneous endoscopic gastrostomy (PEG)

Operation: Gastrostomy

- The steps of the operation are shown in Fig. 21.2
- The child is positioned with a small roll behind their back to elevate the epigastrium
- A short transverse incision is made in the left epigastrium (first third of the imaginary line that runs from the umbilicus to the mid-portion of the left rib cage)
- The ventral surface of the stomach is identified
- Traction sutures are made, midway between the large and small curvature, preferably in the direction of the pylorus rather than the fundus

- Two concentric purse-string sutures are placed around the potential opening site
- The stomach is opened using electrocoagulation
- The catheter is introduced and the purse-string sutures are pulled together in order to form a chimney like structure around the catheter
- The catheter is brought out through the incision
- The stomach is anchored to the anterior abdominal wall
- After removing the catheter the fistula shrinks in 95% of cases spontaneously. An operation is necessary only if spontaneous closure fails

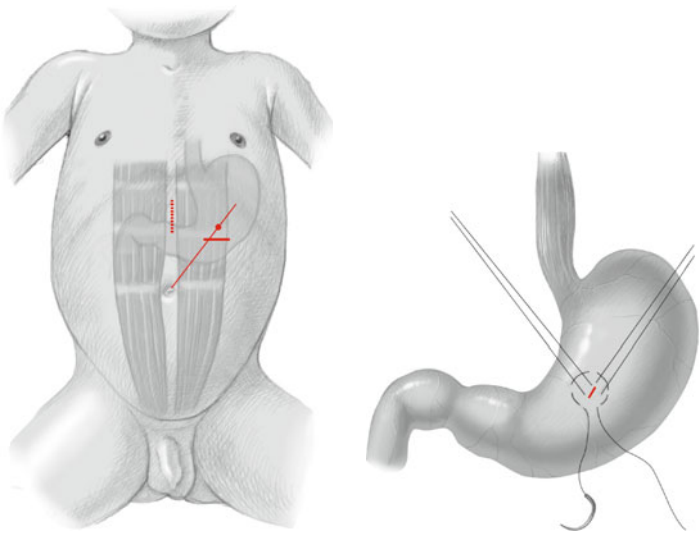


Fig. 21.2 Operative steps: gastrostomy

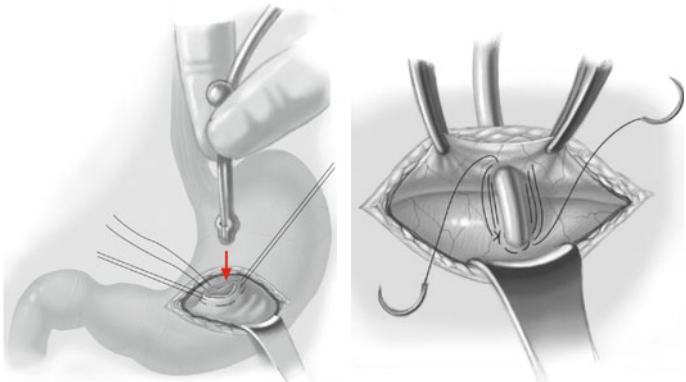


Fig. 21.2 *(continued)* Operative steps: gastrostomy

Operation: PEG

- The operative steps are shown in Fig. 21.3
- Endoscopy with the patient under general anesthetic
- Stomach insufflation
- Transillumination of the gastric anterior wall whilst digital pressure is applied to the proposed gastrostomy site
- Insertion of the cannula, which bears the loop, through the abdominal and gastric wall until it is visible for the endoscopist to grasp
- The loop is pulled out through the patient's mouth
- The catheter is attached to the loop, which is pulled back down through the esophagus into the stomach, to emerge across the gastric and abdominal walls
- Traction is applied to the catheter so that the gastric and abdominal walls are in loose contact
- The external crossbar is slipped over the catheter until it reaches the skin, avoiding pressure on the gastric mucosa or the skin

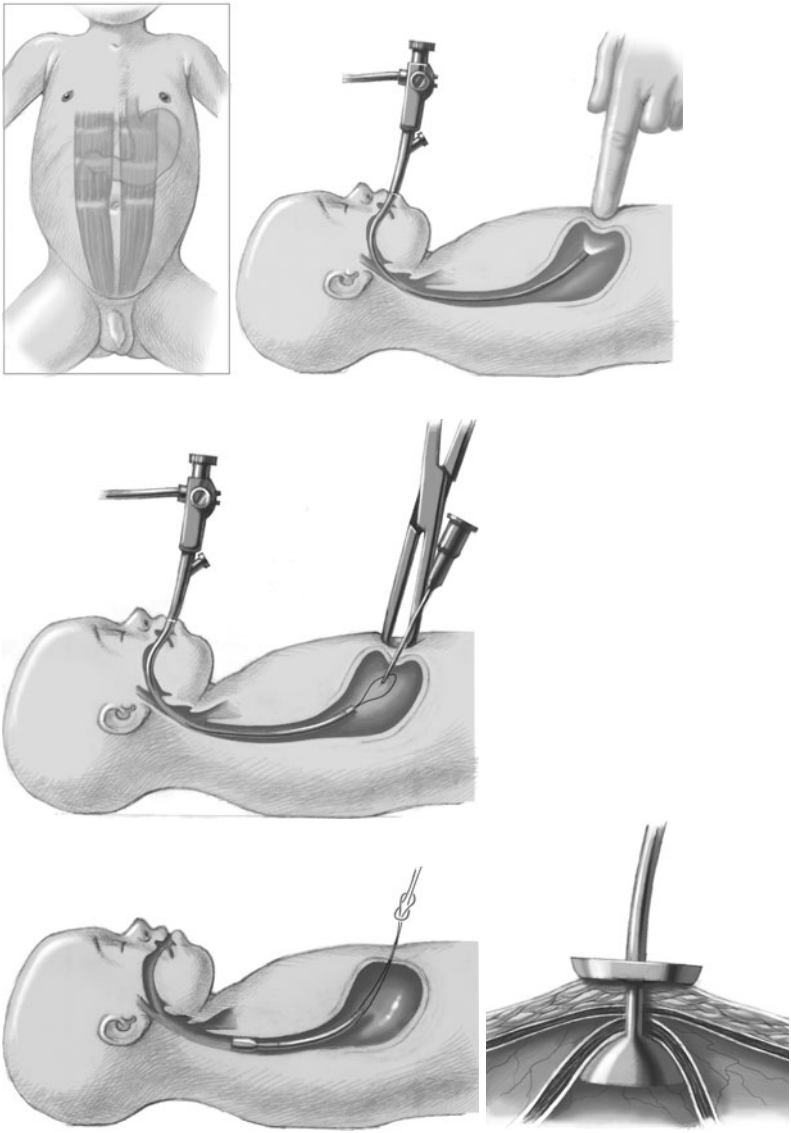


Fig. 21.3 Operative steps: percutaneous endoscopic gastrostomy (PEG)

Gastrostomy button

- A special button, commercially available, can be used either instead of the catheter in an open procedure or after a delay of some days when, the gastric and abdominal wall have fused (Fig. 21.4)
- The distance between the skin level and the stomach can be measured with the small-scale probe provided with the button
- The correct-sized button is then inserted
- The advantage of the button is that the patient has a closed system enabling better mobility and comfort

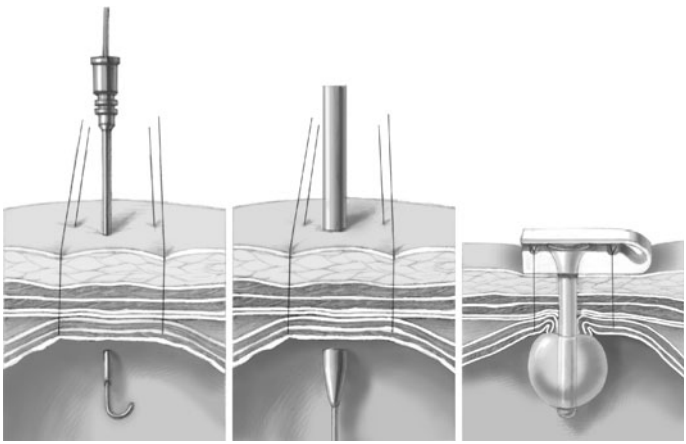


Fig. 21.4 Gastrostomy button

Complications

- Peritonitis in cases of disconnection of the gastric wall from the abdominal wall
- Leakage beside the catheter with irritation of the skin
  - The fistula tends to shrink within hours after catheter removal. The catheter is removed for 8–12 h and then reinserted into the smaller fistula

21.3 Duodenal Atresia and Stenosis

General considerations

- Incidence about 1:10,000 births
- Approximately 70% of congenital intestinal stenoses and 45% of atresias are found in the duodenal region
- 30% have trisomy 21 (Down's syndrome)
- Diagnosis is possible antenatally
  - 17%–57% have associated polyhydramnios
  - Antenatal ultrasonography may identify a dilated stomach and proximal duodenum (double-bubble)
  - Antenatal diagnosis should prompt a detailed search for associated anomalies and amniocentesis
- The majority of cases are diagnosed in the early newborn period
- Other associated anomalies include:
  - Congenital heart disease (~20%)
  - Malrotation (~20%)
  - Esophageal atresia and tracheo-esophageal fistula (~20%)
  - Genito-urinary anomalies (~9%)
  - Anorectal anomalies (~3%)
  - VACTERL association (**v**ertebral abnormalities, **a**nal atresia, **c**ardiac abnormalities, **t**racheoesophageal fistula and/or **e**sophageal atresia, **r**enal agenesis and dysplasia and **l**imb defects)

Classification

- Intrinsic
  - Complete atresia
  - Incomplete stenosis
- Extrinsic
  - Annular pancreas (most common)
  - Pre-duodenal portal vein
  - Malrotation, volvulus and Ladd's bands volvulus
- The obstruction may occur at any level of the duodenum (preampullary and postampullary); 85% of cases are distal to the ampulla of Vater

Signs

- 50% born prematurely with low birth weight (due to polyhydramnios)
- Majority present in first few hours of life with bile-stained vomiting
- Large volumes of fluid can be aspirated from the stomach (more than 20 ml)
- 20% may have preampullary atresia, therefore non-bile-stained vomiting
- Partial obstruction (e.g., "windsock" deformity) may present later – up to several years
- Vomiting leads to dehydration and electrolyte imbalance
- Fullness in the left upper abdominal quadrant
- Delayed passage of the meconium (in 25% of cases)
- Dehydration, hyponatremia, hypochloremia

Preoperative work-up

- X-ray with the typical findings
  - "Double-bubble" (a large air/fluid level in the stomach, a smaller one in the proximal duodenum)
  - No evidence of air in the rest of the abdomen
- An upper gastrointestinal contrast radiograph in cases of partial obstruction (to exclude duodenal stenosis or windsock, annular pancreas, duodenal web or malrotation with Ladd's bands)
- Associated anomalies – other investigations may be necessary, e.g., ECG
- Continuous orogastric (nasogastric) decompression
- Intravenous hydration (correction of dehydration, electrolyte imbalance, and metabolic alkalosis)
- Operation only if the patient's condition is stable

Operation: Kimura duodenoduodenostomy

- The operative steps are shown in Fig. 21.5
- Transverse supra-umbilical incision
- Mobilization of the ascending colon and the hepatic flexure of the colon to expose duodenum (Kocher manuever)
- Proximal duodenum and distal duodenum are mobilized (Kocher maneuver) to assess abnormality and determine operative repair

- A transverse incision is made in the proximal duodenum 1 cm above the atresia
- Care should be taken to avoid damage to the ampullary region
- A longitudinal duodenotomy of the same length is made in the distal atretic segment
- A diamond-shaped anastomosis is thus created
- Anastomosis is fashioned with a single layer of interrupted absorbable sutures (e.g., 6-0 or 5-0 PDS)

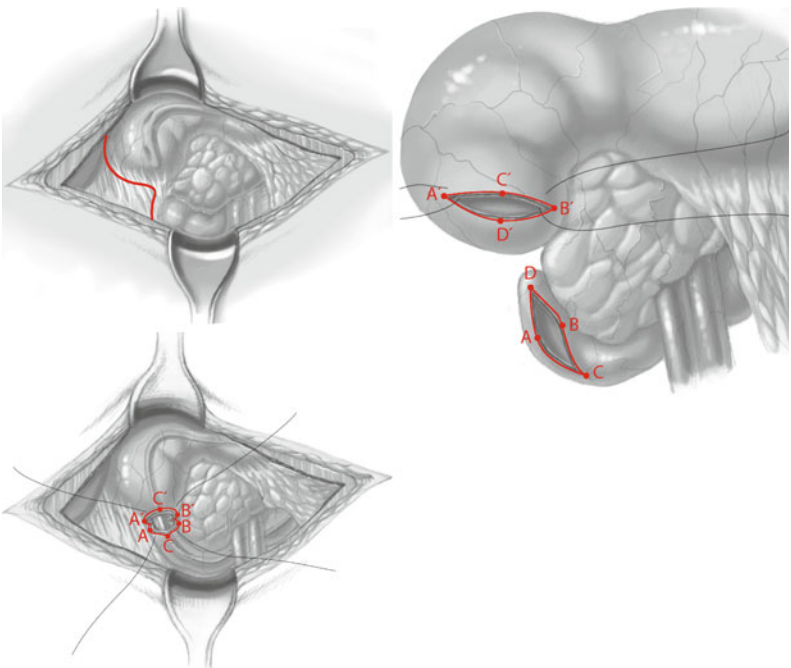


Fig. 21.5 Operative steps: duodenal atresia and stenosis

Operation: side-to-side duodenoduodenostomy

- Parallel transverse duodenotomies are made in the proximal and distal portions of the atretic duodenum
- The two ends are anastomosed in side-to-side fashion as a single layer

Operation: duodenojejunostomy

- This is the procedure of choice if the atretic segments are widely separated to create a tension-free anastomosis
- A duodenotomy is created in the proximal dilated duodenum as described above
- A loop of proximal jejunum is brought through a "window" created in the right mesocolon and anastomosed in side-to-side fashion to the proximal atretic segment. The defect in the mesocolon is closed with interrupted 5-0/6-0 sutures (PDS)

Operation: additional technical points

- Windsock abnormality
  - The operative steps are illustrated in Fig. 21.6
  - Suspect when there is no obvious atresia
  - Passage of a transpyloric feeding tube (8 F) with saline irrigation to localize the obstruction
  - An approximately 3-cm longitudinal incision is made along the transitional zone of the dilated portion and the narrow part of the duodenum
  - The membrane usually originates near the ampulla of Vater; care should be taken to avoid injury
  - The membrane may be either completely or partially excised
  - To perform partial excision a radial incision is made from the center of the membrane toward the duodenal wall. A rim of tissue (1–2 mm) is left circumferentially to reduce the risk of ampullary damage
  - The incision is closed transversely

- Saline should be injected distally (via a catheter) following repair to exclude the presence of distal atresia (rare)
- Check for coexistent malrotation (Ladd's procedure may be necessary)

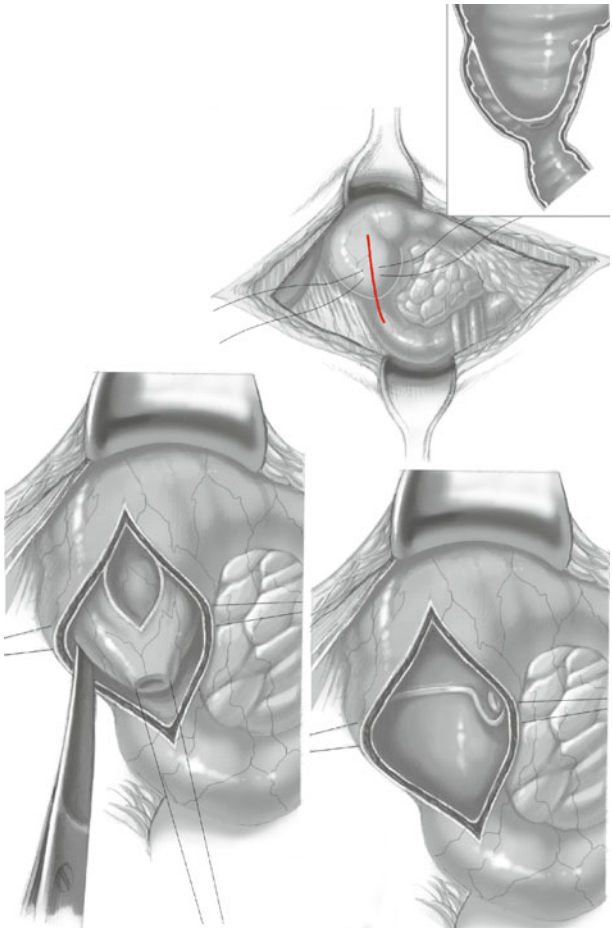


Fig. 21.6 Operative steps: duodenal atresia and stenosis; additional technical points

Postoperative care

- Feeding – duodenal dysmotility may require a period of total parenteral nutrition (TPN, via a neonatal long-line or rarely a Broviac catheter)
- Trans-anastomotic feeding tube may obviate requirements for TPN
- Graded introduction of enteral feeds as bowel motility recovers (minimal gastric aspirates)
- Prophylactic antibiotics for 48 h

Complications

- Early postoperative complications – anastomotic leak with intra-abdominal sepsis
- Long-term complications – recurrent abdominal pain, diarrhea, gallstone formation

Prognosis

- Excellent (95%) – associated anomalies influence prognosis, e.g., Trisomy 21
- Morbidity
  - Megaduodenum
  - Duodenogastric reflux
  - Gastritis and ulceration
- Mortality in patients with neonatal duodenal obstruction is 10%–15% (it depends on associated cardiac and chromosomal anomalies, and on prematurity)

22 Intestinal Congenital Malformations

22.1 Jejunoileal/Colonic Atresia

General considerations

- Atresias can occur at any level of the gastrointestinal tract
- Atresias result most likely after mesenteric vascular accidents caused by intrauterine intussusception, volvulus or internal hernia
- No extraintestinal-associated anomalies are consistently present
- Jejunoileal atresia is the most common cause of congenital intestinal obstruction in newborns (incidence 1:1000 live births)
- About 35% of jejunoileal atresia patients are significantly premature
- Differential diagnosis of jejunoileal atresia includes: meconium disease, meconium plug syndrome, malrotation, Hirschsprung's disease, and intussusception
- Colonic atresia is a rare congenital disorder (comprises 5%–10% of all intestinal atresias)
- Differential diagnosis of colonic atresia includes: malrotation with volvulus, meconium disease, Hirschsprung's disease, and megacystis hypoperistalsis syndrome

Classification

- Classification of jejunoileal atresia is outlined in Table 22.1 and Fig. 22.1
- The classification of colonic atresias is the same as that of jejunoileal atresias

Table 22.1 Classification of jejunoileal/colonic atresia

Type 1	Mucosal (membranous) atresia	Intact bowel wall and mesentery
Type 2	Cord atresia	Blind ends are connected by a solid fibrous cord
Type 3a	Gap atresia	Disconnected blind ends atresia (a portion of bowel and its associated mesentery are missing, leaving two blind segments of bowel)
Type 3b	Apple-peel atresia	Significant loss of bowel length and a large mesenteric defect
Type 4	Multiple atresia	Multiple atresias can be combinations of types 1–3

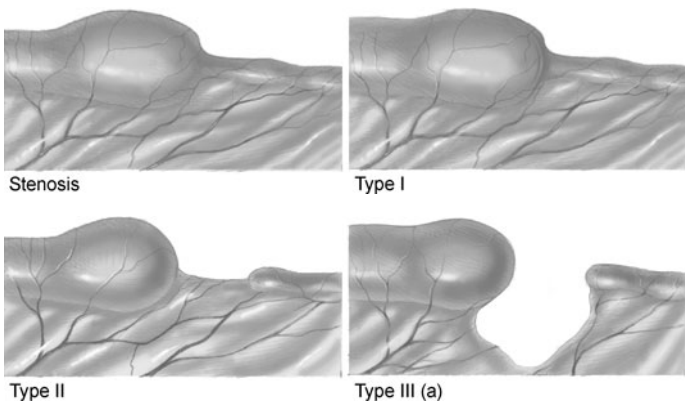


Fig. 22.1 Classification of jejunoileal atresia

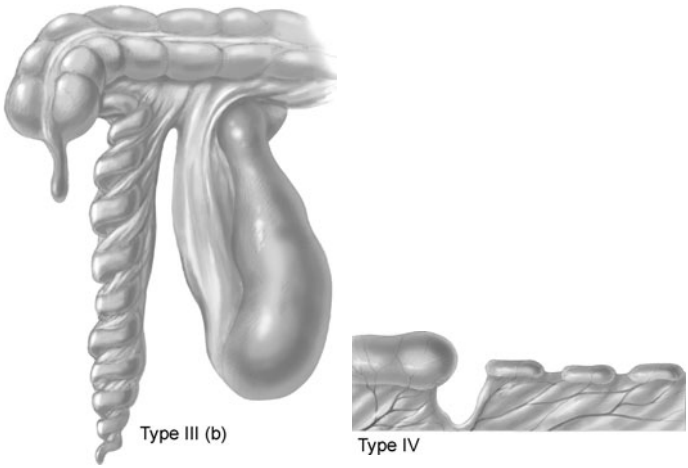


Fig. 22.1 *(continued)* Classification of jejunoileal atresia

Signs

- Polyhydramnios
- Dilated stomach and distended obstructed loops visible on ultrasonography
- Aspiration of large volumes of bilious fluid from the stomach
- Bilious vomiting (80% present on the first day of life, but in 20% it is delayed for 2–3 days
- The higher the obstruction, the earlier and the more forceful the vomiting
- Abdominal distension (the higher the obstruction, the less marked is the distension)
- Abdominal distension at birth is indicative of meconium ileus or peritonitis
- Loops of dilated bowel are frequently visible through the skin and palpable
- Failure to pass meconium (70% of patients fail to pass meconium on the first day of life)
- Dehydration and electrolyte imbalance

Preoperative work-up

- Free-drainage nasogastric tube of an adequate size (10 F) to prevent aspiration
- Intravenous hydration (correction of dehydration and electrolyte imbalance)
- Preoperative broad-spectrum antibiotics
- Patient must be warmed during transport and radiography studies
- Anteroposterior abdominal radiograph in the upright position: air-fluid levels and no gas in the lower part of the abdomen
- The number of air-filled intestinal loops increases with more distal atresia (all loops are dilated to about the same degree)
- A contrast enema (in colonic atresia patients)

Operation

- The operative steps are illustrated in Figs. 22.2 and 22.3
- A supraumbilical transverse incision (transecting the rectus muscles 2–3 cm above the umbilicus)
- Intraperitoneal fluid bacteriological examination
- Identification of proximal and distal atresias
- The lengths of the bowel should be carefully measured along the antimesenteric border (the normal length of the small intestine at birth is approximately 250 cm; in the preterm infant, 160–240 cm)
- Examination of the distal bowel for other sites of obstruction (injection of saline into the lumen of collapsed intestine)
- Partial resection or tapering of the dilated proximal bulbous tip
- Establishment of intestinal continuity with a single-layer primary anastomosis (contraindicated in cases of meconium ileus, peritonitis, and atresia type 3b)
- In case of isolated membranous atresia (type 1) – primary resection and end-to-end anastomosis
- In multiple atresias (when the atresias are close to each other) – resection of them all together and creation of one anastomosis is better than multiple anastomoses

- In right colon atresia – a primary anastomosis. In left or sigmoid colon lesions – a proximal colostomy and staged reconstruction
- In preterm infants with colonic atresia – a proximal colostomy
- In patients with colonic atresia and Hirschsprung's disease – a colostomy (colonic biopsies at operation are important)

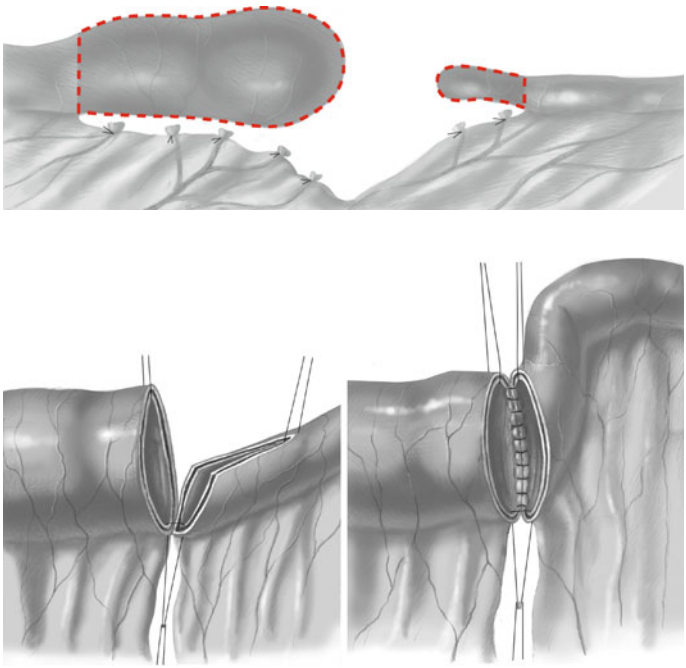


Fig. 22.2 Operative steps: jejunoileal atresia

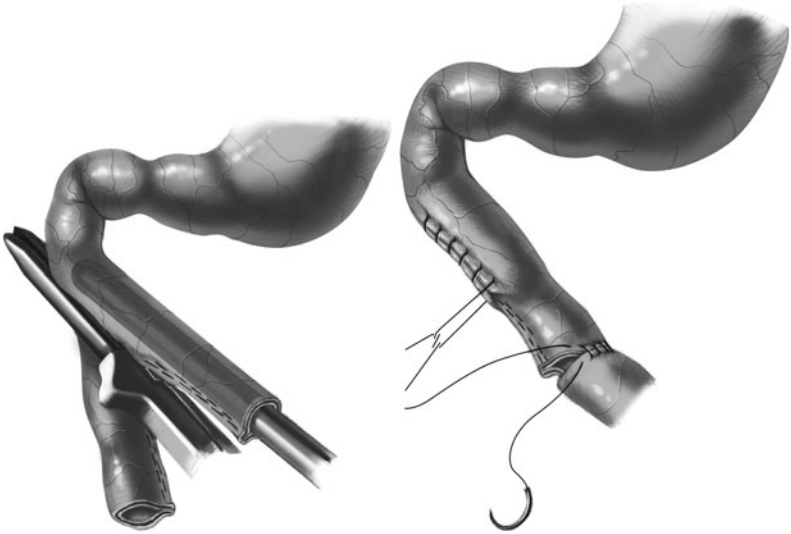


Fig. 22.3 Tapering duodenojejunal plasty

Postoperative care

- Postoperative antibiotic therapy (5–7 days or longer)
- Free-drainage nasogastric tube (for 5–6 days, longer for high jejunal atresias)
- Total parenteral nutrition (until a full diet is tolerated by the GI route)
- Transanastomotic enteral feeding 24–48 h after the operation
- Oral feeding (when the function of GI tract is normal, i.e., spontaneous bowel motions, the gastric drainage fluid is clear and less than 5 $ml \cdot h^{-1}$)
- In cases of jejunoileal atresia all patients should be tested for cystic fibrosis (a sweat chloride test and cytogenic testing for the delta F508 gene mutation)
- Closure of the colostomy and delayed anastomosis after 3 months

Complications

- Anastomotic stricture
- Leakage
- Proximal segment dysfunction

Prognosis

- Survival is 90%–95% (if an adequate bowel length remains and no complications occur)
- Mortality rate depends on birth weight, type of atresia (residual small bowel lengths), associated anomalies, and septic complications

22.2 Malrotation/Ladd's Bands

General considerations

- Malrotation includes any aberration of the gut position
- The incidence of malrotation in the general population is 0.5%
- Malrotation can be present throughout life without clinical signs
- Approximately 60% of malrotation patients present with signs in the neonatal period (40% in the first week)
- Every second to third patient with malrotation has associated anomalies (duodenal atresia, prune-belly syndrome, Hirschsprung's disease)
- Malrotation is an integral component of omphalocele, gastroschisis, and diaphragmatic hernia

Classification

- Embryological intestinal development in the first 12 weeks of gestation is very complicated and has three stages with a variety of clinical presentations (Table 22.2)
- The most common type of malrotation with duodenal obstruction is caused by aberrant peritoneal (Ladd's) bands extending from the colon across the anterior duodenum to the right abdominal wall
- Malrotation and volvulus must be quickly diagnosed and treated operatively to avoid gangrene of the midgut

Table 22.2 Embryological intestinal development and clinical presentation of malformation. (*CW* Clockwise, *cCW* counterclockwise)

Embryonic intestine (blood supply)	Organ	Stage					
		I		II		III	
		Gestational weeks 4–10		Gestational weeks 10–11		Gestational week 12	
		Normal	Malformation	Normal	Malformation	Normal	Malformation
Foregut (celiac trunk)	Stomach Duodenum			cCW 270°			
Midgut (superior mesenteric)	Ileum Jejunum Proximal colon	Physiologic umbilical hernia	Omphalocele Laparoschisis	Retraction of umbilical hernia Cecum cCW 270°	1[a] 2[b] 3[c] 4[d]	Colonic fixation	Mobile cecum
Hindgut (inferior mesenteric)	Distal colon Rectum						

[a]Nonrotation: failure of the normal counterclockwise (cCW) rotation of the midgut. The duodenum and small intestine remain in the right abdomen; the cecum and colon, on the left

[b]Malrotation I: arrested counterclockwise rotation at 180°. The cecum typically resides in a subhepatic position in the right hypochondrium, the duodenojejunal flexure to the right of the midline and the superior mesenteric

[c]Malrotation II: rotation of 180° CW in one plane and 180° cCW in the other. The colon is behind the duodenum and the superior mesenteric artery

[d]Hyperrotation: rotation > 270° cCW. The cecum is in the left abdomen near the spleen

Signs

- Malrotation in the neonatal period may present with acute strangulating ileus (acute midgut volvulus) or intermittent intestinal obstruction (chronic midgut volvulus)
- Clinical signs depend on the specific mechanism of obstruction and vascular occlusion
- Bilious vomiting, pain, and abdominal distension in cases of midgut volvulus
- Blood-stained vomit, bloody diarrhea, dehydration, shock and lethargy (indicating bowel strangulation with ischemia and necrosis)
- Intermittent abdominal pain, cyclic bilious (nonbilious) vomiting, and malabsorption (in chronic midgut volvulus patients)
- Anorexia nervosa (in older patients)
- Duodenal obstruction (bilious vomiting, dehydration, electrolyte and acid–base disturbance) in patients with Ladd's bands

Preoperative work-up

- Plain abdominal radiograph in malrotation with volvulus: double-bubble (distended stomach and proximal duodenum) and no air in the distal small bowel
- Radiographic airless abdomen (when the volvulus has created a closed-loop obstruction)
- Upper gastrointestinal contrast study – the abnormal rotation of the duodenojejunal loop (in simple malrotation) and an abrupt cut-off to passage of contrast (bird's beak) in the distal duodenum (in malrotation with volvulus)
- Contrast enema (for locating the position of the abnormal-sited cecum)
- Ultrasonography (an abnormal relationship of the superior mesenteric vein to the superior mesenteric artery)
- Nasogastric decompression
- Intravenous correction of dehydration and electrolyte imbalance
- Preoperative broad-spectrum antibiotics (in the acute volvulus patient)

- Operation as soon as possible after 1–2 h of intensive resuscitation (acute volvulus is a surgical emergency)
- Chronic midgut volvulus patients are prepared in the same way (preoperative preparation 4–12 h)

Operation strategy

- Due to the variability of malrotations there is no standard operative procedure
- The main aim is to bring the bowel in a position where a volvulus is improbable
- This is not always achieved if the bowel is brought to its normal position
- In most cases the safest position is a nonrotation one
- Prophylactic appendectomy is not necessary nowadays: the appendix can almost always be found laparoscopically

Operation: Ladd's procedure

- The operative steps are illustrated in Fig. 22.4
- A supraumbilical right-sided transverse incision
- Evisceration of the bowel and confirmation of the malrotation
- Untwisting of the volvulus in a counterclockwise manner, if present
- Dissection of the Ladd's bands
- Widening the mesenteric base (until the superior mesenteric artery and vein have no compression)
- Correction of duodenal obstruction (intraluminal duodenal obstruction must be diagnosed and corrected)
- In cases of malrotation and acute midgut volvulus with extensive intestinal gangrene – untwisting of the volvulus, warming, closure of the abdomen and a second-look after 12–24 h (when demarcation lines are clear), resection and anastomosis or stoma formation

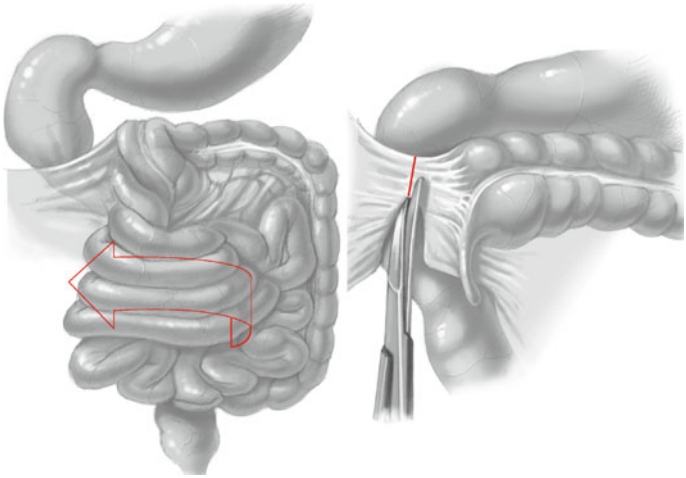


Fig. 22.4 Operative steps: intestinal malrotation

Postoperative care

- Broad-spectrum antibiotic therapy (depends on septic complications)
- Continuous nasogastric aspiration (2–3 days or longer)
- Parenteral nutrition
- Intravenous fluid and electrolyte support
- Oral feeding (depending on postoperative bowel function)
- Stoma closure and anastomosis formation (after 4–8 weeks)
- Postoperative treatment must be individualized to each patient

Prognosis

- Mortality depends on short-gut syndrome (prolonged parenteral nutrition) caused by volvulus and intestinal gangrene
- The incidence of adhesions with bowel obstruction is 5%–7%
- The recurrence rate for volvulus formation after Ladd's procedure is up to 5%
- Prognosis for premature neonates with peritonitis, shock, and sepsis is poor

22.3 Intestinal Volvulus

General considerations

- Intestinal volvulus is a condition in which the intestine twists around the mesentery
- Malrotation is a common cause
- During the process of embryonic gut rotation and fixation, three conditions develop where volvulus is possible:
  - Nonrotation (the midgut volvulus risk is high)
  - Abnormal rotation of the cecocolic limb (the volvulus risk is very high)
  - Abnormal rotation of the duodenojejunal limb (the volvulus risk is low)
- Volvulus during intrauterine life can lead to ischemic atresia
- Intestinal volvulus may occur later in life in situations involving postoperative or congenital adherences, and tumor masses

Signs

- Acute abdominal pain is the most typical clinical feature
- Vomiting is uncommon
- Presence of blood in stools is a late finding
- General status deteriorates rapidly
- Abdominal auscultation demonstrates hyper-peristaltic activity in the early phase followed by complete silence later

Preoperative work-up

- Abdominal plain X-ray in erect position shows abnormal distribution of intestinal gas
- Ultrasonography demonstrates thickening and clumping of the bowel loops
- Doppler ultrasonography may show a decreased flow rate in the mesenteric artery (total volvulus)
- Complete blood count and fluid/electrolyte assessment should be performed with a view to repletion

Operation

- A nasogastric tube (siphon) should be placed for luminal decompression
- A transverse supraumbilical abdominal laparotomy: the base of the mesentery should be located and the volvulus reduced
- After reduction the intestine should be rinsed with warm saline solution and then inspected to document tissue viability and detect abnormal positions
- Rotational anomalies should be treated and congenital adherences dissected
- The intestine should be re-inspected and any necrotic bowel encountered should be excised
- If possible, intestinal continuity should be re-established by primary anastomosis
- A dual enterostomy may be necessary after resection of a significant length of bowel

Postoperative care

- Free-drainage nasogastric tube
- Antibiotic therapy is required only in cases of intestinal resection (antibiotic prophylaxis in other cases)
- Parenteral feeding should be maintained until intestinal transit resumes

Prognosis

- Prognosis is good following early recognition and prompt treatment
- Short gut syndrome is a major risk after extensive bowel resection

22.4 Duplications of the Gastrointestinal Tract

General considerations

- Duplications can occur at any location along the gastrointestinal tract
- Duplication cysts are the most common

- Gastrointestinal tract duplications are lined with intestinal epithelium and their walls have smooth muscles
- Communication with the intestinal tract is possible
- Antenatal diagnosis is possible for cystic structures
- Association with other malformations is common (50% of cases), e.g., vertebral abnormalities

Classification

- Duplications of the esophagus are usually noncommunicating (intramural cysts). The cyst and mucosa must be removed followed by closure of the esophageal wall. The vagus nerve should be dissected and protected
- Duplications involving the stomach are rare. Most are located along the greater curvature. An auto stapling suture device can usually be used. Closure can easily be checked by insufflating air via a nasogastric tube
- Duplications of the duodenum are also uncommon. They may be communicating or noncommunicating. Embedding in the pancreas is possible. The surgeon must bear in mind possible relations with biliopancreatic ducts
- Duplications of the small intestine are typically small cystic formations
- Types of gastrointestinal tract duplications are illustrated in Fig. 22.5

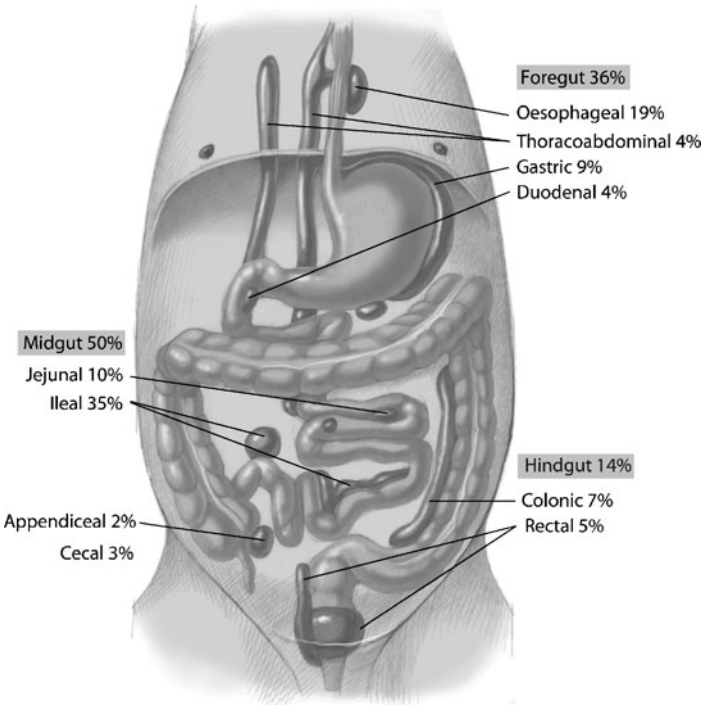


Fig. 22.5 Gastrointestinal tract duplications

Signs

- Often occur in newborns and depend on size and location

Preoperative work-up

- Abdominal X-ray demonstrating abnormal intestinal gas distribution (indirect sign)
- Ultrasonographic examinations can depict cystic formations
- Scintigraphy is useful for detection of ectopic gastric mucosa
- MRI especially if duplication of the esophagus and rectum is suspected

- Gastrointestinal endoscopy is indicated if duplication of the esophagus and rectum is suspected
- Esophagogastric and intestinal transit studies as well as retrograde enema may be useful depending on the location

Operation

- The operative steps for ileal or colonic duplication are shown in Fig. 22.6
- Antibiotic therapy is administered at the time of anesthesia induction (prophylactic)
- The goal of treatment is to achieve complete resection of mucosa, lining the duplication with no sequels to the normal intestine
- Exposure and surgical techniques are dictated largely by anatomic location
- Transverse laparotomy is suitable for abdominal duplications, supraclavicular exposure for duplications of the upper esophagus, and posterolateral thoracotomy for duplications of the distal esophagus
- Duplications must be completely resected
- Primary anastomosis is generally feasible if removal of the adjacent intestine is required
- Segmental resection followed by primary anastomosis may be necessary for deeply embedded cystic duplications
- Mucosal stripping is indicated for extensive tubular duplications. It can be achieved through sero-muscular incisions placed along the duplication, so that the mucosa can be peeled away like a tube. Attention must be paid to peptic ulcers
- Single duplications of the colon should be removed leaving the mesocolon and the ileocecal junction intact if possible

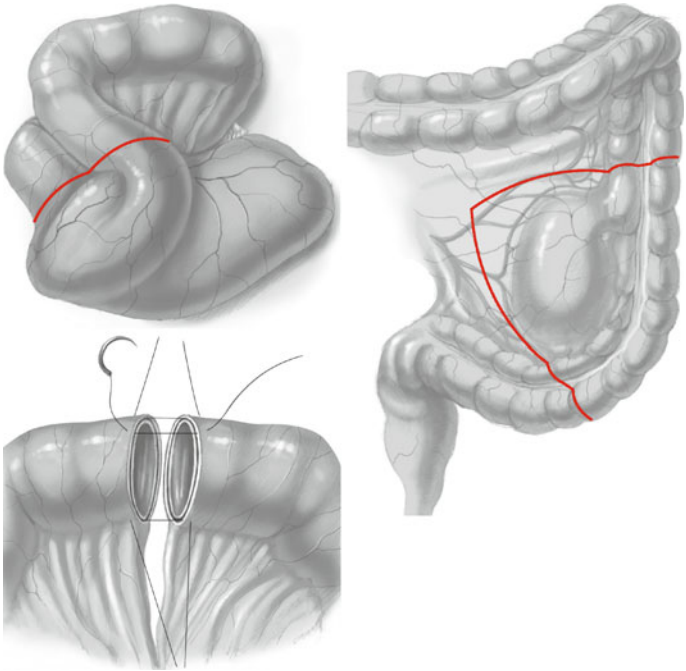


Fig. 22.6 Operative steps: ileal or colonic duplication

Postoperative care

- Short-term parenteral feeding
- Antibiotic therapy is required only in case of intestinal resection

Prognosis

- Prognosis depends mainly on the type of duplication and the presence or absence of communication with the adjacent intestine

22.5 Stoma Complications

General considerations

- Most stomas in childhood are temporary
- However, complications during this period of transition are still not uncommon
- There are many different techniques and modifications
- Meticulous technique reduces complications

Complications

- Stoma stenosis
- Stoma necrosis
- Stoma prolapse
- Peristomal hernia
- Stoma retraction
- Enterocutaneous fistula
- Skin problems (dermatitis, erosions)
- Stoma decubital ulcer

Operation

- In stenosis – widening of opening through the abdominal wall, i.e., by splitting the fascia
- In necrosis – revision with resection and exteriorization of normal vascularized bowel
- In prolapse – revision with retraction and anchoring of the prolapsed bowel, sometimes after resection
- In hernia – revision with resuturing of the peritoneum and narrowing of the fascial defect
- In retraction – revision with bowel mobilization and formation of a new stoma
- In fistula – usually no treatment
- In skin problems/ulcer – careful skin care (best prophylactically)

Prognosis

- Fairly high morbidity, but most complications can be successfully dealt with

22.6 Short Bowel Syndrome

General considerations

- A condition in which the absorptive area of small bowel is too small, leading to insufficient absorption of nutrients
- There are large individual variations, but survival is possible if the ileocecal valve is intact and more than about 20 cm of functioning small bowel is present
- Survival is possible even if the ileocecal valve is missing and more than about 50 cm of functioning small bowel is present
- More common in infants and young children (bowel atresia, neonatal volvulus, necrotizing enterocolitis, gastroschisis), later on, e.g., Crohn's disease
- Loss of part of the ileum has more severe effects than loss of part of the jejunum
- Main purpose is to prevent short bowel syndrome

Physiology

- Figure 22.7 shows the main sites of resorption in the small bowel

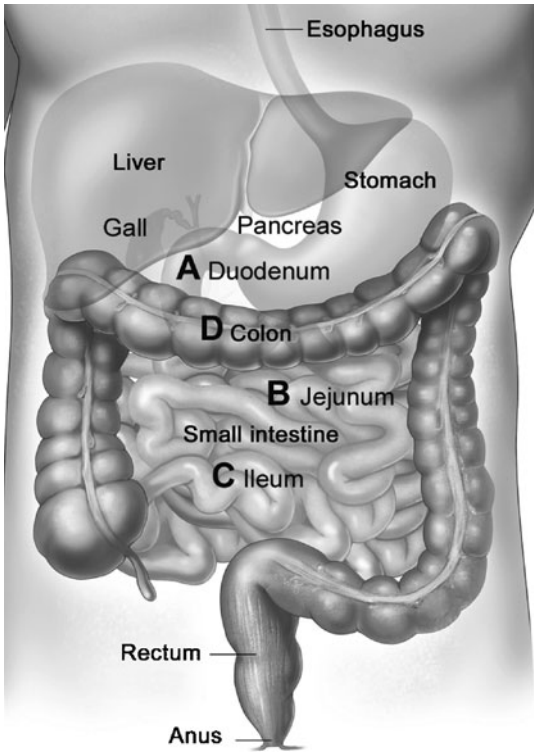


Fig. 22.7 Resorption sites in the small bowel. *A* Electrolytes (Fe, K, Mg), *B* lipophilic vitamins (A, D), protein, fats, monosaccharides (glucose, xylose), disaccharides, hydrophilic vitamins (C, folate, pyridoxine, riboflavin, thiamine), *C* vitamin B12, *D* water

Signs

- Diarrhea, steatorrhea
- Deficiency problems may arise because of defective absorption (e.g., electrolytes, vitamins, fatty acids, weight loss, retarded growth)

Conservative therapy

- Initially total parenteral nutrition, but early enteral feeding to stimulate lengthening and increase the absorptive area (reactive hyperplasia)
- Vitamins, electrolytes, antibiotics

- Gradual adjustment of enteral feeding guided by how well it is tolerated by the bowel
- H_2-receptor blockers (reduce acid hypersecretion), loperamide (decrease bowel motility), somatostatin (reduce pancreatic/biliary secretion), cholestyramine (reduce steatorrhea)
- Expected bowel adaptation occurs during the first 2–3 years of medical therapy

Complications of total parenteral nutrition

- Incorrect placement of central lines
- Infection/sepsis
- Metabolic complications
- Liver damage
- Nephrolithiasis
- Cholelithiasis
- Various abnormal levels (vitamins, electrolytes, trace elements, fat, carbohydrates, proteins, water, etc.)

Operation: emergency procedure

- The easiest and quickest operation should be performed; a stoma at the site of perforation is mostly sufficient
- During the primary operation bowel resection should be kept to a minimum
- When in doubt leave questionable parts and perform a planned second-look operation

Operation: tapering

- The operative steps are shown in Fig. 22.8
- In cases of small bowel dilatation over a short bowel segment tapering could be applied
- This method enables improved peristalsis
- The disadvantage of the method is the loss of mucosa

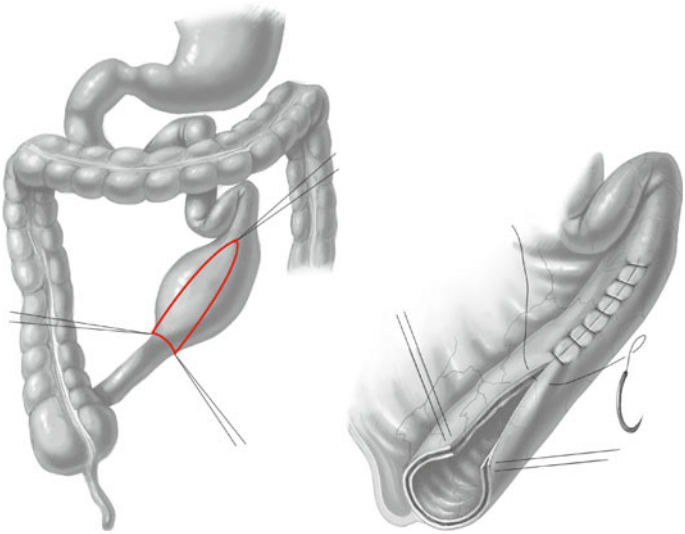


Fig. 22.8 Operative steps: ileal tapering

Operation: Bianchi procedure

- The operative steps are shown in Fig. 22.9
- Bianchi intestinal loop lengthening (1980) takes advantage of the fact that the mesenteric vessels supply each half of the bowel separately
- The mesentery is divided, the bowel is split longitudinally
- The bowel is formed into two separate tubes, which are then joined end to end

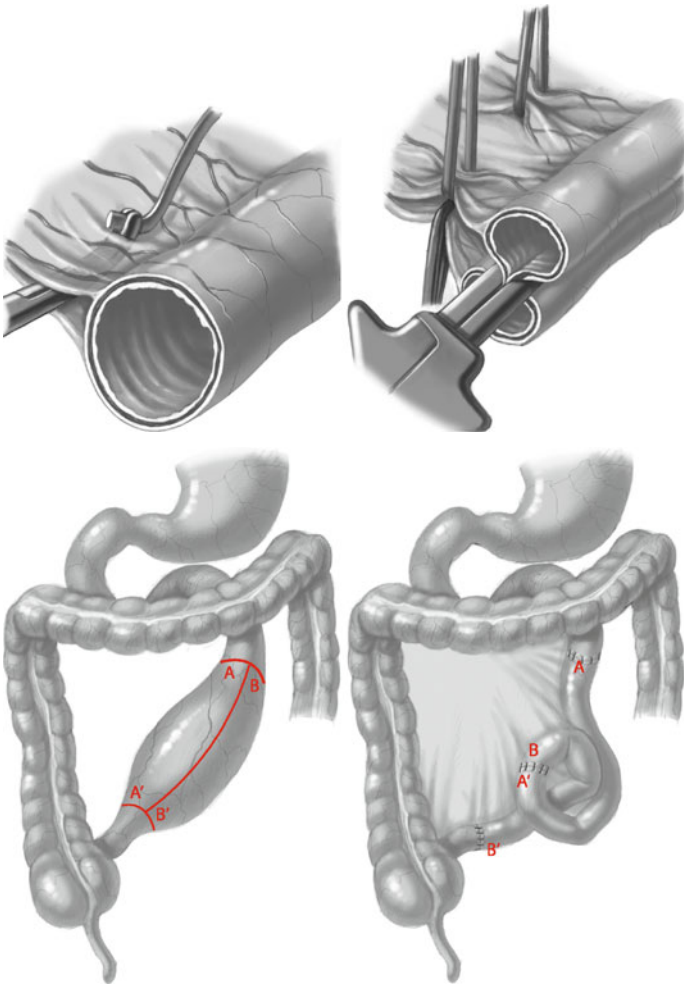


Fig. 22.9 Operative steps: Bianchi procedure

Operation: bowel transplantation

- Small bowel transplantation, with or without simultaneous liver transplantation, has become a valuable alternative recently. Problems with organ shortage, rejection, etc. remain

Postoperative care

- Usual postoperative measures after abdominal operations should be taken
- Regarding nutrition, conservative treatment principles are followed
- Parents are to be taught to manage the central line and infusion pump
- Parents are to be taught to manage the stoma
- Close follow-up necessary

Prognosis

- Depends on the primary condition (length of remaining bowel? ileocecal valve left? sufficient adaptation to oral feeding?)
- Very high morbidity

PART III
SPECIFIC DISEASES

23 Intestinal Diseases

23.1 Meconium Ileus

General considerations

- The most common reason for a meconium obstruction is cystic fibrosis (CF)
- Present in 8%–10% of CF patients at birth
- Accounts for 9%–10% of all neonatal intestinal obstructions
- Uncomplicated cases show impacted meconium mostly at the ileocecal valve
- Complicated cases include:
  - Volvulus
  - Perforation
  - Peritonitis with sepsis
- Antenatal diagnosis is feasible based on ultrasonographic findings: calcified or hyperechogenic bowel, dilated fetal bowel, and areas of calcification throughout the peritoneal cavity in cases of antenatal perforation

Signs

- Signs depend on degree of gastrointestinal obstruction and complications
- Significant abdominal distension may develop during the neonatal period
- The venous network of the abdominal wall may be highly developed
- Ascension of the diaphragm due to meteorism, results in dyspnea

- General status progressively deteriorates with incipient sepsis in cases of perforation due to a parent rapinal procesus
- In cases of perforation the scrotum or labia may have a greenish discoloration due to a patent vaginal procesus

Preoperative work-up

- Anterior abdominal plain X-rays in erect (suspension) and decubitus positions demonstrate a soap bubble appearance in the right part of the abdomen and diffuse calcification or localized intra-abdominal calcification with air-fluid levels throughout the cavity in cases of perforation
- Ultrasonography shows a calcified or hyperechogenic distal intestine, thickened bowel loops, and intestinal dysmotility
- Retrograde hyperosmolar water-soluble enema confirms the diagnosis (microcolon, meconium impaction in the ileum) and can help to resolve intestinal obstruction (repeated if necessary) in 80% of cases if no complications occur
- In cases of persistent obstruction placement of a nasogastric tube, repletion of fluid and electrolyte deficiencies, and surgical treatment should be undertaken as soon as the general condition allows
- An adequate supply of blood should be ordered for potential transfusion since dissection is often associated with massive blood loss in cases of meconium peritonitis

Operation strategy

- Operation is indicated if enemas fail to relieve obstruction
- The objective of the surgical treatment is complete evacuation of meconium proximal and distal to the level of obstruction
- Enterotomy and irrigation using warm saline suffice in most cases
- Temporary decompression of the dilated bowel segment may be needed
- Surgical exploration in cases of antenatal perforation may reveal a large cyst surrounded by a thick membrane containing calcifications or a pseudo-capsule formed by the adherent bowel loops

- Nonviable intestinal segments should be resected. The extent of resection is usually limited due to the resilience of the newborn's intestine. In this case enterostomy (oral and caudal stoma) is a more suitable procedure than primary anastomosis if local conditions or the patient's general status is unstable
- Drainage is unnecessary

Operation: double-barreled ileostomy (Mikulicz)

- The operative steps are illustrated in Fig. 23.1
- Resection of the diseased bowel segment, as little as possible
- The two loops are brought up to the skin side-to-side and sutured to the skin
- Preferably as a nipple valve (see below)

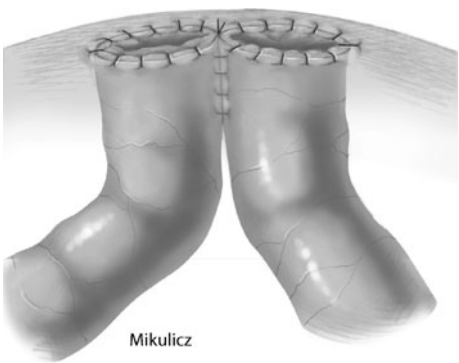


Fig. 23.1 Mikulicz stoma

Operation: Bishop–Koop stoma

- The operative steps are illustrated in Fig. 23.2
- Based on the principle of a Y-Roux anastomosis
- The distal limb is sutured to the skin as an ileostomy
- The proximal limb is sutured as an end-to-side anastomosis to the distal limb

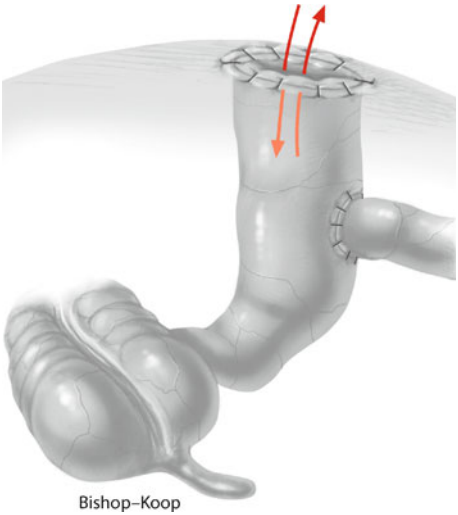


Fig. 23.2 Bishop–Koop stoma

Operation: Santulli stoma

- The operative steps are illustrated in Fig. 23.3
- Based on the principle of a Y-Roux anastomosis
- The proximal limp is sutured to the skin as an ileostomy
- The distal limp is sutured as an end-to-side anastomosis to the proximal limb

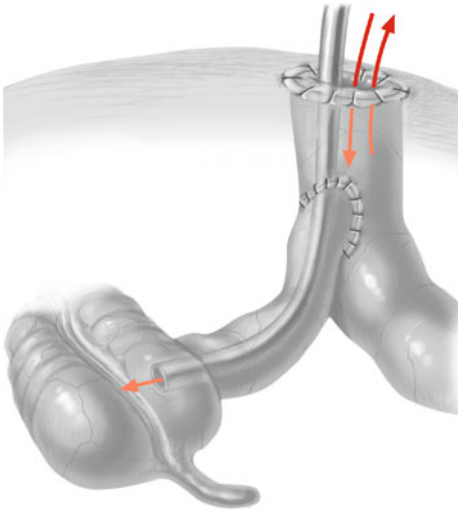


Fig. 23.3 Santulli stoma

Operation: terminal ileostomy

- A 2- to 3-cm-long intestinal segment is used with a serosal surface free of fat
- The seromuscular wall is sutured to the fascia in four quadrants
- The edge of the stoma is then sutured to the skin creating a nipple (Fig. 23.4)

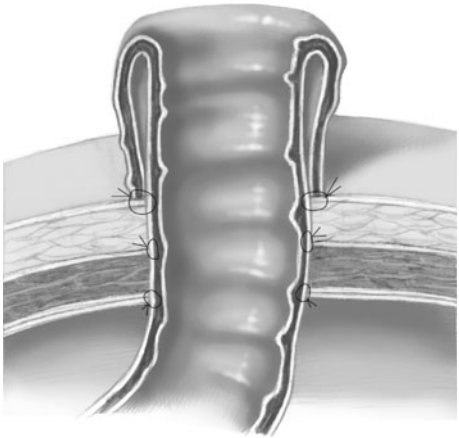


Fig. 23.4 Nipple ileostomy

Postoperative care

- Intensive care is required
- A nasogastric tube (siphon) is maintained
- Parenteral feeding is maintained until peristalsis begins
- Enteral feeding with transfer of chyle from the oral stoma to the caudal stoma can be used if problems occur during parenteral feeding
- Physiotherapy is necessary to avoid pulmonary complications
- Gastrointestinal continuity should be re-established as soon as possible

Prognosis

- Outcome depends on the severity of the obstruction and the presence of peritonitis, birth weight, associated malformations, and the severity of the initial disease if the patient has cystic fibrosis
- Perforation does not always lead to complications

23.2 Necrotizing Enterocolitis

General considerations

- Necrotizing enterocolitis (NEC) is a therapeutic emergency affecting predominantly premature infants
- It is a disease of paradoxes and its etiology remains unknown. The most likely mechanism involves gastrointestinal vascular compromise resulting in bacterial invasion of the portal venous system
- Onset of NEC usually occurs during the first or second week of life after beginning oral feeding in newborns weighing less than 1500 g
- The most common locations are the distal ileum and right colon
- Surgical intervention is required in more than one-third of cases
- Deterioration may be sudden
- Differential diagnosis include volvulus, meconium perforation, and other ileal obstructions

Signs

- Signs of intestinal occlusion (meteorism, gastric secresion increase, transit stop)
- The abdomen is red and distended with extensive collateral circulation resulting in a bright edematous appearance
- Peritonitis is either localized (abdominal mass) or generalized
- Ascension of the diaphragm due to meteorism, results in dyspnea
- Infants are severely ill and deteriorate rapidly
- Shock and sepsis

Preoperative work-up

- Abdominal plain X-rays in erect or lateral and decubitus positions demonstrate pneumoperitoneum, intestinal pneumatosis, and air in the portal vein as a late sign
- Ultrasonography demonstrates intraperitoneal fluid, thickened bowel loops, and bowel and bowel and portal pneumatosis (early sign)

Conservative therapy

Aggressive medical treatment should be started promptly

- Bowel rest with placement of a nasogastric tube for decompression (suction if needed)
- Total parenteral feeding
- Broad-spectrum antibiotic therapy
- Physical, radiographic, and/or ultrasonographic examination should be performed every 6 h for the first 48 h in the ICU

Operation

- Surgery should be undertaken if there is clinical deterioration, if radiographic or ultrasonographic investigations indicate perforation, or if the patient shows the above signs during medical treatment
- Exposure is obtained by transverse supraumbilical laparotomy
- The operative strategy depends on the extent of involvement, with the goal of performing the simplest and most effective procedure necessary to save the child's life
- Direct suture and closure or resection and primary anastomosis may be adequate if the perforation is small with minor peritonitis
- Resection of necrosis with creation of a dual enterostomy (see Fig. 23.1) at the most proximal site followed by resection of nonviable bowel segments distally may be necessary if perforation is associated with severe peritonitis and deterioration of general status
- Resection should be limited to frankly necrotic zones with the possibility of a second-look procedure 12–24 h later
- Simple drainage and abdominal perfusion may be necessary to stabilize low-weight infants (<1500 g) in poor general condition

Postoperative care

- Intensive care is required
- The nasogastric tube is maintained
- Parenteral feeding is maintained until peristalsis begins
- Enteral feeding with transfer of chyle from the oral stoma to the caudal stoma can be used if problems occur during parenteral feeding

- Gastrointestinal continuity should be re-established as soon as possible, however after a minimum of 6 weeks as secondary strictures may occur during this time. The size of the lumen should be checked along the full length of the intestine

Prognosis

- The overall survival rate is 70%–80%
- A short length of small intestine is a predisposing factor for late complications

23.3 Meckel's Diverticulum

General considerations

- Meckel's diverticulum is a remnant of the vitelline duct that connects the midgut to the yolk sac in early embryonic life
- Failure of embryonic obliteration of the vitelline duct (usually after the sixth gestational week) occurs in about 2% of the population and can be considered as a variant of persistent omphalomesenteric duct (distal part)
- Ectopic gastric tissue is present in about 40% of cases (top or bottom of the diverticulum)
- Most school-age patients are asymptomatic and the likelihood of complications is small for older children

Signs

- The most common presentation is gastrointestinal bleeding with melena. If bleeding is severe, stools may be bright red and hematemesis may occur
- Chronic anemia and acute or recurring abdominal pain (diverticulitis) occur in some cases
- Meckel's diverticulum can be the leading point for intussusception
- Vascular abnormalities involving the mesentery of the diverticulum can lead to subocclusion or occlusion of the small intestine

Preoperative work-up

- Abdominal plain X-rays can demonstrate indirect signs of occlusion
- Depending on the size of the abnormality, ultrasonography can be useful
- Technetium 99m radioisotope scanning is performed in order to visualize gastric mucosa (positive result only enhances the diagnosis)
- Complete blood cell count with blood transfusion if necessary
- Prophylactic antibiotic therapy is performed immediately before surgery

Operation

- The operative steps are shown in Fig. 23.5
- Successful treatment can be performed by laparoscopy via the transumbilical route. The diverticulum is extirpated through the umbilical route in most cases, especially if intra-abdominal resection cannot be performed using a stapling device
- Resection should be made crosswise in relation to peristaltic movement to avoid residual stenosis or leaving the base of the diverticulum in place
- If necessary, transverse laparotomy may be carried out via an incision through McBurney's point. After visualization (40–60 cm in front of the ileocecal valve), the diverticulum should be resected along with its base
- For a short-based Meckel's diverticulum, resection may be carried out in the longitudinal direction; however, closure should be performed in the transverse direction by separate suture points. Alternatively use a transverse anastomosis (TA) stapler
- For a diverticulum associated with a broad base or diverticulitis, partial resection of the small intestine is required followed by endtoend anastomosis
- For a diverticulum associated with intussusception, the resection technique depends on the size of the implantation base. In cases of necrosis, the small intestine must be resected followed by endtoend anastomosis
- If Meckel's diverticulum is found incidently, resection is recommended for newborns in all cases and in toddlers depending on signs and local findings; a persistent vitelline duct remnant or palpable ectopic tissue is a good reason to remove the diverticula

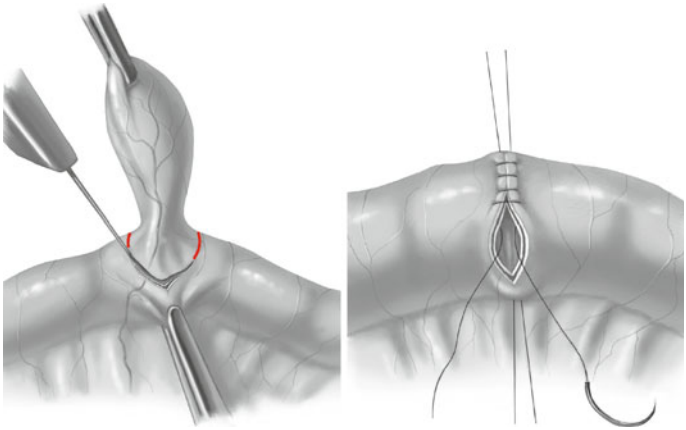


Fig. 23.5 Operative steps: Meckel's diverticulectomy

Postoperative care

- Antibiotic therapy may be administered depending on local findings
- If gastrointestinal anastomosis has been performed, parenteral feeding should be continued until bowel motion resumes

Prognosis

- Prognosis is good depending mainly on the volume of blood lost during initial presentation
- Postoperative morbidity is low

23.4 Intussusception

General considerations

- Intussusception is an acute life-threatening condition in which one segment of the intestine (intussusceptum) enfolds into an adjacent distal segment (intussuscipiens)
- In most cases intussusception involves enfolding of the terminal ileum into the colon

- It typically occurs during the first year of life in boys
- It is the most common cause of intestinal obstruction in this age group
- Prolonged intussusception leads to ischemia and bowel necrosis
- The leading point usually contains thickened hypertrophied lymphoid patches (associated with viral disease)
- Association with Meckel's diverticulum, polyps, and/or tumor is less frequent
- Postoperative invagination (hyperperistaltism) is difficult to diagnose
- Intussusceptum may be carried into the transverse colon and even reach the rectum

Signs

- Predisposing factors include upper airways infection, enteritis with diarrhea, and surgical intervention (heart and abdominal surgery)
- Onset is characterized by the sudden appearance of intermittent paroxysmal crampy abdominal pain
- The child initially acts normal between the cramps, but then becomes lethargic
- Palpation often reveals a sausage-shaped intra-abdominal mass
- Bloody rectal discharge after rectal examination (currant jelly stools) is a late finding
- Vomiting is a sign of occlusive syndrome (the first one is reflux)
- Prolonged intussusception results in dehydration and deterioration of general status (lethargy)

Preoperative work-up

- Abdominal X-ray demonstrates abnormal distribution of intestinal gas
- Obstructive pattern is a late radiographic finding
- Ultrasonography shows the target (anterior) and pseudo-kidney (profile) sign
- Ileo-ileal intussusception can also be seen by ultrasonography (experienced radiologist)
- Contrast enema shows failure of gas contrast to reflux in the small bowel and the chalice or lobster claw sign
- Repletion of fluid/electrolyte deficiencies is necessary in late cases

Conservative therapy

- Nonsurgical reduction of the intussusception can be attempted if diagnosis is achieved early (less than 36 h). Pediatric radiologists use air or hydrostatic contrast enemas under ultrasonographic or radiographic guidance. The child can be lightly sedated and the surgeon is kept on stand-by in case of complication
- Strict criteria for successful reposition are a free flow of contrast into the small bowel, disappearance of diagnostic signs due to the mass, correlating with improvement of clinical status
- The success rate is over 70% in children with no associated disease
- Reduction is easier under general anesthesia (spontaneous reduction can be observed: 5%–10%)
- Perforation can occur if excessive pressure is applied or if the intestine is ischemic

Operation

- The operative steps are illustrated in Fig. 23.6
- Laparoscopic treatment by the transumbilical route should be attempted first
- The surgeon should squeeze the intussusception out between two probes rather than attempt to pull it out
- If laparoscopic treatment fails, right transverse supra- or infra-umbilical laparotomy must be undertaken
- After locating the leading point, reduction can be achieved by pushing the intussusceptum back into place. Reduction of the last part of the segment may require patience
- The intestine should be rinsed in warm saline solution and inspected for necrosis
- If complete reduction cannot be achieved, bowel resection and anastomosis is performed
- Routine fixation of the intestine and/or appendectomy is unnecessary

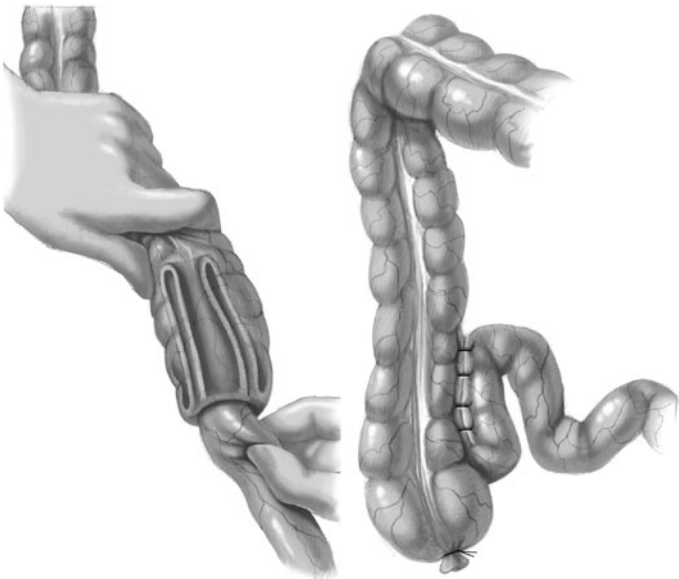


Fig. 23.6 Operative steps: intussusception

Postoperative care

- Antibiotic therapy is not mandatory
- Nasogastric tube placement is performed after bowel resection
- Parenteral feeding should be continued until normal peristalsis resumes
- Oral feeding can be resumed immediately after nonsurgical reduction

Prognosis

- Prognosis is good following early recognition and prompt treatment
- Intussusception recurs in up to 15% of cases after nonsurgical treatment. The family should be informed of this risk

23.5 Appendicitis

General considerations

- Appendicitis is a common cause of emergency abdominal surgery in children
- Variability in clinical findings often leads to misdiagnosis. Recent diagnostic and therapeutic techniques have reduced the number of false appendectomies, as well as the duration of recovery and hospitalization
- The negative laparotomy rate should be less than 10%

Classification

The classification of appendicitis is given in Table 23.1. Chronic appendicitis is a term used in cases of recurrent episodes of pain in the right, lower hemiabdomen. In the authors' opinion this term is wrong.

Table 23.1 Classification of appendicitis

Acute appendicitis	Acute inflammation of the appendix
Suppurative appendicitis	An early stage in which fluid fills the appendix and forms a local abscess
Gangrenous appendicitis	Advanced stage involving necrosis
Perforated appendicitis	Additional rupture allowing intestinal contents to enter the peritoneum

Signs

Interpretation of signs requires a great deal of clinical experience of examination. The hands should be warm and the examination should begin in an area where pain is least intense. The child's attitude should be observed carefully. If the child cries, wait until the inspiratory phase when the abdominal muscles become relaxed again.

- A wide range of signs can be observed
- In most cases pain is initially located periumbilically and subsequently shifts to the right lateral, lower quadrant. Proper assessment of pain is a key element for correct diagnosis. The following factors should be noted:
  - Pain location, type, intensity, irradiation
  - Localized guarding in the abdominal wall
  - Reaction when pressure is applied to McBurney's point
  - Reaction to contralateral decompression
  - Pain at rectal examination (the Douglas cul-de-sac)
- Factor of sedation and fatigue
- Lack of appetite, nausea with or without vomiting, and constipation
- Prodromal diarrhea may occur
- Fever is often present (38°C at the beginning)
- Tachycardia may occur
- Septic shock is a sign of peritonitis
- The examiner should always consider possible atypical clinical presentations in relation to the variability of the appendix position:
  - Retrocecal position with enhancement of posterior pain when extending the lower extremities and frequent association with diarrhea
  - Pelvic position with painful micturition
  - Mesocolic position with possible intestinal occlusion
- Another important point to remember is that perforation is followed by a calm period (closed cavity theory) in which pain subsides until signs of peritonitis appear
- The younger the child, the later the diagnosis is made (often after perforation)
- Repeated examination at close intervals is crucial to determine the evolution of signs

Differential diagnosis

- Chronic appendicitis is not a disease, the slow evolution of the signs leads to the correct diagnosis
- Yersinial ileitis (serological tests), gastroenteritis (stool examination), and regional diseases such as Crohn's disease should be considered

- Other intra-abdominal diseases including mesenteric adenitis, volvulus, intussusception, constipation, rheumatoid purpura, celiac disease, nephrolithiasis, and ovarian alterations in girls must also be ruled out (i.e. ultrasonographic examination)
- Many general surgeons consider negative appendectomy as a minor error and perform surgery liberally. Experienced pediatric surgeons must not share this point of view:
  It is always easy to go for an operation.
  The decision not to operate makes the good surgeon.

Preoperative work-up

- Complete blood count and fluid/electrolyte assessment whereas hyperleucocytosis is not necessarily a sign of appendicitis
- A urine strip test is useful for differential diagnosis of diseases associated with pain during micturition
- Abdominal ultrasonography is a good tool for overall evaluation. However, if the abnormal appendix can be visualized, appendicitis is likely. If the appendix cannot be located, appendicitis cannot be ruled out
- Intraperitoneal effusion is an indirect sign
- The clinical status remains the most important key to diagnosing appendicitis
- Prompt perfusion and rehydration may lower the risk of perforation
- A nasogastric tube is placed if the patient is vomiting
- Antibiotic therapy should be started prior to surgery if perforation is suspected

Operation: open procedure

- The operative steps are shown in Fig. 23.7
- Exposure for conventional appendectomy is obtained by a transverse incision at McBurney's point without muscle section. This incision may be enlarged obliquely above the iliac crest. Opening the sheath of the right grand abdominal muscle should be avoided (risk of infectious contamination)
- After dissection of the large muscles of the abdomen, the peritoneum is opened and the appendix is located

- After isolation of the structures, the appendix is dissected at the base
- The stump of the appendix can be inverted in case of false diagnosis or normal appendix (not systematically)
- Layer-by-layer closure of the abdominal wall is performed

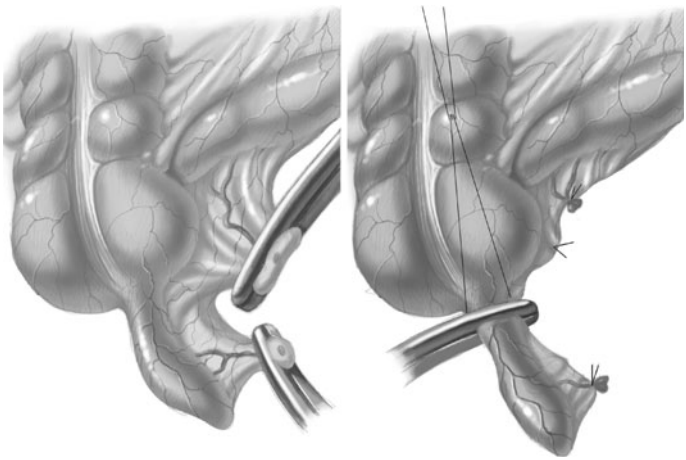


Fig. 23.7 Operative steps: appendicitis

Operation: laparoscopic appendectomy

- After production of pneumoperitoneum using the open technique (umbilical port), the first step consists of laparoscopic exploration of the abdomen
- If possible, the appendix should be mobilized, brought out through the umbilical port, and removed conventionally (one port technique)
- If mobilization is not possible, two more ports are placed in the right and left inguinal folds under visual control and appendectomy performed as follows:
  - Attachment of a clamp to the end of the appendix and preparation of the appendicular mesentery

  - Ligation of the base of the appendix using a suture or Roeder node. Alternatively resection can be performed using an automatic stapling device
  - Dissection of the appendix and extraction
- In cases of perforated appendicitis, the feasibility of laparoscopy depends on the surgeon's skills
- If exploration is normal, and a reason for the pain has been found (ovarian cysts, torsion of paraoophoric cysts, etc.), the appendix can be left in place

Operation: further considerations

- Regardless of the technique, the presence or otherwise of Meckel's diverticulum should always be verified. If a diverticulum is discovered, resection is advisable even in patients with acute appendicitis
- The need for drain placement and intraperitoneal lavage is controversial. Best results are obtained by placing a drain through a counter incision and doing the lavage with warm isotonic saline solution
- In patients with abscess and cecal problems (vitality) drainage-interval appendectomy can be performed 6–10 weeks after drainage only
- Prophylactic appendectomy may be indicated to avoid the possibility of future diagnostic errors in patients with isolated malrotation
- Conservative treatment with antibiotics alone can be proposed in exceptional situations such as operative contraindications, acute phase of oncological therapy, etc.

Postoperative care

- A nasogastric tube is unnecessary except after treatment of perforated appendicitis
- Antibiotic therapy should be included as part of the treatment of perforated appendicitis
- Feeding depends on resumption of intestinal transit
- In patients presenting with peritonitis, infusion should be continued at least for the first 72 h and until feeding is resumed

- Cicatricial and residual intraperitoneal abscesses can be drained by percutaneous puncture or by placement of a tube drain (with lavage), if close ultrasonographic or CT scan surveillance is feasible and the patient's clinical status is good

Complications

- Residual abscess
- Postoperative occlusion due to adherence
- Sterility due to tube dysfunction

Prognosis

- The mortality rate is low in pediatric centers
- Morbidity rate often continues to be too high

24 Anorectal Diseases

24.1 Bowel Washout

General considerations

- Colonic washout is indicated when complete emptying of the colon is required
- Washout can be achieved retrogradely by enemas or antegradely by total bowel washout
- Retrograde emptying is preferred, as whole bowel washout requires usually the use of a nasogastric tube, which most children find uncomfortable
- Bowel washout is recommended for the following procedures:
  - One-stage operations for Hirschsprung's disease
  - One-stage operations for anal anomalies (rectovestibular fistula)
  - Colon resections for Crohn's disease
  - Closure of colostomies
  - Proctocolectomy and ileoanal anastomosis for ulcerative colitis
  - Ileocolonoscopy

Retrograde washout

- Keep the patient on a liquid diet for 3–4 days prior to operation
- Empty the bowel with oral stimulant (bisacodyl) laxatives one day before the operation
- Give an enema (physiologic saline, phosphate enema, docusate enema) the evening before the procedure or on the morning of the procedure

Total bowel washout

Indicated in cases where complete cleanliness of the colon is required (diagnostic ileocolonoscopy) and in cases when reasonable emptying of the colon cannot be guaranteed. Polyethylene glycol (PEG) solution is the most commonly used washout solution.

- Washout is performed the day before the procedure
- Starting dose 25 $ml \cdot kg^{-1} \cdot h^{-1}$
- Administered orally or by a nasogastric tube
- Washout is stopped when stool is clear liquid
- Monitoring of the patient includes measurement of their weight; electrolytes should be controlled if the patient develops any general signs
- Washout is well tolerated in the great majority of patients
- If the patient develops signs during washout the rate of fluid administration should be decreased or the procedure discontinued

Contraindications

- Organic bowel stenosis
- Significant abnormalities in blood electrolytes
- Poor general condition
- In Hirschsprung's disease washouts should be performed using a large bore rectal tube

24.2 Chronic Constipation

General considerations

- Constipation is a problem in modern society
- False or unbalanced nutrition habits support constipation
- 5%–10% of all healthcare visits for children are because of constipation

Definition

Constipation is diagnosed when:

- The patient has a bowel movement twice or less in a week
- The patient has other signs consistent with chronic constipation
- Duration of the signs is longer than 6 weeks

Chronic constipation is considered severe when:

- There is recurrent fecaloma formation
- Patient suffers from overflow soiling
- The constipation is refractory to dietary measures and standard oral laxative therapy

Causes of chronic constipation

- Idiopathic (functional) constipation (>90%)
- Central nervous system impairment
- Anal anomalies
- Neuronal outflow obstruction (dysganglionosis)
- Chronic anal fissures
- Metabolic (hypothyreosis, hypercalcemia)
- Starving
- Psychogenic (anorexia nervosa, severe psychosocial problems)

Differential diagnosis

- The differential diagnoses of idiopathic constipation and Hirschsprung's disease are detailed in Table 24.1

Table 24.1 Differential diagnoses of idiopathic constipation and Hirschsprung's disease

	Hirschsprung's disease	Idiopathic constipation
Onset	Neonatal	Usually > 6 months
Soiling	No	Common
Enterocolitis	Common	No
Radiology ▪ Megacolon ▪ Transitional zone	 Almost always Almost always	 Common No
Manometry	Missing rectoanal reflex	Positive rectoanal reflex
Histology	Missing ganglion cells	Normal ganglion cells
Histochemistry (acetyl-cholinesterase staining)	Increased activity	Normal activity

Signs

- Signs of severe chronic constipation are listed in Table 24.2

Table 24.2 Signs of severe chronic constipation

Age at onset	
0–1 year	25%
1–5 years	70%
>5 years	15%
Boys:girls	2:1
Previous attempts to treat	90%
Soiling	70%
Unsuccessful toilet training	70%
Pain at defecation	70%
Abdominal pain	50%
Occasional blood in stool	25%
Poor appetite	30%
Wetting	20%
Primary psychopathology	20%
Rectal prolapse	3%

Aberrant forms of childhood chronic constipation

Aberrant forms of childhood chronic constipation are seen in:

- Slow transit constipation:
  - Adolescent females
  - No localized colonic dilatation
  - Slow whole colonic transit
  - Poor/moderate response to laxatives
- Paradox pelvic floor contraction:
  - Can be found in a subset of patients (10%–20%) with idiopathic constipation
  - Anal sphincters contract instead of relax when rectum is stimulated
  - Treatment similar as for idiopathic constipation
  - May respond to biofeedback

Investigations

The main focus in the diagnostic work-up is to make the distinction between organically induced constipation, which requires surgical therapy, and functional constipation which can be managed medically.

- Anal anomalies and local anal causes can be ruled out by careful clinical examination
- Systemic and central nervous system (CNS) causes can be ruled out by clinical history and examinations
- The common problem in small infants is differential diagnosis between Hirschsprung's disease and functional constipation
- Clinical history
  - Onset of signs
  - Duration of signs
  - Frequency of bowel movements
  - Soiling
  - Dietary habits
  - Modes of previous treatment
  - Other signs (wetting, etc.)
  - Psychosocial factors
- Clinical examination
  - General impression
  - Nutritional status
  - Emotional status
  - Abdominal palpation
  - Anal inspection
  - Rectal examination
- Contrast enema
  - Only in unclear cases and in those with poor response to optimal medical therapy
  - If contrast enema is consistent with functional constipation, no further examinations are needed
  - If contrast enema is inconclusive or refinements in medical therapy are without success, further examinations are indicated

- Transit time studies
- Anorectal manometry
- Rectal biopsies

Management of severe chronic constipation

Conservative and medical treatment can be initiated based on history and clinical examination, provided there are no features suggesting a specific organic cause. The duration of this treatment is at least as long as the duration of signs.

Primary counseling should consist of:

- Adjustment of diet
  - Increased water in diet
  - Increased fiber in diet
- Emptying the bowel (if fecaloma formation and overflow soiling)
  - Large enemas for 1–3 days
  - Confirm evacuation
  - Avoid long-term enema regimens
- Establish toileting routines
  - Stimulant laxatives (bisacodyl, senna, picosulfate)
  - Start with high doses, taper slowly according to response
  - Regular daily toileting, after a major meal (gastro-colic reflex)
- Maintenance of bowel movements
  - Bulk laxatives
  - Start when the dose of stimulant laxatives is approximately one-third of the starting dose
  - Again, start with high doses

Operative treatment is usually not indicated. There is no evidence to support anal dilatations or sphincter myotomies in the management of chronic constipation. However, sphincter myotomy may be indicated for chronic anal fissures with high anal resting tone.

24.3 Anorectal Manometry

General considerations

- Anorectal manometry is applied for studying anorectal physiology
- Common variables measured at anorectal manometry include:
  - Anal resting tone
  - Anal sphincter squeeze
  - Presence of rectoanal relaxation reflex
  - Rectal sensibility
- The study is usually performed with a water-perfused or steady-state electronic system with one or several measurement channels
- There are no universal normal values in children as the manometric equipment varies in performance and accuracy
- It is important that each unit develops institutional normal values

Indications

- Diagnosis of Hirschsprung's disease
- Diagnosis of internal sphincter achalasia
- Evaluation of anorectal physiology following anorectal surgery
- Evaluation of anorectal physiology in patients with anal incontinence
- Differential diagnosis of idiopathic constipation (in selected cases)

Technique of anorectal manometry

- Bowel washout the day before examination
- Infants and small children (<5 years) require sedation
  - Ketamine is preferred as it does not interfere with sphincter tone
  - Benzodiazepines and inhaled anesthetics lower the sphincter tone
- The anorectal sphincter profile can be assessed with constant pull-through or segmental stationary techniques
- The rectum is stimulated by a latex balloon
- Reliable assessment of voluntary sphincter squeeze cannot be achieved in children younger than 5–6 years

Typical findings in anorectal manometry

- See Table 24.3 for typical findings in anorectal manometry

Table 24.3 Typical findings in anorectal manometry

	Resting pressure	Voluntary squeeze	Rectoanal relaxation reflex	Rectal sensibility
Hirschsprung's disease	↑ Normal	(Normal)	Missing	(Normal)
Internal sphincter achalasia	↑ Normal	Normal	Missing	↓
Idiopathic constipation	↓ Normal	Normal	Normal ↑ Threshold	↓
Incontinence (following anal anomaly repair)	↓	↓	Missing or low amplitude	Normal ↓
Neurogenic anal incontinence	↓ Normal	No or rudimentary squeeze	Normal	Missing ↓

24.4 Rectal/Colonic Biopsy

General considerations

- Rectal biopsy is obtained for the diagnosis of Hirschsprung's disease or other dysganglionosis
- Open biopsies can be taken as partial or full-thickness samples or using a rectal suction device
- Usually one biopsy is sufficient, provided it is representative and has been taken from the proper level of the rectum:
  - 1 cm above the dentate line, preferably from the dorsal rectum wall
  - Must contain enough submucosa and muscularis mucosae
- Routine staining includes standard staining and histo- or immunohistochemical studies to demonstrate neuronal elements of the bowel wall

Noblett biopsy forceps

- This forceps has two openings 2 cm apart (Fig. 24.1). After suction of the mucosa and submucosa a cylindrical scalpel moves and cuts the biopsy samples

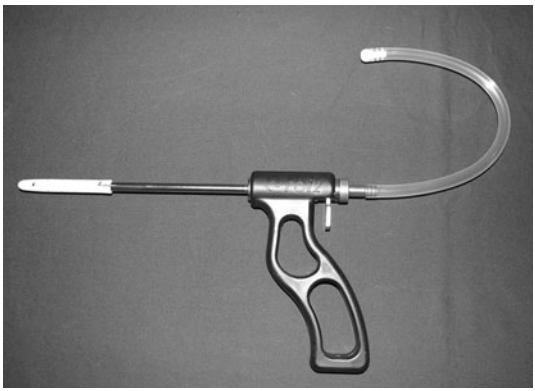


Fig. 24.1 Noblett biopsy forceps

Operation: open or laparoscopic biopsies

- Open biopsy samples can be taken or laparoscopically as partial or full-thickness samples
- In the abdomen the apparent transitional zone is selected
- Seromuscular biopsy samples are taken from the antimesenteric surface if possible without entering the lumen (Fig. 24.2)

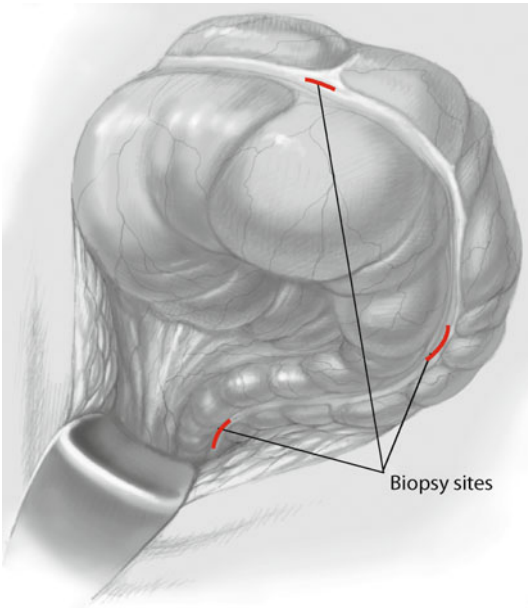


Fig. 24.2 Biopsy sites in Hirschsprung's disease

24.5 Hirschsprung's Disease

General considerations

The craniocaudal migration of ganglion cells of the bowel begins at the 12th week of gestation. Hirschsprung's disease (aganglionosis of the distal bowel) is caused by a deficiency of this migratory process. The cessation of ganglion cell migration is caused either by a defect in the cells or an abnormal microenvironment that blocks further migration of the cells. The earlier the migration stops, the longer is the aganglionic part of the distal bowel. The aganglionic bowel in Hirschsprung's disease is unable to relax and the propulsive wave stops in the proximal end of the aganglionic segment. This dynamic obstruction causes dilatation of the bowel proximal to the aganglionic segment.

- The reported incidence of Hirschsprung's disease range between 1:3000 and 1:5000
- Male:female ratio is 4:1
- Familial incidence of Hirschsprung's disease ranges between 7% and 10%
- Several gene mutations are associated with Hirschsprung's disease, the most frequent are mutations in the RET proto-oncogene

Classification

- Hirschsprung's disease classification is given in Table 24.4

Table 24.4 Classification of Hirschsprung's disease

Rectosigmoid (classical)	75%–80%
Long colonic	10%
Total colonic	10%
Jejunoileal	< 5%

Signs

- Almost all patients with Hirschsprung's disease have signs as neonates
- The acute signs may subside and the child subsequently has bouts of intestinal obstruction, especially when he or she starts having solid foods
- Delayed meconium stool (>24–48 h)
- Constipation
- Vomiting
- Poor feeding
- Abdominal distension
- Clinical and radiological signs of low bowel obstruction
- Enterocolitis is the most feared complication of Hirschsprung's disease
- About 15%–20% of patients develop enterocolitis either before or after the definitive surgical treatment; more commonly before the operation with the following signs:

- Abdominal distension
- Foul-smelling loose stools
- Fever
- Rapidly developing signs of sepsis

Preoperative work-up

- History suggesting Hirschsprung's disease
- Contrast enema
  - Megacolon (may be insignificant in a neonate)
  - Transitional zone showing the difference in caliber
- Anorectal manometry
  - Missing rectoanal reflex
- Rectal biopsy
  - No ganglion cells
  - Thick nerve trunks and increased activity in lamina propria and mucosa in acetylcholinesterase histochemistry

Conservative therapy before definitive surgery

- Continue with oral feeds
- There is no urgency between diagnostic work-up and the timing of definitive surgery for the responders
- Daily emptying of the bowel with saline enemas (30–50 ml in a neonate)
- Most patients tolerate weeks or months of conservative treatment if bowel washouts are performed regularly
- The patients who do not comply with this regime are those with a long segment disease and many of those have Down's syndrome

Operation strategy

The aim of the surgical treatment of Hirschsprung's disease is to resect or by-pass the aganglionic bowel that causes the functional obstruction. Today many patients, even in the neonatal period, have primary surgery without preliminary bowel diversion

- However, bowel diversion is required in some patients:
  - Those with fulminant enterocolitis
  - Those who are not manageable with routine bowel washouts
  - Those with long segment or total colonic aganglionosis who are not planned to have a neonatal repair

Operation: colostomy

- The aim is to establish a leveling colostomy to a bowel segment above the transitional zone with normal innervation and caliber
- The colostomy operation should be scheduled so that a specialized pathologist is available to evaluate the frozen sections removed
- This type of stoma can be subsequently pulled-through without the need to confirm ganglion cell status
- The optimal site of the stoma is the left abdomen at or below the level of the umbilicus
- The technique of stoma formation can vary provided the following basic rules are followed:
  - Both stoma ends should be fixed to all layers of the abdominal wall
  - Both stoma barrels should be separated by a fascia or skin bridge (Fig. 24.3)
  - Brooke-type stoma maturation makes stoma care easier

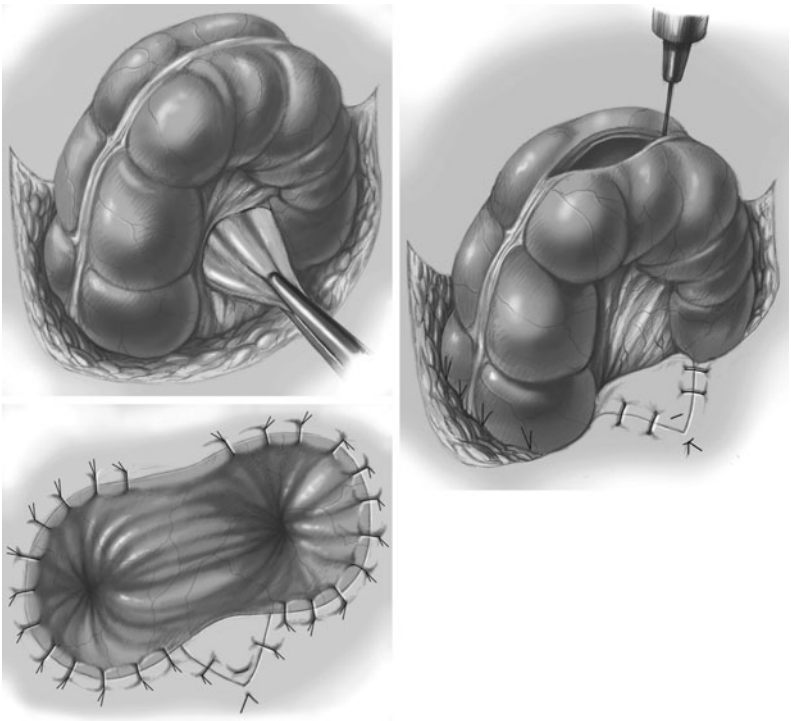


Fig. 24.3 Operative steps: skin bridge colostomy

Operation: preparation

- Preoperative bowel washout
- Antibiotics initiated at anesthesia induction
- Lithotomy position
- Bladder catheter

Operation: Duhamel retrorectal pull-through

- The aganglionic bowel proximal to the rectum is mobilized (Fig. 24.4)
- Intrapelvic dissection is continued on the back wall of the rectum until the level just above the dentate line is reached

- The rectal stump is closed with sutures or a stapler
- A hemicircumferential incision is made in the back wall of the anal canal 0.5–1.0 cm above the dentate line
- The ganglionic bowel is pulled-through and anastomosed to the opening in the posterior wall of the anal canal
- A side-to-side anastomosis is created between the rectum and the pulled-through bowel by a gastrointestinal anastomosis (GIA) stapler. Some surgeons prefer to perform another stapler firing abdominally through the opened rectal stump and enterotomy in the anastomosis through the abdomen, connecting the opened rectal stump with the pulled-through colon. This decreases the risk of septum formation between the rectum and the pulled-through colon, but requires an abdominal side-to-side anastomosis that increases the risk of postoperative infective complications
- The abdominal phase of the Duhamel operation is now commonly performed laparoscopically

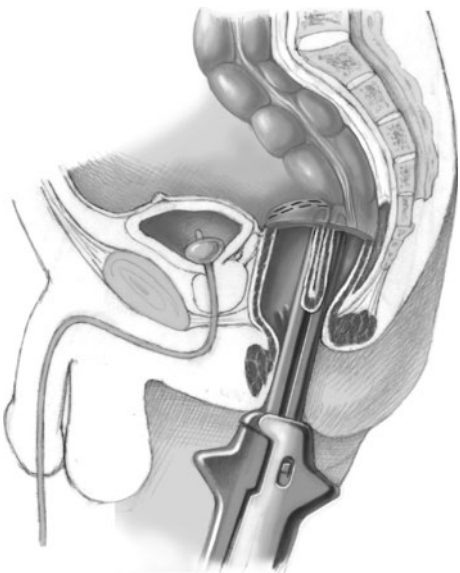


Fig. 24.4 Duhamel operation

Operation: Swenson rectosigmoidectomy

- Low mobilization of the rectum anteriorly down to 2 cm above the dentate line and posteriorly 1 cm above the dentate line (Fig. 24.5)
- The rectum is everted through the anus and the anterior wall is hemicircumferentially transected
- Ganglionic bowel is pulled through the opening in the anterior rectal wall
- An oblique anastomosis between ganglionic colon and distal rectum is performed outside the anal canal. In the anterior wall the anastomotic line is higher from the anal verge
- The Swenson operation can also be used for a totally transanal pull-through for Hirschsprung's disease

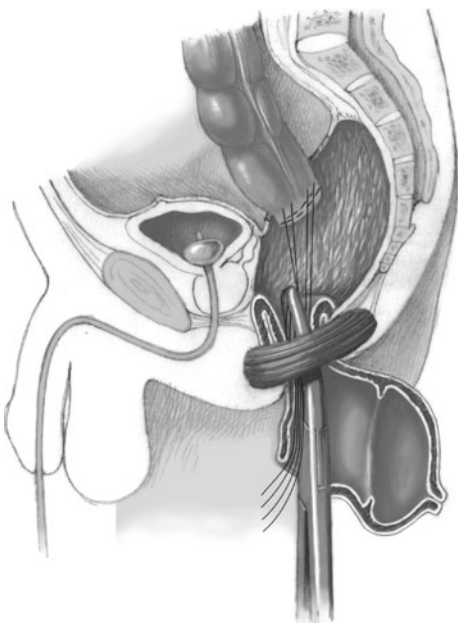


Fig. 24.5 Swenson's pull-through operation

Operation: Soave transabdominal endorectal pull-through

- The operation is illustrated in Fig. 24.6
- The aganglionic bowel is mobilized to the level of the peritoneal reflection
- The seromuscular layer of the bowel wall is incised circumferentially
- An abdominal mucosectomy is performed and extended distally to the level just above the dentate line
- The mucosal tube can be transected at level of the distal end of the dissection or everted through the anus
- The ganglionic bowel is pulled through and anastomosed to the anal canal at the level of the transection of the mucosal tube
- This operation has also been modified with laparoscopic dissection of the colon and a transanal mucosectomy

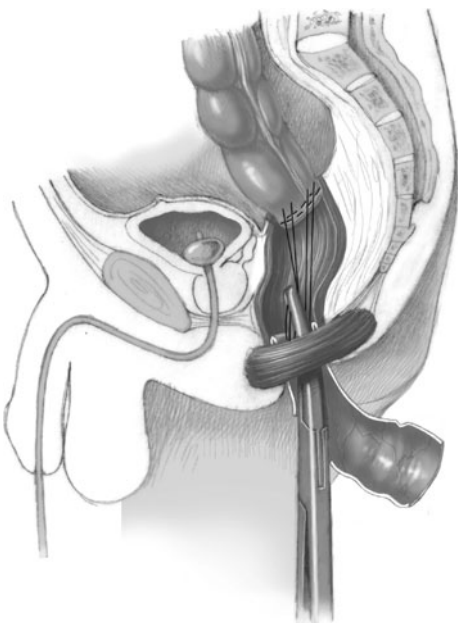


Fig. 24.6 Soave endorectal pull-through

Operation: transanal endorectal pull-through (De La Torre)

- The bowel mobilization is performed completely through a transanal approach:
  - The operative steps are illustrated in Fig. 24.7
  - A circular incision in the mucosa about 1 cm above the dentate line is made
  - The mucosa is dissected for about 8–10 cm leaving the muscular cuff intact until the peritoneal reflection is reached. However, nowadays some surgeons prefer a shorter cuff (2–4 cm)
  - At this point the muscular cuff is opened and the surgeon has access to the abdomen
  - The aganglionic bowel is pulled successively out through the anus and the rectal and sigmoid vessels are divided
  - The muscle cuff is split on the posterior side, thus decreasing the risk of enterocolitis and improving functional results
  - The healthy colon is then sutured circularly to the resected site at the dentate line
  - Patients with classic Hirschsprung's disease may be operated on by the totally transanal approach
  - In older children, up to 55 cm of bowel can be resected transanally

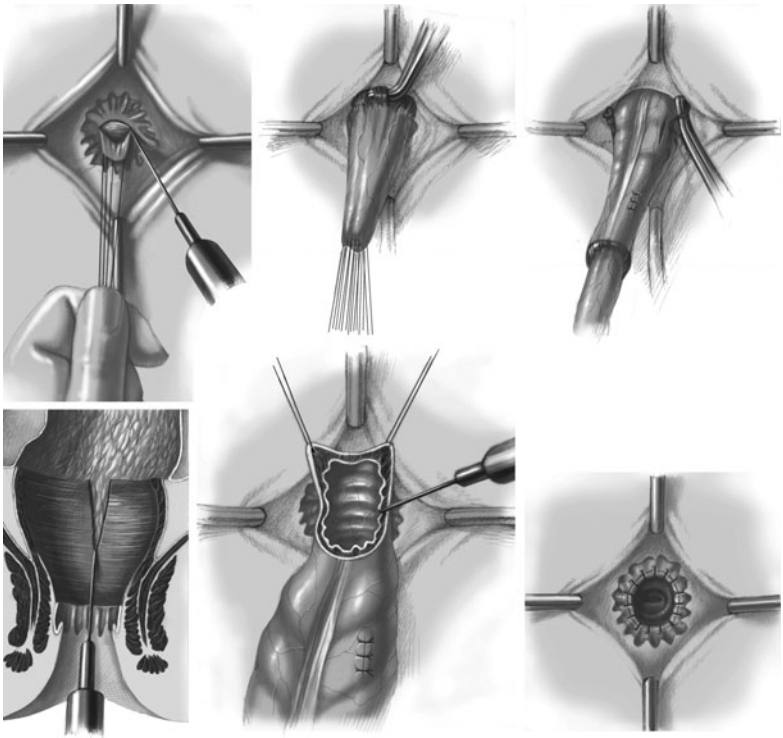


Fig. 24.7 Transanal pull-through

Operation: anterior resection of Rehbein

- The rectum is dissected circumferentially to a point about 3–4 cm above the dentate line (Fig. 24.8)
- The anastomosis between the rectum and ganglionic proximal bowel can be performed abdominally by hand-sewn stitches or by a circular stapler
- If a stapler is used, both the rectal stump and the distal end of the ganglionic bowel are closed with a purse string suture over the anvils of the end-to-end anastomosis (EEA) stapler

- The stapler is tightened and fired
- A circular unbroken doughnut of bowel wall between the jaws of the stapler confirms a successful stapler anastomosis

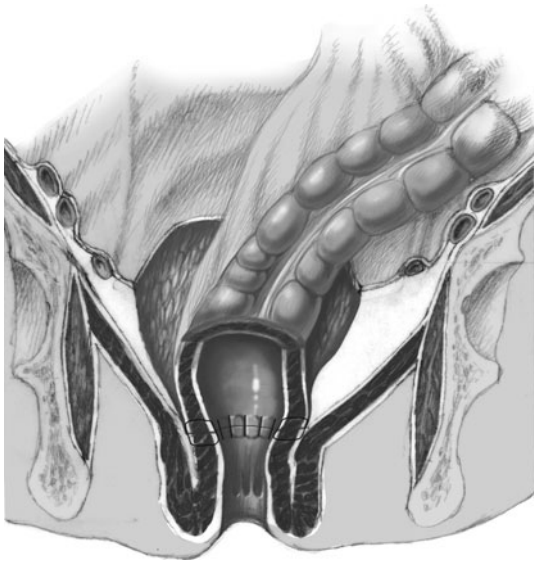


Fig. 24.8 Rehbein's operation

Operation: sphincteromyotomy

- Indicated in patients with internal sphincter achalasia (see Sect. 24.6) or who develop sphincter muscle achalasia after an operation
- Digital dilatation of the anus after the patient has had a general anesthetic
- Semicircular incision in the skin about 1 cm below the dentate line
- Submucosal dissection for at least 2 cm above the incision
- Resection of a muscle strip about 0.5 cm wide and about 3 cm long
- The incision is closed and an anal tampon inserted for 12 h

Postoperative treatment

- A nasogastric tube is not used routinely
- Bladder catheter is kept in place as long as the patient has an epidural catheter or an opiate infusion (2–3 days)
- Enteral feeding is allowed when the first bowel motions are noted (1–2 days after operation)
- Routine total parenteral nutrition is not applied
- Antibiotic therapy is continued for 3 days
- The anus is calibrated 14 days after the operation
- The anal dilatation regime is only used if there is a clear anastomotic narrowing
- Patients who have undergone totally transanal pull-through require less postoperative analgesia, and resume bowel function and enteral feeding significantly faster than those with associated laparotomy or laparoscopy

Prognosis

- Beyond childhood, 80%–90% of patients with repaired Hirschsprung's disease resume normal or nearly normal bowel function and control
- During childhood a significant proportion of patients suffer from diminished fecal control (10%–40%) and to a lesser extent from recurrent constipation (10%–30%)
- Postoperative enterocolitis occurs in 10%–30% of patients
- There are no significant differences in terms of functional outcome between the major surgical techniques. The following findings however are reported as general trends in the literature but have not been confirmed by controlled studies:
  - Recurrent constipation appears to be more common following the Rehbein operation
  - Soiling is more common following the Swenson procedure
  - Enterocolitis is more frequent after an endorectal pull-through

24.6 Allied Hirschsprung's Disease

- **Internal sphincter achalasia**
  - General considerations
    - Internal sphincter achalasia is characterized by the lack of relaxation of the internal sphincter
    - The condition is much less common than Hirschsprung's disease (1:20,000–30,000)
  - Signs are similar to idiopathic constipation but response to optimal medical management is poor or completely lacking
  - Diagnosis
    - Anorectal manometry, showing lack of internal sphincter relaxation reflex
  - Treatment of internal sphincter achalasia is internal sphincter myotomy
  - Prognosis
    - Between 65% and 80% of patients respond favorably to myectomy, the rest still need laxatives to maintain bowel function

- **Hypoganglionosis**
  - General considerations
    - Hypoganglionosis is characterized by a small number of ganglion cells in the neuronal plexuses of the bowel wall
    - The ganglion cells are also smaller than normal
    - Hypoganglionosis is a typical feature of the transitional zone between ganglionic and aganglionic bowel in Hirschsprung's disease
    - Hypoganglionosis occurs also independently but is an uncommon disorder
  - Signs are similar to idiopathic constipation
  - Diagnosis
    - Full-thickness rectal biopsy showing the typical hypoganglionosis
    - Anorectal manometry findings are inconsistent

- Treatment
  - Controversial results
  - Most patients respond acceptably to conservative treatment
  - Patients with recalcitrant signs and lacking internal sphincter relaxation may respond to internal sphincter myectomy
  - The role of bowel resection for hypoganglionosis is unclear

- **Intestinal neuronal dysplasia**
  - General considerations
    - Intestinal neuronal dysplasia is characterized by hyperplastic ganglions with an increased number of ganglia cells per ganglion
    - Another type of intestinal neuronal dysplasia with hypoplastic sympathetic innervation is very rare
    - Incidence of true intestinal neuronal dysplasia is difficult to estimate as similar histological findings are common in the developing bowel during infancy
    - Similar histological changes may also occur in obstructed bowel and in inflammatory bowel disease
    - Intestinal neuronal dysplasia is a clinical entity but probably more uncommon than anticipated
  - Signs
    - Similar to idiopathic constipation in small children
    - Early-onset signs in infants with intestinal neuronal dysplasia mimic Hirschsprung's disease
  - Diagnosis
    - Rectal biopsy samples show characteristic findings
    - Anorectal manometry findings are inconsistent
  - Management of intestinal neuronal dysplasia is controversial
    - Conservative therapy is preferred in infants as histological changes of intestinal neuronal dysplasia tend to diminish and eventually disappear
    - Medical management is similar to that for chronic constipation

- Neonatal patients with intestinal neuronal dysplasia and severe obstruction may benefit from temporary fecal diversion (3–6 months)
- Some surgeons advocate bowel resections as in Hirschsprung's disease for intestinal neuronal dysplasia but as yet there are no controlled studies concerning the efficacy of surgical treatment

24.7 Anorectal Malformations

General considerations

- Anorectal malformations develop early during the first 2 months of gestation, however the cause is unclear
- The agents causing anorectal malformations have general noxious effects on the developing fetus; therefore, associated anomalies are common
- Embryologically, the fistula between the rectum and the urogenital tract or perineum is an ectopic anus
- The voluntary external sphincters develop in their normal position
- In the western world the incidence is 1:3300–5000
- 60% of patients are male
- Male patients tend to have more severe malformations than females ones

Classification

- Two classification are most commonly used:
  - Wingspread classification (Table 24.5) stratifies anomalies as high, intermediate or low
  - Pena classification (Table 24.6) is based on the location of the fistula between the bowel termination and bowel outlet (this is the classification used in this chapter)

Table 24.5 Wingspread international classification

	High	Intermediate	Low
Female	Anorectal agenesis	Rectovaginal fistula	Anovestibular fistula
	Rectovaginal fistula	Rectovestibular fistula	Anocutaneous fistula
	No fistula	Anal agenesis	Anal stenosis
	Rectal atresia		
		Cloaca	
		Rare malformations	
Male	Anorectal agenesis	Bulbar fistula	Anocutaneous fistula
	Rectoprostatic fistula	Anal agenesis	Anal stenosis
	No fistula		
	Rectal atresia		
		Rare malformations	

Table 24.6 Pena classification

Female	Perineal fistula	
	Vestibular fistula	
	Persistent cloaca	<3 cm common channel
		>3 cm common channel
	Imperforate anus without fistula	
	Rectal atresia	
Male	Perineal fistula	
	Rectourethral fistula	Bulbar
		Prostatic
	Rectovesical fistula	
	Imperforate anus without fistula	
	Rectal atresia	

Typical types of anorectal malformations in females

- These are illustrated in Fig. 24.9
- **Perineal fistula**
  - Perineal fistula presents as a fistulous opening between the anal site and vestibulum without a median bar
  - Fistula opening more anterior to the sphincter complex than in males
  - Fistula opening is not within the sphincter complex
  - Perineal groove between bowel outlet and vestibulum is common
- **Anterior perineal anus**
  - Normal or moderately stenotic anus (50% of cases) situated anteriorly right behind the vestibulum
  - Normal sphincter support posteriorly and laterally, poor sphincters anteriorly
- **Rectovestibular fistula**
  - The fistula is situated in the posterior fourchette of the vestibulum, behind the hymeneal ring and is as common as a perineal fistula
  - Voluntary sphincters are usually well developed
  - The rectum and posterior vaginal join a common wall for 2–4 cm
  - Vaginal anomalies are common
- **Rectovaginal fistula**
  - A rectovaginal fistula is exceedingly rare
- **Cloaca**
  - Urinary tract, vagina, and rectum join in a common channel; the rectum usually drains to the inferior posterior wall of the vagina but can drain anywhere in the system
  - The length of the common channel is usually between 1 cm and 5 cm but may be longer
  - Patients with a relatively long common channel tend to have poor sphincters
  - The orientation and anatomy of the cloaca are extremely variable
  - Obstructive uropathy is not uncommon; most patients have some sort of urinary tract abnormalities
  - Vaginal and uterine duplications occur in 50% of cases, often associated with neonatal hydrocolpos
  - Agenesis of Mullerian structures is not uncommon

- Most patients have sacral anomalies that are more severe in patients with a long common channel

- **Anorectal malformation without a fistula**
  - Uncommon in females
  - Most patients have Down's syndrome
  - Normally there is a short distance between the blind ending rectum and a usually well-formed anal pit
  - Well-developed sphincters
- **Rectal atresia**
  - As in males (see below)

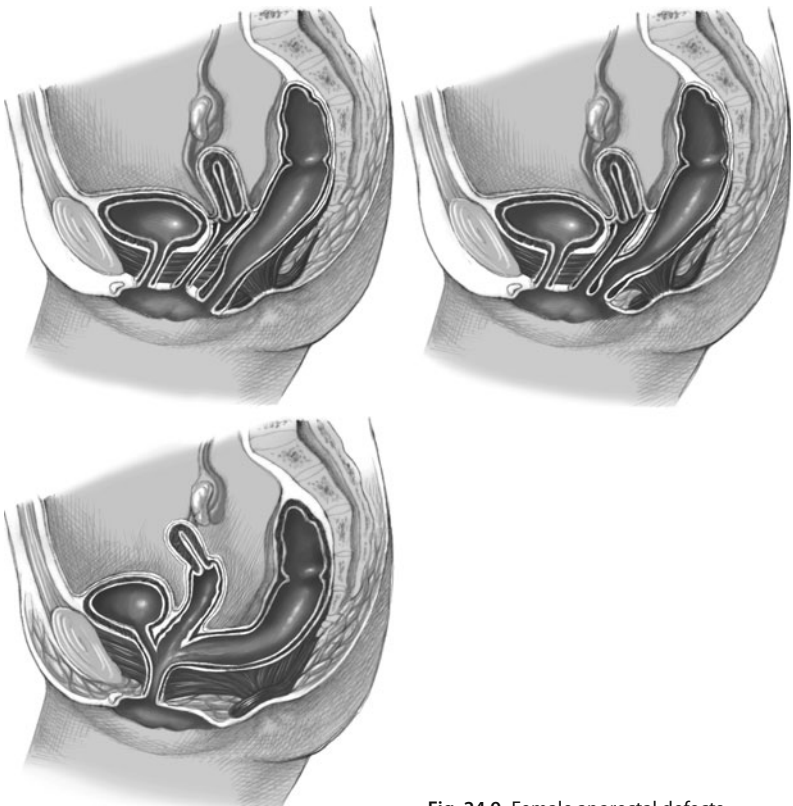


Fig. 24.9 Female anorectal defects

Typical types of anorectal malformations in males

Anorectal malformations in males are illustrated in Fig. 24.10. In all following anorectal malformation types, except in rectal atresia, the bowel ends either in a fistula or is blind and has all the features of a normal anus, i.e., terminal transitional epithelium, anal glands and internal sphincter. If there is no perineal fistula the patient is likely to have a urethral fistula

- **Perineal fistula**
  - Usually associated with a median bar
  - Opens usually along or at the tip of the median bar
  - May open also on either side of the median bar (covered anus)
  - Distal rectum is partially surrounded by the voluntary sphincter complex
- **Rectobulbar fistula**
  - The fistula opens into the urethra at the bulbar urethra
  - Usually there is a relatively long common wall between the urethra and rectum
  - Voluntary sphincters are usually well developed
  - Usually there is a visible pit at the anal site
- **Rectoprostatic fistula**
  - The fistula opens into the urethra at the level of the prostate
  - The rectum and prostatic urethra join a common wall
  - Voluntary sphincters are more hypoplastic than in bulbar fistula
  - Sacral deformity is more severe than in bulbar fistula
  - A pit may or may not be visible at the anal site
  - Some patients have a "flat bottom" suggesting severe sphincter hypoplasia
- **Recto-bladder neck fistula**
  - Relatively uncommon
  - The fistula opens into the bladder neck
  - Usually there is no significant common wall between the rectum and bladder
  - Severe sacral deformities
  - Usually the patient has a flat bottom with very poor voluntary sphincters

- **Anorectal anomaly with no fistula**
  - Rectum ends blindly behind the urethra
  - The blind end usually extends to near a well-formed anal pit
  - Well-developed voluntary sphincters
  - A typical anorectal anomaly in patients with Down's syndrome
- **Rectal atresia**
  - Etiology likely to be a local vascular abnormality
  - Normal-looking anus ending blindly above the dentate line
  - Normal voluntary sphincter muscles
  - Proximal blind-ending bowel is usually connected to the blind-ending anus by a short distance of fibrous tissue
  - Both structures may be separated only by a membrane

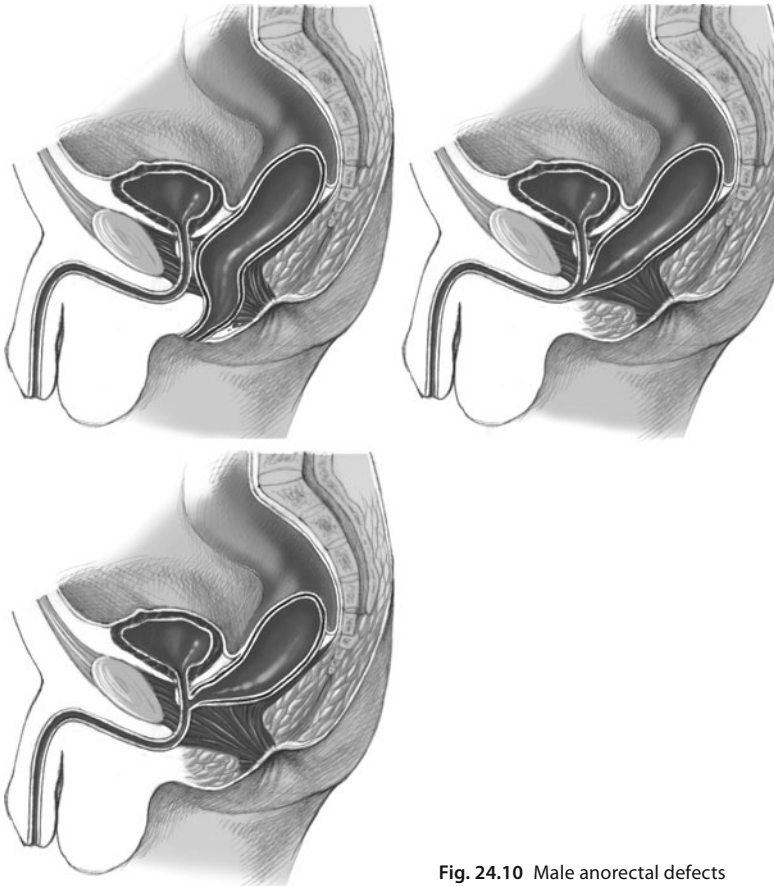


Fig. 24.10 Male anorectal defects

Preoperative work-up

- Clinical examination discloses the definitive type of anomaly in the majority of cases
- Cross-table lateral plain X-ray may be used in doubtful cases. However, they should not be performed before 18–24 h after birth
- Early perineal ultrasonographic examination
- CT or MRI may have advantages especially when trying to answer the question of sphincter muscle integrity

- Ultrasonographic examination is indicated for the detection of associated anomalies:
  - Urinary tract malformations
  - Cardiac malformations
  - Spinal cord anomalies

Associated malformations

- The overall incidence of associated anomalies is 65% (Table 24.7).

Table 24.7 Incidence of anomalies associated with anorectal malformations in males

Anomaly	Incidence (%)
Urogenital anomalies	45
Skeletal anomalies (excl. coccyx)	30
Gastrointestinal anomalies	20
Cardiovascular anomalies	15
Chromosomal anomalies	10
CNS anomalies	5
Others	15

Operative strategy

The surgical procedure for the management of anorectal malformations depends on the type of the anomaly, which has to be ascertained during the first 24 h of life. Life-threatening associated anomalies that require urgent management (i.e., severe cardiac anomalies, obstructive uropathy, esophageal atresia, etc.) have to be ruled out.

- **Colostomy needed**
  - In males with:
    - Rectourethral fistula
    - Rectovesical fistula
    - No fistula (if distance between rectum and skin is unclear or >1 cm)
    - Rectal atresia

- In females with
  - Cloaca
  - Vestibular fistula (may be repaired when a neonate without a stoma)
  - No fistula (if distance between rectum and skin is unclear or > 1 cm)
  - Rectal atresia

- **Colostomy is performed**
  - As a split colostomy in the proximal sigmoid or distal descending colon
  - Risk for stoma prolapse is less than in more proximal stomas
  - Stool quality is better than in more proximal stomas
  - Washout of the distal blind-ending bowel is easier
  - Cloaca patients with missing or hypoplastic Mullerian structures need a transverse colostomy, as the use of the sigmoid bowel is the preferred method for vaginal reconstruction later
- **Definitive operation after neonatal colostomy**
  - 3–8 weeks later
  - Definition of the exact anatomy of the malformation by distal loopogram
  - Intermittent catheterization or urinary diversion (vesicostomy) in patients with obstructive uropathy (especially cloaca)
  - Drainage of hydrocolpos (preferably external) in cloaca patients
- **Neonatal repair possible**
  - In males with:
    - Perineal fistula
    - No fistula (if distance between rectum and skin is clearly < 1cm)
  - In females with:
    - Perineal fistula
    - No fistula (if distance between the rectum and skin is clearly < 1cm)
    - Rectovestibular fistula – in experienced hands!
- Suggested techniques for repair are listed in Table 24.8

Table 24.8 Suggested techniques for repair. (*PSARP* posterior sagittal anorectoplasty, *PSARVUP* posterior sagittal anorectovaginourethroplasty, *TUM* total urogenital mobilization)

Males	
Perineal fistula	Minimal PSARP or cutback
Rectourethral fistula	PSARP
Rectovesical fistula	PSARP ± laparotomy
No fistula, high	PSARP
No fistula, low	Minimal PSARP
Rectal atresia	PSARP
Females	
Perineal fistula	Limited PSARP
Vestibular fistula	Limited PSARP
Cloaca	PSARVUP ± laparotomy ± TUM
No fistula, high	PSARP
No fistula, low	Minimal PSARP
Rectal atresia	PSARP

Operation: PSARP in males with rectourethral or recto-vesical fistula (Fig. 24.11)

- Prone position with raised pelvis
- Bladder catheter, confirm proper placement, may go through the fistula to the rectum
- Voluntary sphincters identified by a muscle stimulator
- Midline incision from the sacrum to the identified and marked anal site
- The incision is extended in the midline through parasagittal fibers of the external sphincter and levator ani
  - In standard PSARP the whole external sphincter complex is split
  - The distal part of the sphincter complex may be left intact as there is a loose tissue plane in the center of the external sphincter that can be easily identified and dilated to accommodate the rectum
  - Endopelvic (Waldeyer's) fascia encompassing the rectum is incised in the midline

- In patients with a vesical fistula the rectum may not be mobilized through this approach:
  - The patient is turned supine and a low transverse laparotomy performed
  - Abdominal closure of the fistula and mobilization of the rectum
  - Distal rectum is pulled through to the sacroperineal incision and the patient turned prone again
- Back wall of the bowel termination is opened and the fistula site identified
- The rectum is separated from the urethra and prostate:
  - Urethra, prostate, and rectum join a common wall for a variable distance
  - Care should be taken to avoid injury to the urethra, prostate, and seminal vesicles
  - As much as possible of the bowel termination at the fistula site should be preserved as it contains the internal anal sphincter
  - The urethral end of the fistula is closed
- The rectum is mobilized to reach the anal site with mild tension
- If the blind-ending rectum is extremely dilated, it can be tapered by resecting the posterior wall of the dilated pouch
- The anterior wall of the sphincter funnel is reconstructed and the rectum is pulled through
- The posterior wall of the sphincter funnel and parasagittal fibers are reconstructed
- Anoplasty is performed between perineal skin and bowel outlet
- Alternatively, if the sphincter complex is not split completely, the rectum is pulled through the dilated channel in the center of the sphincter funnel
- The posterior wall of the levator and external sphincters are reconstructed
- Anoplasty is performed as above

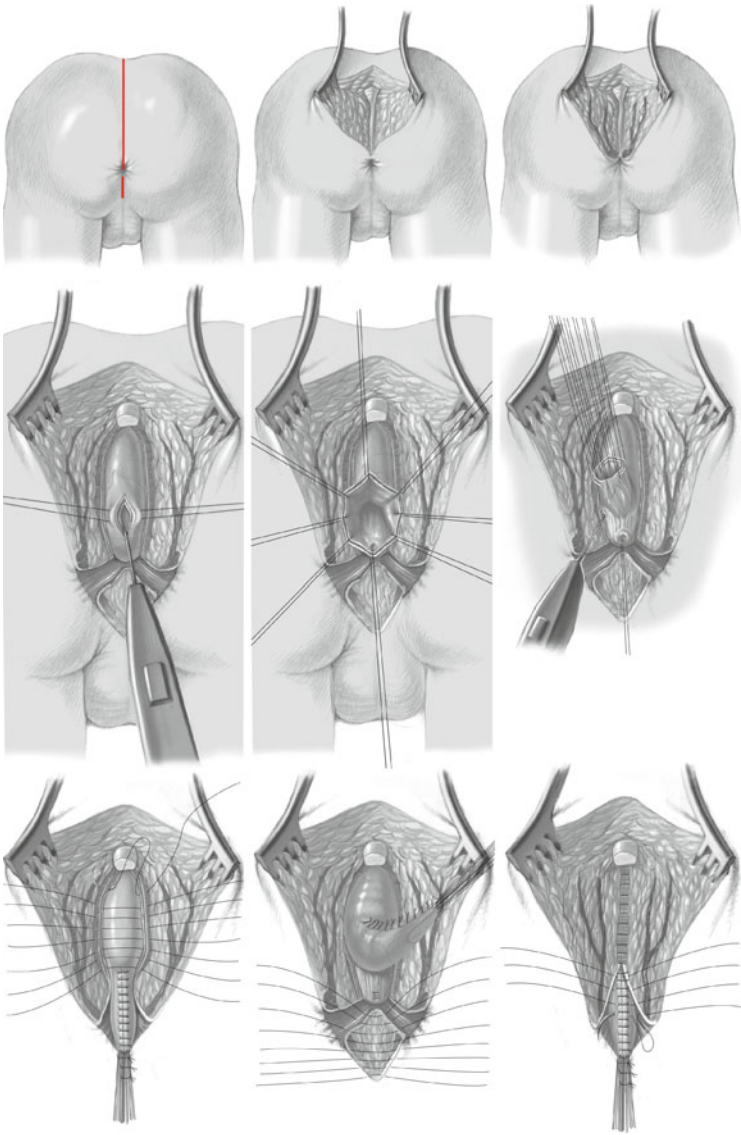


Fig. 24.11 Operative steps: repair of male anorectal defects

Operation: minimal PSARP in perineal fistula in males

- Anal site is identified by muscle stimulation
- Incision from the anal site to and around the fistula
- Fistula and terminal rectum are mobilized
- Anterior part of the sphincter funnel is split in the midline
- The terminal bowel is placed in the center of the sphincters
- Closure of the anterior part of the sphincter funnel and anoplasty

Operation: cut-back in perineal fistula in males

- The fistula is incised over a small probe until the posterior wall of the anal canal is reached
- The mucosal edge of the bowel termination is sutured to anal skin

Operation: limited PSARP in females with vestibular or perineal fistula (Fig. 24.12)

- Supine position
- Anal site is identified by muscle stimulation
- Skin incision from the anal site to and around the fistula
- The fistula is mobilized from the surrounding perineal or vulvar skin/mucosa
- The fistula is separated from the vagina
  - There is a common wall between the vagina and rectum for a variable distance
  - The dissection is extended above the level of the common wall
  - Injury to the rectum should be avoided, small tears in the vaginal wall are easily repaired and heal rapidly
  - If the rectum is injured a colostomy should be performed if not performed earlier
- The rectum is separated from lateral muscular attachments
- Midline dissection is extended through the anterior wall of the sphincter funnel to the anal site
- The mobilized rectum is pulled in the midst of the sphincters
- The perineal body is repaired in at least two layers of sutures
- A formal anoplasty is performed
- The vestibular fourchette and posterior hymen are reconstructed

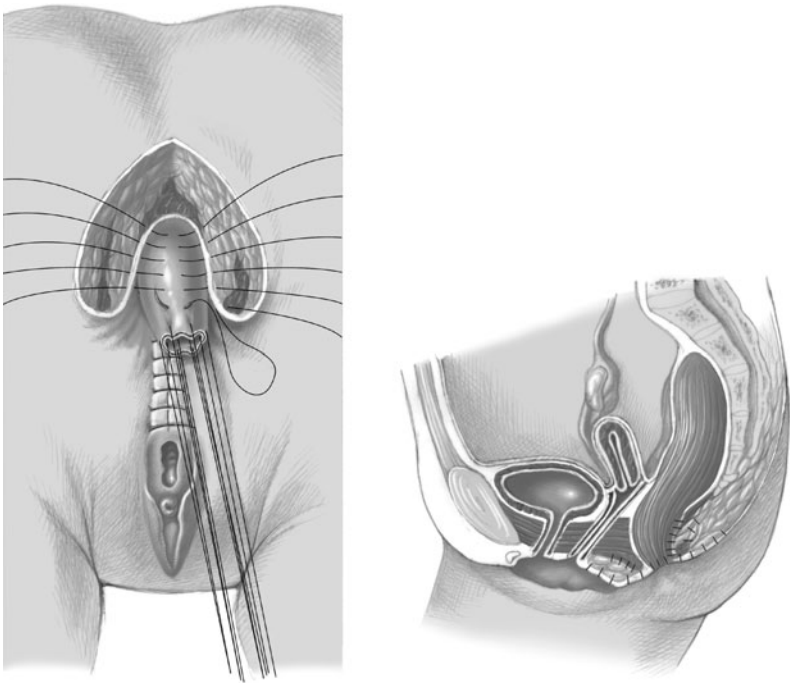


Fig. 24.12 Operative steps: repair of female anorectal defects

Operation: PSARP for anal malformations without a fistula

- Approach as in a standard operation for rectourethral fistula
- The bowel termination may lie very near the urethra in males and vagina in females
  - There may be a common wall between the rectum and genitourinary organs
  - In patients with Down's syndrome the bowel termination is usually clearly separate from the genitourinary tract
  - The bowel termination extends usually quite low within the sphincter funnel

Operation: PSARP for rectal atresia

- Approach as in standard PSARP for rectourethral fistula
- The sphincter complex is split just as much as is needed to identify the blind-ending tip of the anal canal
- The proximal rectal pouch is mobilized to reach the anal canal
  - The rectal pouch may need tapering or resection if it is extremely dilated
- Pushing a Hegar dilator through the anus easily identifies the proximal tip of the atretic anal canal
- A direct anastomosis is performed between the opened anal canal and proximal bowel
- Closure of the sphincter complex and skin as in standard PSARP

Operation: PSARVUP for cloaca (Fig. 24.13)

- The aim of the operation is the simultaneous reconstruction of the urethra, vagina, and rectum
  - The urethra is reconstructed from the common channel or the native urethra if total urogenital mobilization is applied
  - The vagina is reconstructed from available native vaginal structures if possible and placed in a functional and cosmetically appropriate position
  - If native vagina is hypoplastic or missing, a vagina is reconstructed from a bowel flap or tube, preferably using sigmoid colon
  - The rectum is mobilized and located in the center of the available sphincter structures
- The position of the patient and surgical approach are the same as described for rectourethral fistula
  - A silicone catheter is inserted into the bladder if possible; if not, into the vagina
  - The incision extends from the sacrum to the opening of the common channel
  - Midline split of the whole sphincter complex and perineal body is required in all cases

- The posterior wall of the rectum is entered above its confluence to the vagina
- The rectum is separated from the vagina or vaginas, preserving as much tissue at the fistula site as possible
- The separation of the rectum is usually easier than in rectourethral fistula
- In cases where there is a large vagina(s), minor damage to the vaginal wall is not critical, and vaginal tapering is required anyway

- If the common channel is clearly longer than 3 cm, successful mobilization of the rectum and separation of the vagina from urethra and bladder neck usually requires a laparotomy
- Separation of the vagina from the urinary tract is a demanding procedure
  - Injury of the urethra is to be avoided by all means!
  - Minor tears in the anterior wall of the vagina are not critical and may be repaired without major consequences
- If the common channel is narrow there is no need to open it
  - A wide common channel requires tapering over an indwelling catheter
- In cases where there is a short common channel (clearly less than 3 cm) and adequate urethral length, the urethra and vaginal can be mobilized en bloc (total urogenital mobilization):
  - Lateral mobilization
  - Ventral mobilization behind the pubic symphysis extends up to the bladder neck
  - The common channel is used to augment the vulval mucosal lining
  - Native urethra and vagina are retained en bloc and sutured to the vulva in an appropriate position
- If the vagina is hypoplastic it is augmented abdominally by a segment or flap of sigmoid or ileum:
  - The uterus or uteri are anastomosed to the bowel vagina; it is useful to retain a rim of vagina where proximal bowel remnant vagina are sutured

- If the vagina and uterus are absent the proximal remnant of the vagina is closed
- It is important to make the size of the vagina appropriate to the patient's age: a too large and redundant vagina may cause excessive mucous secretions and may prolapse

- The vagina is sutured to vulvar skin/mucosa exactly behind the urethral opening
- Perineal body is reconstructed behind the vagina with at least two rows of interrupted sutures
- The rectum is placed in the center of the sphincter complex and the back wall of the sphincter funnel and the parasagittal fibers are reconstructed

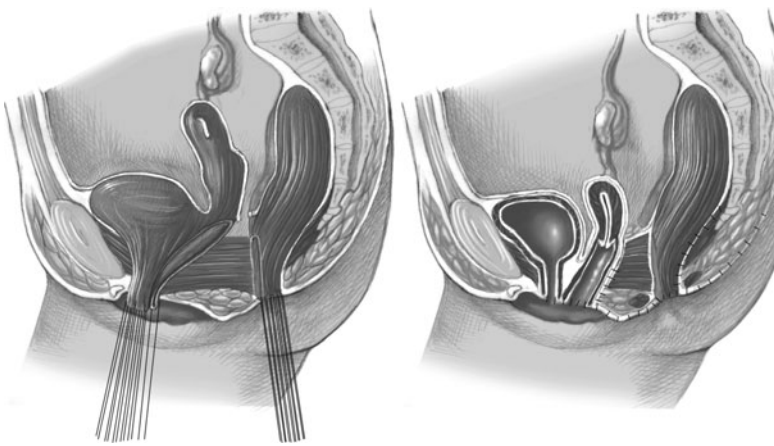


Fig. 24.13 Operative steps: cloaca

Postoperative care

- Patients with standard PSARP have a bladder catheter for 3–5 days
- Patients with limited PSARP need a bladder catheter for 2–3 days depending on the need for opioid analgesics
- Patients with a cutback do not need a bladder catheter
- Antibiotic prophylaxis is continued as long as the patient has the bladder catheter
- Anal dilatations with Hegar dilators are started 2 weeks postoperatively
  - The usual starting size is Hegar 6–10, depending on the patient's age
  - The dilation is performed once a day
  - The dilator size is changed to the next size up each week
  - The final size is Hegar 13–14 in infants younger than 6 months and 15–16 in patients older than 6 months
  - The colostomy is closed when the final size is reached
  - The dilatations are continued after closure of the stoma for 4–6 weeks at increasing intervals (2 weeks once a day, 2 weeks every other day, 2 weeks twice a week)
- Patients require vigorous follow-up during the first 2 years of life
  - During the first year, every 3–6 weeks
  - During the second year, every 3 months
  - Further follow-up every 3–12 months depending on functional outcome
- Constipation, which is the most common early functional problem (50%–70% of patients), usually develops during the first 2 years postoperatively
  - Treatment starts early and requires long-term use of laxatives
  - Daily or every other day evacuations should be ascertained
  - If the patient develops overflow incontinence an enema regimen is initiated
- The follow-up regime includes monitoring and management of the associated anomalies
  - Urogenital anomalies
  - Spinal abnormalities
  - Other gastrointestinal anomalies

Functional problems

- Fecal continence in patients with urethral or vesical fistula or cloaca develops significantly more slowly than in normal individuals
  - Constipation should be continuously treated in all patients
  - Between the ages of 3 and 5 years, 35% of patients may develop normal bowel function without soiling or constipation that requires medical treatment
  - The degree of continence depends on the integrity of the sphincter muscle and the severity of sacral malformations
  - Good prognostic signs are cleanliness during the night, clean intervals between bowel motions and the absence of fecal accidents
  - Patients with no fistula, bulbar urethral fistula or rectal atresia have better functional outcome than those with higher anomalies
- Patients with perineal or vestibular fistula may develop normal bowel function at the same age as normal individuals
  - Constipation is especially common and should be looked after and treated vigorously
  - Poor fecal continence is due to untreated constipation, mental retardation or use of inappropriate surgical technique at the primary repair
- Constipation disappears in the majority of patients when they pass puberty

Treatment of long-term fecal incontinence

- Antipropulsive medication
- Low-residue diet
- Retrograde bowel washout regimen
- Antegrade (continent appendicostomy or cecal button) bowel washout regimen
- Permanent (or temporary) colostomy
- Secondary sphincter repairs:
  - Usually by PSARP
  - May improve continence but normal bowel function unlikely
  - Indicated only if the primary pull-through was performed so that the sphincter funnel is largely intact

- Long-term results following gracilis, gluteus or levator muscle plasties are unpredictable and usually inferior to redo PSARP
- Artificial sphincter prostheses and stimulators are still at the developmental stage

Prognosis

- Final prognosis cannot be assessed before the child is clean and can control the stools
- Depends on the severity of the primary defect
  - Degree of sacral abnormalities
  - Height of the rectourogenital communication
- Severity of constipation
  - Successful early management is associated with good functional outcome
  - Need for rectosigmoid resection is associated with poor prognosis
- The approximate percentage of patients having normal bowel function beyond childhood without fecal soiling or the need for medication or protective aids is given in Table 24.9
  - Patients have had optimal surgical management
  - Patients have been followed-up regularly

Table 24.9 Approximate percentage of patients with normal bowel function after puberty

Anomaly	Percentage with normal bowel function (%)
Perineal fistula	90
Vestibular fistula	80
Bulbar urethral fistula	70
Prostatic-urethral fistula	50
Bladder neck fistula	10
Cloaca	50
Rectal atresia	90
No fistula	70

24.8 Anal Fissures

General considerations

Anal fissures are a common problem in children

- Usual onset after 6 months of age
- Etiology unclear; not always associated with constipation
- Vicious circle: fissure causes painful defecation, child holds back defecation, accumulating a large and hard stool that opens the fissure again, fissure healing is impeded
- In children anal fissures may occur anywhere in the anal circumference
- Chronic posterior fissures are uncommon in children
- Crohn's disease often presents as chronic deep anal fissures that may be multiple

Signs

- Fresh blood on stool or after defecation
- Painful defecation
- Secondary constipation
- Fissure can be diagnosed by inspection; may be situated anywhere around anal circumference
- Shallow tear in the skin–anal mucosa junction
- Rectal examination unnecessary and painful if fissure readily seen
- Endoscopy is not required for diagnosis

Conservative therapy

- Stool softeners
- Preferably bulk laxatives (lactulose)
- Enemas should be avoided
- Stimulant laxatives should be avoided
- Protection of anal skin (petrolatum jelly)
- Treatment is continued until normal painless stool pattern is reinstituted

Operative strategy

- Surgery indicated if medical management is not successful
- Aim is to lower the high resting pressure

- Anal dilatation under general anesthesia
  - Not recommended as may cause permanent sphincter damage
  - Unpredictable effect
- Rarely needed in acute fissures
- Less than 5% of anal fissures require surgery
- Chronic posterior fissures with scar formation
- Success expected if the patient has high anal resting pressure
- Biopsy samples of atypical fissures should be taken because of possible Crohn's disease

Operation

- Lateral subcutaneous sphincter myotomy
  - General anesthesia
  - Anal speculum inserted in the anal canal without anal dilatation
  - Lateral incision in the perianal skin
  - The scalpel is inserted into the intersphincteric groove
  - The blade of the scalpel is turned towards the mucosa of the anal canal
  - The distal half of the internal sphincter is severed without damaging anal mucosa
  - No permanent sphincter damage
  - Very little postoperative pain
  - Chronic fissures heal rapidly
  - Laxative therapy should be continued for at least 6 weeks postoperatively
- Excision of the fissure
  - General anesthesia
  - Anal speculum inserted in the anal canal
  - Fistula excised and wound closed primarily
  - The results appear to be comparable to those for myotomy
  - Laxative therapy continued for at least 6 weeks postoperatively
- Botox injection
  - Injection in the four quadrants in the intersphincteric groove

Prognosis

- Most fissures heal with appropriate medical management
- Recurrences are relatively uncommon
- Results of appropriate surgical management are excellent

24.9 Anal Fistulas

General considerations

- Anal fistulas have a dual age of presentation
  - During first year of life, when the majority present in males
  - In puberty, where females may also develop an anal fistula
  - An atypical age and mode at presentation indicates a biopsy of the fistula to rule out Crohn's disease
- An infection in an anal crypt leads to abscess formation that bursts into perianal skin
  - Infant fistulas are always straight and go through the subcutaneous part of the external sphincter
  - Pubertal patients may develop more complex adult-type fistulas
- There are several types of anal fistula: intersphincteric, trans-sphincteric, extra-sphincteric, and submucosal (Fig. 24.14)

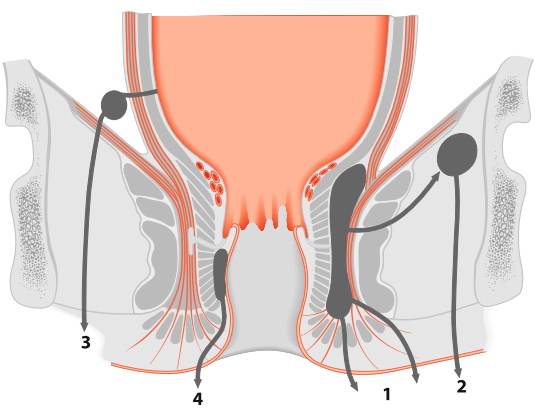


Fig. 24.14 Anal fistulas (*1* intersphincteric, *2* trans-sphincteric, *3* extra-sphincteric, *4* submucosal)

Signs

- Most infants with fistulas have a history of pre-existing perianal abscess
- Approximately half of the infants with perianal abscess develop a fistula
- Defecation may be painful
- Discharge of putrid material from the fistula opening
- Many patients have very few signs
- Long asymptomatic periods with occasional flare-ups

Conservative therapy

- If the patient has an abscess it should be drained surgically
- Antibiotic therapy usually unnecessary
- Patients should be followed-up to detect development of a fistula
- Patients with established and symptomatic fistula should be operated
- Patients with no signs may be followed-up; many fistulas heal spontaneously

Operation

- General anesthesia
- Insertion of an anal retractor
- Probing of the fistula
- Care has to be taken to identify the correct anal crypt that is associated with the fistula tract
- If a wrong crypt is opened the fistula will recur
- Fistula tract is laid open
- The mucosal tract may be excised
- Postoperative care consists of cleaning the perianal region
- Local packs or ointments unnecessary
- Rapid healing in 2–3 weeks
- Pubertal patients are treated in the same way as adults who have an anal fistula

Prognosis

- The prognosis of infant anal fistula is excellent
- Recurrences are not expected if primary management is appropriate

- Adolescent patients present in the same way as adults with fistula; recurrences are not uncommon

24.10 Rectal Prolapse

General considerations

- Rectal prolapse is a relatively common, self-limiting problem in children 1–3 years old
- Two types – mucosa only or full thickness with all layers of the wall
- Most are idiopathic but it is also seen in neuromuscular disease such as myelomeningocele, in severe constipation, severe diarrhea, cystic fibrosis, etc.

Signs

- History of repeated prolapse, e.g., after defecation, straining or crying, etc.
- At first spontaneous reduction, later on reduction by parents is required
- Sometimes mucosal bleeding, sometimes edema
- Prolapsed rectum is usually 2–3 cm long with radial mucosal folds. Full-thickness prolapse usually with circular folds

Preoperative work-up

- History usually reveals the diagnosis
- Careful rectal examination
- Occasionally sigmoidoscopy/colonoscopy
- Search for polyps, parasites
- Sweat test to exclude diagnosis of cystic fibrosis
- Soften stools with, for example, lactulose or a high-fiber diet
- Limit straining, improve diarrhea or constipation

Operation

- Rarely needed, only when no spontaneous cure occurs

- In mucosal prolapse – submucosal injection under general anesthesia in four quadrants with a sclerosing agent (e.g., Phenolic oil, 4×2 ml). Can be repeated if not successful
- In full-thickness prolapse a laparoscopic rectopexy should be performed

Postoperative care

- Continue to keep stools loose with dietary measures and/or medication

Prognosis

- In most cases very good

24.11 Colonoscopy

General considerations

- The technical development of instruments has made it possible to perform colonoscopy in young children too, thus increasing its diagnostic and therapeutic importance
- General anesthesia for most children, or sedation, is needed
- Plan the investigation to be followed by an operation, if necessary

Preoperative work-up

- Fluids only on the day before
- In most (older) children an oral enema, e.g., Golytely® or Picolax®

Instruments

- Flexible instruments, 10 mm in diameter for young children and 15 mm for older
- At least one working channel in all age groups
- Length of instrument is from 50 cm (newborn) to 120 cm (adults)
- Radiological control not necessary; it is usually sufficient to go by anatomical landmarks and use of a light source through the abdominal wall

Operation

- Start with rectal palpation to widen the anus and facilitate introduction of the instrument
- Position the child lying left-side down
- If necessary change to the supine position
- Inflate air, as little as possible, and pass the rectum
- Do not stretch the rectosigmoid to pass more easily
- When vision is reduced, back the instrument off a little. Avoid forcing and blind introduction
- To pass angles and curves straighten and rotate the instrument left and right
- For complete investigation it is necessary to inspect the last 10 cm of the distal ileum. When this is difficult, use the biopsy instrument to guide the way into the small bowel
- During instrument introduction only make a coarse inspection. During instrument extraction make a detailed inspection and perform treatment if appropriate, e.g., polyp extraction
- After investigation evacuate as much fluid and air as possible to avoid postoperative pain

Postoperative care

- Some hours rest for recovery is all that is needed
- Usual postoperative monitoring

Complications

- Bleeding after biopsy or polypectomy is possible but rare
- Bowel perforation is very uncommon
- Ruptured spleen or liver is extremely rare

24.12 Large Bowel Polyps

General considerations

- Mostly benign and singular; may be part of a syndrome, e.g., juvenile polyposis, familial adenomatous polyposis, Peutz–Jegher syndrome, etc.
- Affects boys more often than girls, age 3–10 years
- Juvenile polyps are benign hamartomas, usually within 10 cm of the anus
- Most polyps are located in the rectum (70%, sometimes seen prolapsed), sigmoid (15%), or remaining colon (15%)
- In about 25% of cases there can be more than one polyp without it being a polyposis syndrome
- Malignant degeneration is uncommon, but in Peutz–Jegher syndrome it is about 2%; in juvenile polyposis it is about 6%; and in familial adenomatous polyposis it is inevitable in the long run
- Usually an outpatient treatment

Signs

- Low intestinal (red) bleeding, sometimes chronic
- Abdominal pain

Preoperative work-up

- Rectal enema to clean the bowel
- General anesthesia

Operation

- Start with rectal palpation to determine if the polyp is very distal
- If so, ligate the base and remove the polyp
- For a more proximal polyp put the child tilted to the left
- Use a rectoscope to examine up to the sigmoid
- Localize the polyp and remove it with forceps
- Stop bleeding with cautery

- Place the cautery loop around the base of the polyp and coagulate carefully. Fluid around the polyp must be avoided as it can transmit current to normal bowel
- Be careful to touch only the polyp when coagulating and avoid contact between the neck of the instrument and normal bowel
- Draw out the polyp with the sling

Postoperative care

- Observe for a couple of hours in the daypatient ward
- Histopathological investigation necessary

Prognosis

- Up to one-quarter of children have further polyps. The polyps in juvenile polyposis syndrome are considered premalignant
- Polyps are removed when symptomatic or if the previously removed polyp is malignant

25 Inflammatory Bowel Diseases

General considerations

- Differs from adult inflammatory bowel disease (IBD) because of its more severe clinical course
- Includes ulcerative colitis, Crohn's disease, and indeterminate colitis
- Incidence about 5–8 per 100,000 per year below 16 years of age. About one-quarter have a familial occurrence
- Chronic, idiopathic inflammation

25.1 Ulcerative Colitis

General considerations

- Unknown etiology
  - Infections
  - Allergic
  - Genetic
  - Dietary
  - Psychogenic
  - Environmental
  - Multifactor
- Age usually 10–20 years
- Usually starts in the rectum, progressing proximally
- In about 95% of cases the rectum is involved. Ulcerations of mucosa and submucosa

- Chronic inflammation is complicated by perforation, bowel obstruction, and pseudopolyps
- Colon cancer, increasing in risk with duration of the disease, starting at about 4% after the first 10 years rising to about 20% after 20 years

Signs

- Diarrhea with mucus and bleeding
- Colicky pain
- Vomiting
- Fever, sepsis
- Anorexia, weight loss, growth retardation
- Chronic anemia
- Tachycardia
- Shock due to toxic megacolon
- Extracolonic manifestations such as erythema nodosum, pyoderma gangrenosum, arthralgia, arthritis, uveitis, stomatitis

Preoperative work-up

- Colonoscopy including taking biopsy samples from different levels
- Rectoscopy
- Abdominal ultrasonography
- Contrast enema
- Anal rectomanometry to assess sphincteric function
- MRI
- Continued medication (sulfasalazine and prednisolone, the latter at an increased dose)
- Bowel enema

Conservative therapy

- Sulfasalazine orally or locally, 20–30 $mg \cdot kg^{-1}$ body weight divided into three to six doses
- Corticosteroids orally, locally or intravenously, e.g., prednisone 1–2 $mg \cdot kg^{-1}$ body weight

- Supplementary vitamins and iron
- When acute, hospitalization with bowel rest, total parenteral nutrition including trace elements and vitamins, intravenous corticosteroids, antibiotics
- Endoscopic controls

Operation strategy

- Absolute indication and emergency surgery in perforation, toxic megacolon, ileus or severe bleeding not responding to therapy
- Relative indications and elective surgery in failure to thrive and growth retardation, poor quality of life, failure of conservative therapy, anemia

Operation: emergency operation

- Ileostomy and colectomy with closure of rectum according to Hartmann
- Transanal, endorectal ileal pull-through with S-pouch, J-pouch, straight ileoanal, etc.

Operation: elective operation

- Diverting ileostomy for 3 months
- In case of intact rectal mucosa – endorectal ileal pull-through with a pouch
- As an alternative – continent ileal reservoir according to Koch
- In cases of affected rectal mucosa – first conservative treatment and then as above
- Continent proctocolectomy with J-pouch (Fig. 25.1)
  - Jack-knife position
  - Transanal mucosectomy
  - Laparotomy (midline incision)
  - Colectomy with subtotal proctectomy
  - Creating of a J-pouch using a GIA stapler
  - Pull-through of the pouch through the muscular cuff
  - Suturing of the pouch to the anus

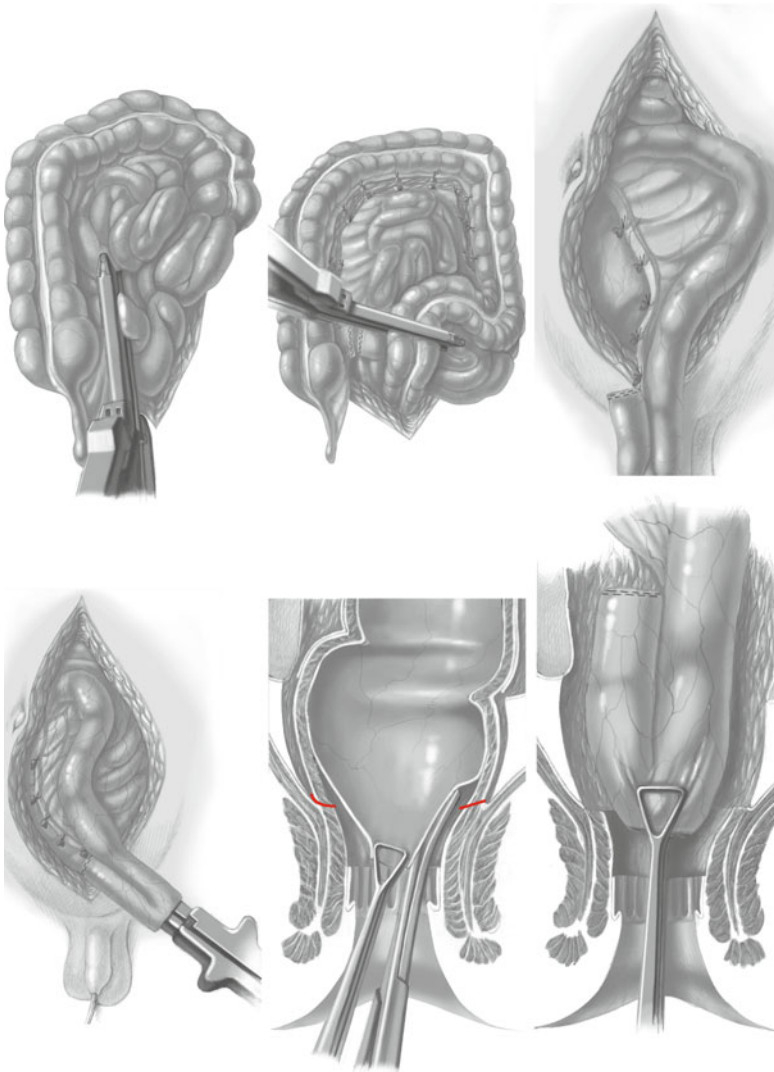


Fig. 25.1 Operative steps: colectomy and J-pouch

Summary of the procedures

- A summary of the procedures is given in Table 25.1

Table 25.1 Summary of procedures for ulcerative colitis

Operation	First operation	Second operation	Third operation
Emergency	▪ Subtotal colectomy ▪ Closure of rectum according to Hartmann ▪ Single lumen ileostomy	▪ Total colectomy ▪ J-pouch or transanal endorectal pull-through ileostomy	Taking down the ileostomy
Elective	▪ Total colectomy ▪ J-pouch or transanal endorectal pull-through ileostomy ▪ Double lumen ileostomy	Taking down the ileostomy	

Postoperative care

- Gastric tube to evacuate the stomach
- Feeding may start as soon as the child seems willing to eat, otherwise total parenteral nutrition until bowel movements begin
- Antibiotics for 5 days
- Soft drain transanally for 3 days
- Reduction of steroid dose to preoperative levels, with subsequent total withdrawal
- Teach the parents how to take care of the ileostomy

Prognosis

- Long term, about 10% experience soiling/incontinence
- Increased number of bowel movements, most children have 4–6 a day
- Complications include pouchitis (chronic reservoir inflammation), stricture formation, ileus
- Re-operation rate due to pouch problems is roughly 5%

25.2 Crohn's Disease

General considerations

- Regional enteritis
- Unknown etiology. Genetic predisposition? Antigenic influence on the mucosa? Mucosal vasculitis, micro infarcts? Chronic inflammation?
- Can involve the total gastrointestinal tract, commonly distal small bowel and colon. Small bowel only (about 25%) and colon only (30%)
- Focal, transmural inflammation with skip lesions
- Micro abscesses with mucosal ulceration
- Fistula formation common
- Multidisciplinary treatment

Signs

- Anorexia with weight loss
- Abdominal pain
- Diarrhea
- Fever
- Anemia
- Perianal problems, painful fissures, fistulas, abscesses
- Extraintestinal manifestations, arthritis, conjunctivitis, iritis, growth retardation, delayed puberty

Preoperative work-up

- Increased C-reactive protein, leukocytosis, anemia, hypoproteinemia
- Small bowel follow-through to diagnose stenosis, fistulas
- Ultrasonography with estimation of the bowel wall thickness
- Colonoscopy including biopsy

Conservative therapy

- Sulfasalazine 40–70 $mg \cdot kg^{-1} \cdot day^{-1}$
- Prednisone 1–2 $mg \cdot kg^{-1} \cdot day^{-1}$ (max. 60 mg) for 6–8 weeks
- High-calorie, high-protein diet

- Sometimes bowel rest with total parenteral nutrition with high calorie intake, 60–80 kcal·kg^{-1}·day^{-1}, electrolytes, vitamins, trace elements, antibiotics
- Metronidazole 15–20 mg·kg^{-1}·day^{-1}
- Methotrexate in steroid resistance, or possibly tumor necrosis factor alpha (TNFα) blockers
- Endoscopic controls

Operation

- Surgical treatment seldom necessary in childhood
- Absolute indication – perforation, ileus, toxic megacolon
- Relative indication – fistula, stenosis, severe perianal disease, failure of conservative therapy
- Bowel-preserving technique a principle
- Perform as limited a bowel resection as possible, with primary anastomosis if the margin is disease free (Fig. 25.2)
- Stricturoplasty preferable – longitudinal incision, transverse suturing

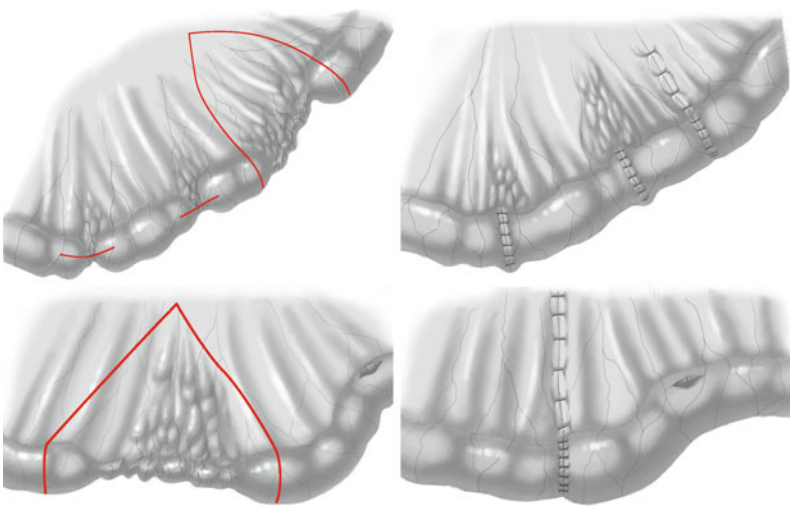


Fig. 25.2 Operative steps: bowel resection

Postoperative care

- Gastric tube to evacuate the stomach
  - Feeding may start as soon as the child wants to eat, otherwise parenteral nutrition until bowel movements start
- Antibiotic therapy for 5 days
- Successive reduction of steroids to preoperative dosages

Prognosis

- Chronic disease. Multidisciplinary management is desirable
- High incidence of recurrence, about 60%–70% within the first year
  - Re-operations not uncommon, about 50% during the first 10 years, increasing with long-term disease
- Substantial morbidity

26 Blunt Abdominal Trauma

General considerations

- Nonpenetrating violence to the abdominal wall with possible damage to parenchymatous organs
- Constitutes about 5% of all accidents in childhood
- Modern diagnostic methods have greatly facilitated diagnosis
- Possible injury to spleen, liver, stomach, duodenum, pancreas, small and large bowel, kidney, ureter, bladder, and diaphragm. Single or multiple organs affected
- Never forget child abuse as a possible cause of injury!

Concomitant injuries

- Rib fractures, lung injury
- Skull fractures, brain injury
- Fractures of the extremities
- Injury to abdominal wall muscles
- Soft tissue injury

Signs

- Sometimes, surprisingly mild signs at first
- Often increasing signs of parenchymatous organ damage
- Abdominal pain
- Referred pain to left shoulder (in ruptured spleen)
- Hypovolemic shock
- Increasing abdominal circumference

- Peritonitis
- Guarding
- Palpable tumor

Emergency care

- According to ATLS® (**A**dvanced **T**rauma **L**ife **S**upport)

Preoperative work-up

- Blood count, blood typing, cross match
- Amylase analysis (pancreatic injury)
- Urine analysis (hematuria)
- Pulmonary X-ray (effusion, hemorrhage, pneumothorax)
- Ultrasonographic examination, CT, MRI (parenchymatous injury)

Conservative therapy

- Basic attitude in abdominal trauma in children is conservative treatment, even in cases of substantial rupture of parenchymatous organs
- Thorough investigation and careful diagnosis
- Strict bed rest
- Careful, continuous monitoring (general condition, blood pressure, pulse rate, pain)
- In many cases ICU
- Parenteral nutrition during intensive care period
- Varying time in hospital [2–5 days, based on Spleen Injury Scale, grade I–IV on CT, according to American Pediatric Surgeons Association, Trauma Committee]

Operation strategy

- Free intraperitoneal gas
- Persistent shock despite therapy
- Need of transfusion more than 40 $ml \cdot kg^{-1}$ body weight
- Peritonitis

- **Spleen rupture**
  - Conservative, spleen-preserving management is important and almost always possible
  - In rare cases requiring surgical intervention, suturing, using fibrin glue or partial resection, should be first choice
  - Total splenectomy is occasionally required and autotransplantation of splenic tissue may be attempted
  - Abdominal drain to detect continuous bleeding
- **Liver rupture**
  - Conservative treatment possible in many cases
  - In case of uncontrollable bleeding, emergency laparotomy
  - Control of hepatoduodenal ligament by Pringle maneuver
  - Concomitant thoracotomy sometimes necessary to control hepatic veins
  - Surgical therapy depends on nature of injury (suture, partial resection, fibrin glue, tamponade)
  - Be careful to suture leaking gall capillaries to avoid bile peritonitis
  - Abdominal drain to detect continued bleeding or bile leakage
- **Bowel perforation**
  - Primary suture or anastomosis after resection is often sufficient
  - Resection in larger injuries with necrotic bowel
  - Protecting stoma is usually not necessary
- **Duodenal rupture/hematoma**
  - Careful exploration necessary (Kocher maneuver)
  - Primary suture and gastric tube in most cases. Carefully check the papilla of Vater and ensure bile flow
  - In rare cases duodenoduodenostomy or duodenojejunostomy
  - If only a hematoma is found, further surgery is usually not necessary
- **Pancreatic rupture**
  - In most cases conservative treatment (gastric tube, total parenteral nutrition)
  - In rare cases, suturing of the pancreatic duct or partial resection may be necessary
  - Posttraumatic pseudocysts may subsequently develop which may be treated sufficiently with percutaneous drainage

- **Diaphragmatic rupture**
  - Explorative laparotomy
  - Primary suture
  - Pleural drain, possibly abdominal drain

Postoperative care

- Depending on type of injury, but basic surgical principles prevail
- In splenectomy, especially in young children, OPSI (overwhelming post-splenectomy infection) must be considered and *Streptococcus pneumoniae* prophylaxis applied
- Prophylactic therapy with penicillin is also recommend if infections develop

Prognosis

- Depends on type of injury
- In most cases, long-term prognosis is good

27 Liver and Pancreas

27.1 Nesidioblastosis/Congenital Hyperinsulinism

General considerations

- Preferred term is congenital hyperinsulinism
- Incidence 1:50,000 live births
- Unregulated secretion of insulin in pancreatic β-cell hyperplasia due to alterations of the K^+-ATP channel (mutations of four genes, for the following: Kir6.2 and sulfonylurea, glucokinase, glutamate dehydrogenase receptor)
- Persistent hypoglycemia is often resistant to therapy

Classification

- Classification of congenital hyperinsulinism is shown in Table 27.1

Table 27.1 Classification of congenital hyperinsulinism

Pathology	Pathological correlate	Cases
Diffuse pancreas involvement	β-cells with abnormal large nuclei and abundant cytoplasm	60%
Focal adenomatous hyperplasia	Apparently normal β-cells	40%

Signs

- Signs are related to severe persistent hypoglycemia in the neonatal period
- Blood glucose of <40 $mg \cdot dl^{-1}$ in premature and term babies
- Bradycardia
- Irritability, fatigability, convulsions
- Jitteriness, tremulousness, tachycardia
- Poor feeding
- Inappropriate sweating, coma

Preoperative work-up

- Glycemic profile
- Selective transhepatic catheterization for sampling of blood glucose and insulin levels in the pancreatic veins
- Ultrasonography (not always contributory)
- CT (not always contributory)
- MRI (not always contributory)

Diagnosis

- Criteria for diagnosis are given in Table 27.2

Table 27.2 Criteria for diagnosis of congenital hyperinsulinism

Insulin	Inappropriate plasma levels of insulin in the presence of hypoglycemia Plasma glucose <40 $mg \cdot dl^{-1}$ simultaneous with elevated plasma insulin >13 $\mu U \cdot ml^{-1}$ (glucose:insulin ratio <3:1)
Glucose substitution	High glucose requirement to maintain normoglycemia >10 $mg \cdot kg^{-1} \cdot min^{-1}$
Fat	Low plasma β-hydroxybutyrate and free fatty acids <1.0 $mmol \cdot l^{-1}$
Glucagon	Inappropriate glycemic response to intravenous glucagon (rise of >30 $mg \cdot dl^{-1}$ in serum glucose level)

Conservative therapy

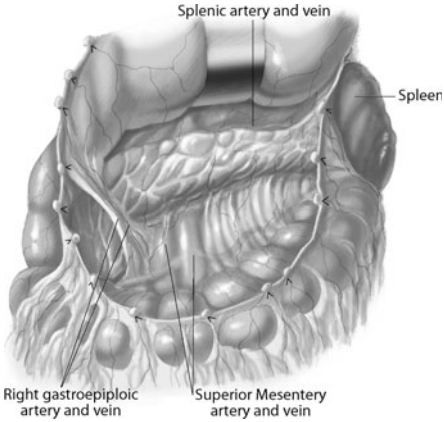
- Maintenance of normal glycemia by glucose infusion (up to 15–20 $mg \cdot kg^{-1} \cdot min^{-1}$) and/or high-calorie enteral feeding
- Diazoxide, insulin antagonist (up to 20 $mg \cdot kg^{-1} \cdot day^{-1}$) (however, many side-effects)
- Followed by octreotide or glucagon for diazoxide-resistant cases
- Hydrochlorothiazide (synergistic action with diazoxide)

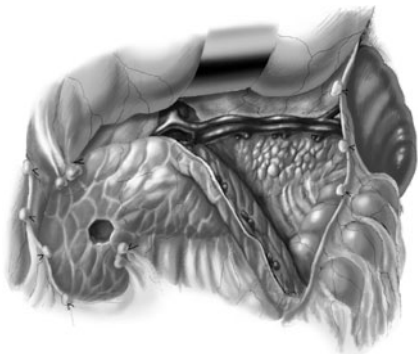
Surgical therapy

- Purpose is to reduce the mass of insulin-producing β-cells
- Control of hyperinsulinism is achieved through the extent of pancreas resection
  - For diffuse form, usually 95%
  - For focal form, less radical excision (after pancreatic venous sampling and with the help of frozen-section biopsies during surgery)
  - Resection of more than 98% may result in endocrine and exocrine insufficiency
  - Resection of less than 95% may result in failure to cure or recurrence

Operation steps

- The surgical procedure is shown in Fig. 27.1
- Make an upper transverse abdominal incision
- Open the omentum major
- Expose the pancreas after mobilization of the duodenum (Kocher maneuver)
- Perform the resection: options include the tail, body, uncinate process and the majority of the head, sparing the spleen
- Dissect the pancreas and ligate all small pancreatic arterial and venous branches
- Leave a sliver of pancreatic tissue to the left of the common bile duct and on the surface of the duodenum
- Ligate the pancreatic duct with a nonabsorbable stitch
- Seal the pancreatic parenchyma with collagen glue or equivalent
- Ensure peritoneal drainage





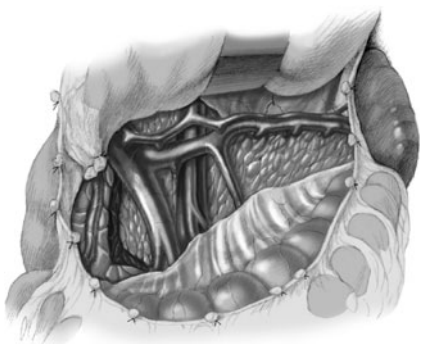


Fig. 27.1 Operative steps: pancreas resection

Postoperative care

- Gastric tube on suction
- Intravenous nutrition for 5–7 days
- Frequent serum glucose tests
- Administration of insulin as required
- Antibiotics for 5 days
- Peritoneal drain to be removed 3–4 days postoperatively
- Follow adaptation for 4–6 months

Prognosis

- 50% cure after >95% resection, 19% cure after <95% resection
- Better results in focal disease assuming complete excision and normal remaining pancreas
- Diabetes mellitus in 15%
- Possible exocrine insufficiency

27.2 Congenital Biliary Atresia

General considerations

- Incidence: 1:10,000 live births
- End result of a destructive inflammatory process, unknown etiology (viral, toxic)

Differential diagnosis (causes of conjugated hyperbilirubinemia)

- Intrahepatic hypoplasia (Alagille's syndrome)
- Alpha-1-antitrypsin deficiency
- Thick bile syndrome (inspissated bile syndrome) after hemolysis
- Neonatal hepatitis (intrauterine viral infection), giant cell hepatitis
- Sepsis with jaundice
- Cystic fibrosis
- PFIC (progressive familial intrahepatic cholestasis or Byler disease)

Classification

- Table 27.3 and Fig. 27.2 give the classification of congenital biliary atresia

Table 27.3 Classification of congenital biliary atresia

Type 1	Atresia of the choledochal bile duct with patent proximal ducts
Type 2	Atresia of the common hepatic duct, residual patency of proximal ducts
Type 3	Atresia of the entire extrahepatic duct system, involving right and left hepatic ducts towards the porta hepatis

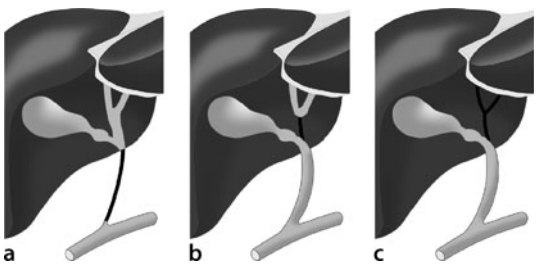


Fig. 27.2a–c Types of biliary atresia. **a** Atresia of ductus choledochus, **b** atresia of ductus hepaticus communis, **c** atresia of ductus hepaticus communis, ductus hepaticus dexter and sinister

Signs

- Early jaundice (first 36 h after birth)
- Jaundice may appear at 3 weeks
- Total bilirubin >12 μmol·l^{-1} in term infants, >15 μmol·l^{-1} in premature infants
- Prolonged jaundice >8 days in term infants and >14 days in prematures
- Conjugated bilirubin >15% of total bilirubin
- Nonpigmented stools
- Dark urine
- Increased size of liver and spleen
- Later on ascites
- Associated malformations (malrotation, situs inversus, polysplenia, preduodenal portal vein, absent inferior vena cava, cardiac defects)

Preoperative work-up

- Exclude infections, metabolic, endocrine disease or genetic disorder
- Blood sample: bilirubin, transaminases, alkaline phosphatase, γ-glutamyl transferase (γ-GT), coagulation factors
- Cytomegalovirus (CMV) and hepatitis virus
- Urine: bilirubin, urobilinogen (lacking)
- Ultrasound (not very specific), exclude choledochal cyst
- Liver biopsy (in 10% there is difficulty of interpretation)
  - Transcutaneously as a needle biopsy (beware of Bleeding!; small amount of material obtained)
  - Laparoscopically (bleeding control, adequate amount of material obtained). Can also be combined with a cholangiography
- In special cases (not mandatory)
  - Hepatobiliary excretion scans (technetium-labeled agents)
  - Percutaneous cholangiography (easier laparoscopically)
  - Endoscopic retrograde cholangio-pancreatography (ERCP)
- Vitamin K, i.v. (1 $mg \cdot day^{-1}$) for 4 days preoperatively
- Blood typing, cross-match
- Perioperative antibiotics

Surgical therapy

- Perform the operation early, before liver fibrosis or cirrhosis
- Place the patient in a prone position on the operating table to permit cholangiography
- Make an incision usable for possible future liver transplantation (transverse upper abdominal subcostal incision)
- Be prepared for possible Roux-en-Y anastomosis

Operation: Kasai portoenterostomy/extended hepatoportoenterostomy

- The surgical procedure is illustrated in Fig. 27.3
- Inspect the abdominal cavity (search for malrotation, situs inversus, preduodenal portal vein). Confirm diagnosis of biliary atresia (gallbladder may be hidden between segments 5 and 4)
- Dissect the hepatoduodenal ligament
- Mobilize the liver

- Mobilize the gallbladder from its liver bed; used as a guide to the fibrous remnant of the hepatic duct. This will lead to the porta hepatis
- Prepare the hepatic artery, right and left branches
- Ligate all lymphatic vessels
- Prepare the portal vein; follow the right and left branches as far as possible and expose the porta hepatis behind the bifurcation of the portal vein (for extended hepatoportoenterostomy; not performed in the original Kasai procedure)
- When necessary, exteriorize the liver
- Ligate all small portal branches to the caudate lobe
- Section Arantius' ligament (or ligamentum venosum), which helps mobilization of the left portal branch
- Widely expose the porta hepatis
- Cholecystectomy: excise remaining tract and tissue of porta hepatis flush with liver capsule
- Extensive numbers of bile ducts present posteriorly and laterally
- Ensure hemostasis; replace liver; create Roux-en-Y loop
- Level of jejunal section approximately 10 cm from the Treitz ligament
- Length of jejunal loop at least 50 cm, placed in retrocolic position
- Make a 3-cm-long incision on the antimesenteric border
- Make an anastomosis of jejunal loop to tissue at the porta hepatis, going far on both lateral sides, posteriorly on the caudate lobe and anteriorly on the quadrate lobe
- Any cystic dilatation not containing bile is considered not to be communicating and should be excised

Operation: portocholecystostomy

- Occasionally, gallbladder and distal bile duct are not affected by the atretic process and the gallbladder can be used for the anastomosis at the porta hepatis

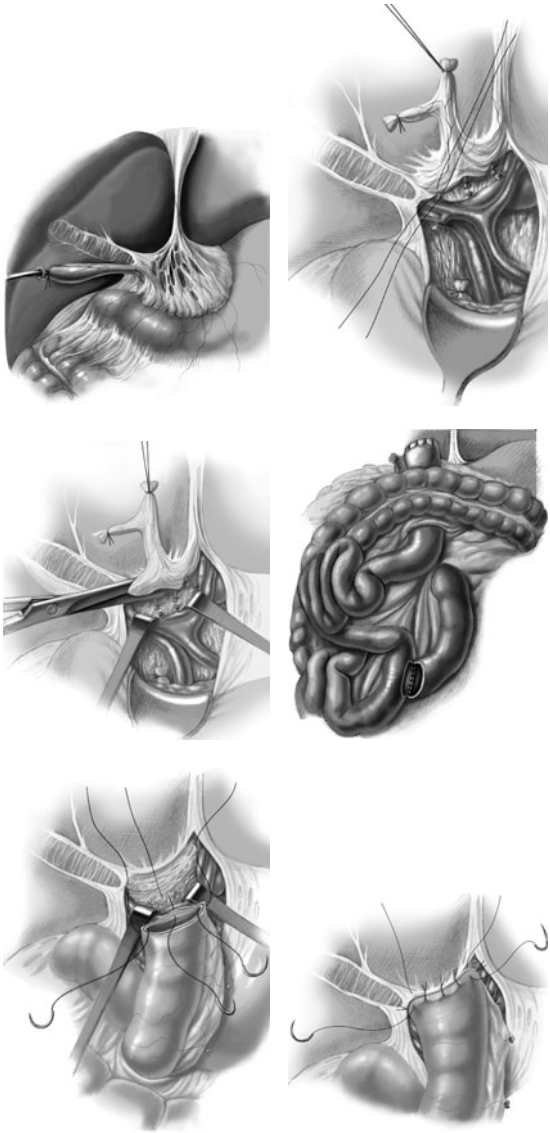


Fig. 27.3 Operative steps: extended hepatoportoenterostomy

Operation: hepaticojejunostomy

- If the residual segment of the proximal bile duct is long enough, a hepaticojejunostomy is feasible (rare type I cystic lesion)

Postoperative care

- The main problem is ascending cholangitis (best prevention: long loop of jejunum)
- Gastric tube on suction
- Initially total enteral nutrition (TEN), but enteral nutrition as soon as possible
- Antibiotics for 5–7 days, discuss continuous (for 2–3 months) prophylactic oral antibiotics (cephalosporin)
- Prednisolone, although there is no general agreement on steroids, we recommend
  - days 1, 2: 5 $mg \cdot kg^{-1}$
  - day 3: 2 $mg \cdot kg^{-1}$
  - days 4–14: 1 $mg \cdot kg^{-1}$
- Cholestyramine (if bile flow)
- Choleretics (ursodeoxycholine)
- Vitamins A, D, E, K

Prognosis

- Depends on grade of liver fibrosis or cirrhosis (late operation, more liver destruction)
- When operation is performed in first 6–8 weeks of life, chances of obtaining bile flow is approximately 70%–90%; beyond 12 weeks of age chances decrease to 35%
- 5-year survival rate with native liver is approximately 60%
- Portoenterostomy and transplantation are now complementary procedures and give good quality of life and a 80%–90% survival rate

27.3 Choledochal Cyst

General considerations

- Incidence: 1:100,000 live births
- Female:male ratio 3:1 to 4:1
- 60% of cases are diagnosed before 10 years of age
- Most frequent etiology
  - Common pancreaticobiliary channel
  - Pressure in pancreatic duct higher than in the bile duct
- Reflux of pancreatic juice in the common bile duct damages endothelium, causing cystic dilatation
- Other etiologies are also possible, such as obstruction of the distal common bile duct and genetic reasons

Classification

- Classification of choledochal cyst is given in Table 27.4 and Fig. 27.4

Table 27.4 Classification of choledochal cyst

Type 1	Cystic or fusiform dilatation of choledochus (most frequent)
Type 2	Choledochus diverticulum
Type 3	Choledochocele
Type 4	Combination of intrahepatic and extrahepatic cysts (second most frequent)
Type 5	Isolated intrahepatic duct cysts, single or multiple (Caroli's disease)

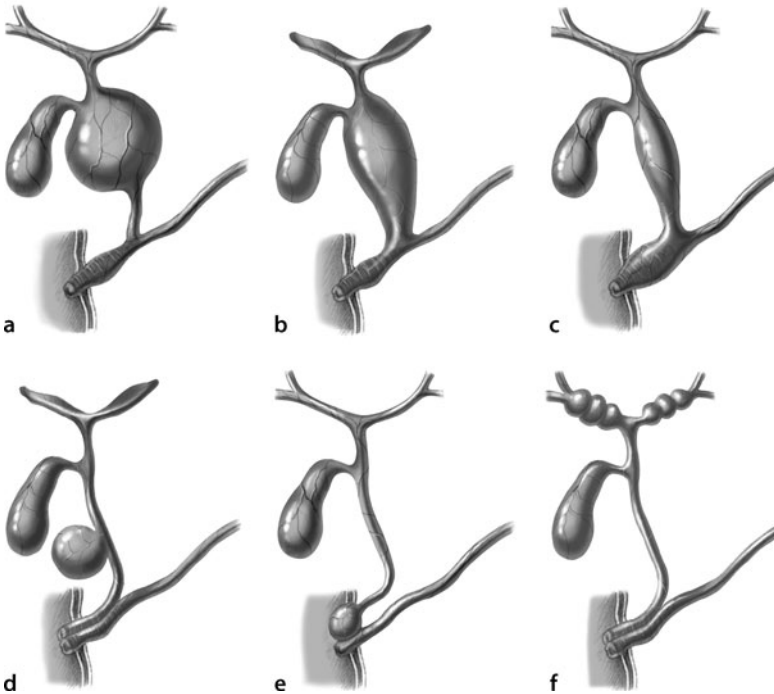


Fig. 27.4a–f Types of choledochal cysts. **a** Cystic dilatation, **b** fusiform dilatation, **c** without dilatation, **d** cystic diverticulum, **e** choledochocele, **f** intrahepatic bile duct dilatation

Signs

- Usually during the first decade of life
- Some are asymptomatic (prenatal diagnosis)
- This classic triad only present in 6% of cases
  - Abdominal mass
  - Intermittent episodes of jaundice
  - Intermittent episodes of abdominal pain
- Recurrent cholangitis
- Pancreatitis

Complications

- Biliary calculi
- Pancreatic duct calculi
- Pancreatitis
- Cyst rupture, biliary peritonitis
- Portal hypertension
- Liver fibrosis or cirrhosis
- Cholangiocarcinoma

Preoperative work-up

- Occasionally diagnosed antenatally
- Ultrasonography
- CT or magnetic resonance cholangiopancreatography (MRCP)
- ERCP
- Percutaneous transhepatic cholangiography
- Hepatobiliary scintigraphy
- Not all investigations are necessary, decision according to infrastructure

Operation

- The operative steps are illustrated in Fig. 27.5
- Early operation prevents complications
- Occasional acute pancreatitis that is resistant to conservative treatment with a gallbladder under tension requires percutaneous drainage of the gallbladder, in order to achieve resolution of pancreatitis before surgery
- Aim of surgery is complete removal of the cyst
- Cystenterostomy should not be done, because of potential complications (cholangitis, cholelithiasis, pancreatolithiasis, biliary cirrhosis, cholangiocarcinoma)
- Perioperative antibiotics
- Make a high transverse incision
- Check appearance of liver and spleen
- Perform liver biopsy
- Sample bile aspirated from the cyst for culture and pancreatic enzyme concentration

- Cholangiography to delineate precise anatomy, intrahepatic ducts and pancreaticobiliary junction
- Mobilize the gallbladder and cystic duct
- Care must be taken to avoid damage to an aberrant right hepatic artery, usually very adherent to the cystic wall
- Lift the cyst and gallbladder with a tape placed under the bile duct (pay attention to the portal vein, often adherent to the posterior wall of the cyst)
- Dissect the common hepatic duct at its bifurcation
- Further dissect the bile duct to within the head of the pancreas
- Remove the entire cyst; oversew the distal duct end
- In difficult cases, open the cyst and remove mucosa from the bottom of the cyst
- Possible calculi and debris of intrahepatic and pancreatic ducts should be cleared (intraoperative endoscopy)
- Make a roux-en-Y loop anastomosis to the hepatic duct bifurcation (wide hilar anastomosis)
- Occasionally, a transduodenal sphincteroplasty is necessary (difficulty in removal calculi from long common channel)

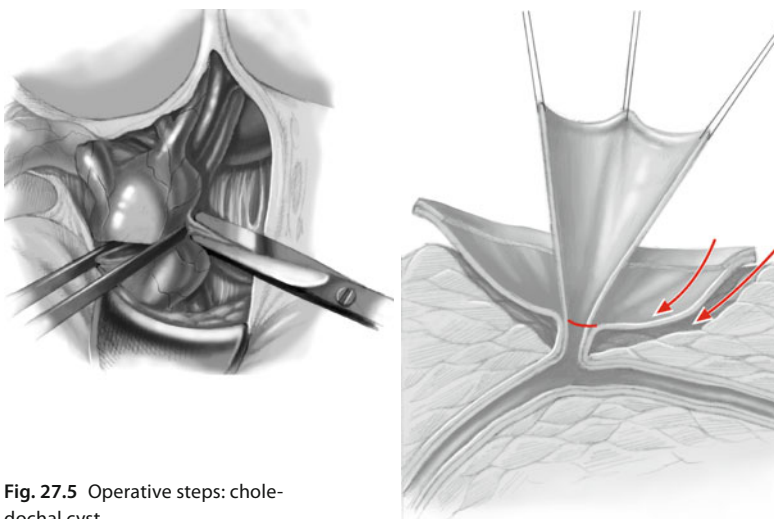


Fig. 27.5 Operative steps: choledochal cyst

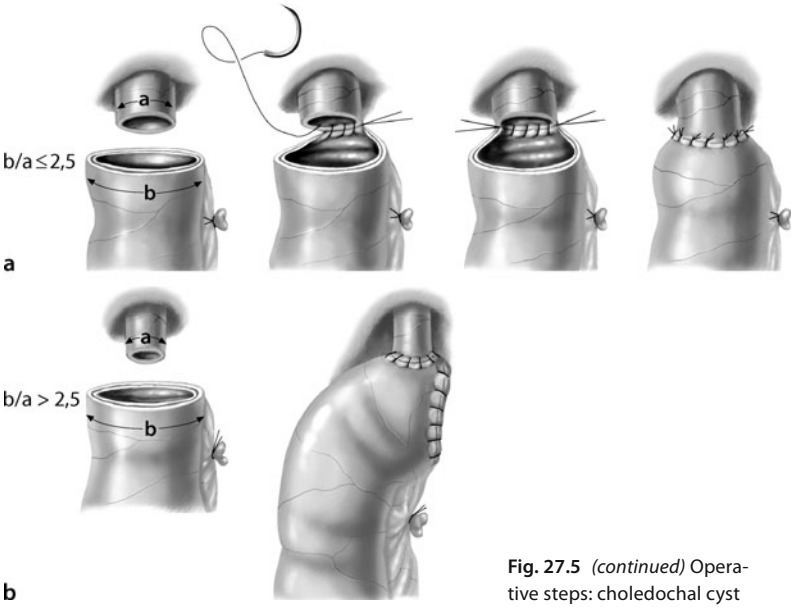


Fig. 27.5 *(continued)* Operative steps: choledochal cyst

Postoperative care

- As in biliary atresia, without prednisone and choleretics

Prognosis

- Low mortality
- 10% complications: cholangitis, pancreatitis, anastomotic stricture (even late), calculi, cholangiocarcinoma
- Operation before 5 years of age limits complications

27.4 Cholelithiasis

General considerations

- Not as frequent as in adults. More frequently discovered within the last three decades, probably because of improvements in diagnostic techniques (ultrasonography)

- Two peaks of appearance: first in infancy and second in early adolescence with a steady increase thereafter
- Female:male ratio 1:1 in infancy, 2–4:1 in prepuberty
- Table 27.5 equates pathogenic mechanisms with risk factors for cholelithiasis

Table 27.5 Pathogenic mechanisms and risk factors for cholelithiasis

Pathogenic mechanisms	Risk factors
▪ Increased bilirubin secretion ▪ Excess bilirubin pigmen	Hemolytic disorders
	Sickle cell anemia
	Thalassemia
	Spherocytosis
	Syndromes associated with hyperbilirubinemia
▪ Perturbation of enterohepatic circulation ▪ Biliary stasis ▪ Lithogenic bile	Short bowel syndrome or ileal resection
	Medication (ceftriaxone, somatostatin, ciclosporin)
	Cystic fibrosis
	Total parenteral nutrition
	Wilson's disease
Malformations of biliary tree	Biliary stricture or cysts
	Post-cholangitis stenosis

Signs

- May be asymptomatic
- In infancy nonspecific signs (poor feeding, vomiting, irritability)
- Abdominal pain (right upper quadrant or epigastrium, nausea, vomiting)
- Ileus
- Biliary colic, acute cholecystitis, choledocholithiasis with obstructive jaundice, pancreatitis

Preoperative work-up

- Detailed history, search for any general condition
- Ultrasonography
- X-ray of abdomen without preparation
- Magnetic resonance cholangiography
- ERCP, with possible papillotomy

Operation: open, conventional cholecystectomy

- Make a right subcostal incision
- Dissect the hepatoduodenal ligament and gallbladder
- Ligate and section the cystic artery
- Dissect the cystic duct and perform a cholangiography in order to visualize concrements in the duct
- Ligate and section the cystic duct
- Visualize the choledochus and excise the gallbladder in an anterograde fashion
- Ensure hemostasis of the gallbladder bed
- Drainage not mandatory
- T-tube drainage in cases of choledochus exploration

Operation: laparoscopic cholecystectomy

- Has become the standard technique (Fig. 27.6)
- If gallbladder is under tension, it may be punctured and its extremity grasped with instruments
- Gallbladder dissection begins close to Hartmann's pouch; window created above and behind the cystic duct and artery
- Cholangiogram
- Section the cystic artery and cystic duct between clips
- Dissect the gallbladder in a retrograde fashion
- Remove the gallbladder through the umbilical port (incision can be widened)
- Remove the entire pneumoperitoneum
- Choledocholithiasis must be treated (through laparoscopy, conversion to open exploration or postoperative ERCP and extraction)

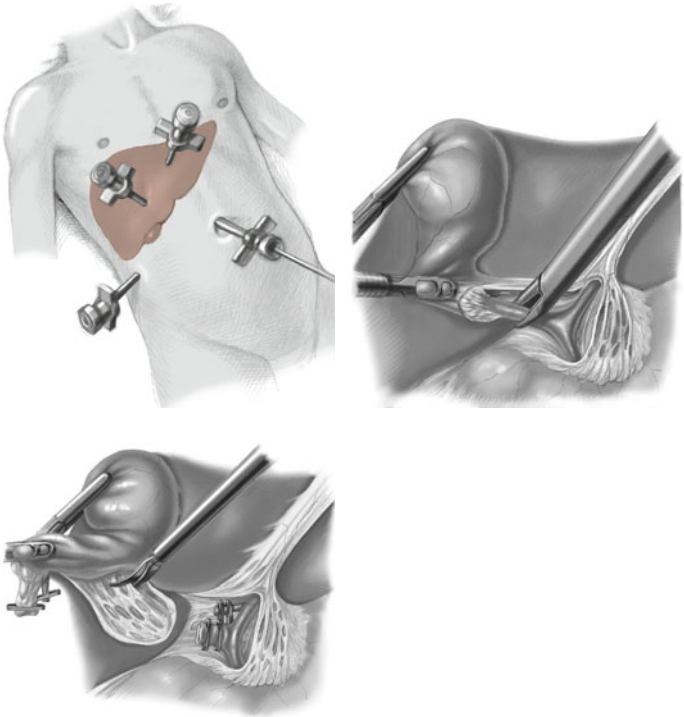


Fig. 27.6 Operative steps: cholecystectomy

Postoperative care

- Rapid recovery after laparoscopy
- Standard postoperative care according to any intra-abdominal procedure

Prognosis

- Good

27.5 Parasitic Liver Cysts

General considerations

- *Echinococcus granulosus* is found more commonly in endemic areas such as Mediterranean shores of Europe, but *Echinococcus alveolaris* can also be found
- Humans are the intermediate host contaminated orally by ingestion of parasitic eggs
- The eggs liberate scoleces, which migrate through the bowel wall into the portal vein and then to the following organs where they develop into a hydatid
  - 70% in the liver
  - 20% in the lung
  - 10% in other organs
- The hydatid cyst usually presents as a space-occupying mass, either univesicular with a thin capsule or multivesicular with a thick capsule (Fig. 27.7)
- The disease can be complicated if there is a biliary fistula or rupture into a hollow cavity occurs
- *Echinococcus alveolaris* behaves like a tissue-invading malignant tumor
- The liquid tumor has a variable size and, with time, the host considers it a foreign body and tends to push it to the organ's periphery

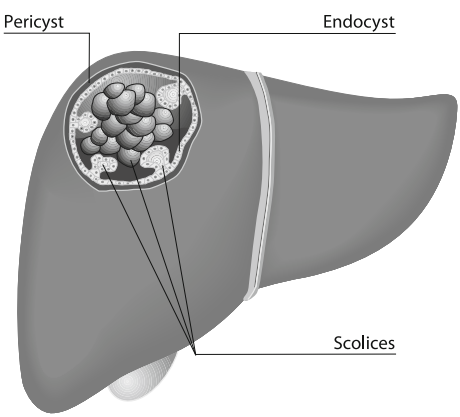


Fig. 27.7 Echinococcus cyst

Signs

- Signs differ during the two periods of parasite development: there is a pretumoral stage with a few signs and the tumoral stage that has more signs
- Palpable mass in the abdomen
- Pain due to the distension of the liver capsule
- Cholangitis if rupture into the biliary tract has occurred
- Fat intolerance

Preoperative work-up

- Blood examination showing eosinophilia, which is however not specific for parasitosis
- Ultrasonographic examination
- Plain X-ray of the abdomen potentially showing a calcified cyst
- CT shows a round mass when the cyst is univesicular or irregular multiple cyst-like structures when the cyst is multivesicular. In the latter case, the fibrotic capsule can be calcified
- Serologic tests are not in routine practice
  - Cassoni test is an immunofluorescence reaction
  - Weinberg test is a complement fixation reaction

Operation

- Make a transverse costal incision for good exposure and control of the liver pedicle
- During dissection pay close attention so as to avoid cyst rupture with dissemination of the fluid content in the peritoneum, which may lead to further infestation
- Operative cholangiography is required before and after treatment of the cyst to look for a major biliary fistula, which should be closed
- The area around the cyst must be isolated from the rest of the peritoneal cavity to avoid contamination by dressings immerged in 20% NaCl
- Puncture the cyst with injection of 20% NaCl solution for 10 min, with the aim of killing the parasite

- Suction out the cyst contents
- The first-line treatment is cystectomy (not pericystectomy due to its high risk of bleeding and complications)
- Hepatic resection may be considered
- In univesicular cysts the hydatid membrane should be excised in toto. The thin fibrotic capsule can be left in situ. The remaining cavity can be closed without drainage; it will progressively disappear following regeneration of the neighboring parenchyma
- For multivesicular cysts, aspirate the contents through a big trocar, inject 20% NaCl, and subsequently open the capsule and cyst. The capsule must be resected (pericystectomy) by cautiously peeling it off the parenchyma and vessels. Major biliary openings should be sutured. Preferably the remaining cavity should be closed and drained. A multivesicular cyst requires major surgery and the patient should remain in experienced hands

Postoperative care

- Gastric drainage
- Analgesia
- Antibiotics

Prognosis

- Good with complete cure if operation has been performed appropriately
- Relapse can occur if the abdominal cavity has been contaminated or when some parts of a multivesicular cyst have been left in place

27.6 Nonparasitic Liver Cysts

General considerations

- Congenital cysts, originating from the biliary system
- Clear liquid content; wall covered with biliary-type epithelium
- Localization most often superficial

Signs

- Most often completely asymptomatic
- In case of space-occupying mass, this is palpable
- Pain is exceptional
- Rupture in the peritoneum is exceptional
- Liver function tests are normal

Preoperative assessment

- Most often, they are discovered fortuitously
- Doppler ultrasound usually suffices to make the diagnosis
- In case of doubt, CT or MRI can be indicated

Surgical treatment

- Usually not indicated because patient is symptomless
- When the cyst volume is great and is symptomatic, de-roofing usually suffices

Postoperative care

- Gastric tube
- Intravenous fluid until resumption of bowel movements
- Abdominal drainage to be removed when discharge stops
- No antibiotics required

27.7 Benign Liver Tumors

General considerations

- General considerations are given in Table 27.6

Table 27.6 General considerations for benign liver tumors

Hemangioma	▪ Localized or diffuse in the liver (Morbus Osler) ▪ Arterio-venous shunts may lead to heart insufficiency
Hemangioepithelioma	▪ Infantile hemangioepithelioma, a benign tumor ▪ Epitheloid hemangioendothelioma, a malignant tumor with slow evolution
Hamartoma	Solid tumor, with a cystic content developing from the liver parenchyma
Adenoma	Should be distinguished from well-differentiated hepatocellular carcinoma; can transform into a malignant lesion

Signs

- Most often asymptomatic
- Palpable mass in the right upper quadrant of the abdomen
- Pain can be caused by distension of the liver capsule
- In hemangioma
  - Thrombocytopenia (Kasabach–Merritt syndrome)
  - Disseminated intravascular coagulation and anemia due to hemorrhage in the hemangioma
  - Heart insufficiency in the presence of arterio-venous shunts (Morbus Osler)
- In adenoma possible rupture into the abdominal cavity with internal bleeding

Preoperative work-up

- Ultrasonography including Doppler examination
- CT
- MRI
- Angiography in special cases (reduced due to modern imaging techniques)
- Technetium scintigraphy can help to differentiate between adenoma and focal nodular hyperplasia

Therapy

- In hemangioma
  - Conservative treatment with corticoids and interferon gamma
  - Transarterial embolization
  - Hepatic artery ligation
  - Surgical resection may be indicated in selected cases with Thrombocytopenia (Kasabach–Merritt Syndrome)]
- In infantile hemangioepithelioma
  - No treatment or surgical resection in selected cases
- In epithelioid hemangioendothelioma
  - Surgical resection, which can require a total hepatectomy (e.g., Budd–Chiari syndrome) and liver transplantation
- In hamartoma
  - Surgical resection
- In adenoma
  - Surgical resection is always indicated because of the risk of malignant transformation

Operation

- Techniques of surgical resection see Sect. 28.10

Postoperative care

- Gastric tube
- Intravenous fluid until resumption of bowel movements
- Abdominal drainage to be removed when discharge stops
- No antibiotics required

Prognosis

- Hemangioma: depends on heart damage
- Hemangioepithelioma: best prognosis given by total resection
- Hamartoma: good prognosis
- Adenoma: good prognosis

27.8 Liver Transplantation

Indications

- End-stage chronic liver diseases
  - Cholestatic origin (biliary atresia is the most frequent indication followed by Byler's disease-progressive familial intrahepatic cholestasis)
  - Alagille's syndrome
  - Cholestatic cirrhosis of unknown etiology
  - Cirrhosis of unknown etiology, post-hepatitis B cirrhosis
  - Inborn error of metabolism
    - With normal liver function, i.e., Crigler–Najjar, Wilson disease
    - With evolution towards cirrhosis, i.e., alpha-1-antitrypsin deficiency, tyrosinemia, glycogenosis cystic fibrosis
- Acute liver failure
- Unresectable liver tumor (mostly hepatoblastoma)

Preoperative work-up

- Depends on the main disease

Operation: hepatectomy and orthotopic transplantation

- Post-mortem (cadaveric) liver grafts
  - Full-size liver
  - Reduced size liver ("cut-down")
  - Split liver
- Living-related liver graft (intrafamilial donor, mostly parental)

Postoperative care/immunosuppression

- Theoretically, liver-transplanted children must take immunosuppressive drugs lifelong to prevent rejection
- The current incidence of acute rejection is around 20%–40% (mostly during the first 3 months post-transplantation), reversible by temporary increase of immunosuppression

- Incidence of chronic rejection is very low (less than 5%) in contrast with renal or cardiac transplantation
- Protocols for weaning of immunosuppression long-term after transplantation and for tolerance induction are currently under investigation

Prognosis

- Long-term survival
  - In experienced centers, long-term life expectancy exceeds 90%
  - Overall results with living-related liver graft are somewhat superior to those of cadaveric transplant; the main advantage is avoiding mortality whilst on the waiting list and being sure of the graft quality
- Quality of life
  - With steroid-containing immunosuppressive regimens, catch-up of growth is delayed until after the first post-transplantation year
  - With steroid-free immunosuppressive regimen, catch-up of growth can start as early as the second trimester after liver transplantation
  - Studies have shown that most children have full physical, mental, and intellectual rehabilitation

28 Pediatric Surgical Oncology

28.1 Pediatric Tumor Patients

Malignant neoplasms are relatively rare in children. The annual incidence of cancer in children is 14/100,000; the probability for a newborn to develop cancer within the first 15 years of life is 215/100,000 or 1 in 470 children. Treatment of a child with cancer requires an interdisciplinary tumor team (pediatric oncologist, pediatric surgeon, radiotherapist, psychologist, pathologist, etc.). As a result of this teamwork and of multinational studies more than 70% of all malignancies in children today are cured (10-year survival). Information and guidance for parents are major issues in pediatric oncology. The aim of treatment is complete remission of the tumor with the lowest possible toxicity, chance of recurrence and risk of late sequelae. Compared to adults, carcinomas are very rare in children while central nervous system (CNS) tumors, sarcomas, and blastomas are more frequent.

Therapeutic options

- Table 28.1 outlines therapeutic options and their indications

Table 28.1 Therapeutic options and their indications

Option	Indication
Chemotherapy only	Better results with conservative treatment
Preoperative chemotherapy	Down staging, initially inoperable tumors
Operation only	Radical operation possible without major risk or mutilation
Postoperative chemo-therapy	Incomplete resection, metastasis
Radiation	Large tumor mass, incomplete resection, metastasis

Classification and staging

Classification and staging of the tumor are prerequisites for the planning of treatment and are usually made according to the following:

- The patient's history
- Physical examination
- Biopsy and histology result
- Imaging (X-ray, ultrasound, CT scan, MRI)
- Intraoperative findings
- Bone marrow aspirate
- Laboratory results [blood, urine, cerebrospinal fluid (CSF)]

Epidemiology of pediatric malignant tumors

- The epidemiology of pediatric malignant tumors is given in Table 28.2

Table 28.2 The epidemiology of pediatric malignant tumors

Type of tumor	Specific disease	Incidence (%)
Leukemias		33.1
	Lymphoid leukemia	27.4
	Acute nonlymphocytic leukemia	4.9
	Chronic myeloid leukemia	0.5
	Other leukemias	0.3

Table 28.2 *(continued)* The epidemiology of pediatric malignant tumors

Type of tumor	Specific disease	Incidence (%)
Lymphomas		12.2
	Non-Hodgkin lymphoma	5.8
	Hodgkin's disease	5.2
	Burkitt's lymphoma	1.2
Leukemias		33.1
	Lymphoid leukemia	27.4
	Acute nonlymphocytic leukemia	4.9
	Chronic myeloid leukemia	0.5
	Other leukemias	0.3
CNS tumors		21.4
	Ependymoma	2.1
	Astrocytoma	9.8
	Primitive neuroectodermal tumors	4.8
	Other gliomas	1.7
	Other CNS tumors	3.0
Sympathetic nervous system		8.3
	Neuroblastoma	8.2
	Other	0.1
Retinoblastoma		2.0
Renal tumors		6.0
	Nephroblastoma	5.9
	Renal carcinoma	0.1
Hepatic tumors		1.0
	Hepatoblastoma	0.8
	Hepatic carcinoma	0.2
Bone tumors		4.5
	Osteosarcoma	2.3
	Chondrosarcoma	0.1
	Ewing's sarcoma	2.1

Table 28.2. *(continued)* The epidemiology of pediatric malignant tumors

Type of tumor	Specific disease	Incidence (%)
Soft tissue sarcomas		6.6
	Rhabdomyosarcoma	3.6
	Fibrosarcoma	0.7
	Other	2.3
Germ cell tumors		3.4
	Intracranial and intraspinal	0.9
	Other nongonadal	0.9
	Gonadal germ cell tumors	1.4
	Gonadal carcinoma	0.1
Carcinomas		1.4
	Adrenocortical	0.1
	Thyroid	0.7
	Nasopharyngeal	0.1
	Malignant melanoma	0.1
	Other and unspecified carcinomas	0.3
Unspecified malignancies		0.1

Source: German Childhood Cancer Registry, 1995–2004, malignomas in children (<15 years).

28.2 Hemangioma/Vascular Malformations

General considerations

Vascular lesions are divided in two types: hemangiomas and vascular malformations resulting from errors occurring during the development of vascular structures (arteries, veins, capillaries, lymphatics)

- Hemangiomas – the most common benign tumor of infancy
  - Incidence – about 2.5% of term infants (increases to 10% by 1 year of age)
  - Cutaneous hamartomatous lesions of blood vessels, usually present at birth, can grow rapidly in the first few months
  - Hemangiomas may be capillary, venous, arterial or mixed and can either be entirely superficial or have a deeper component
  - Approximately 80% of hemangiomas grow as a single tumor; 20%, in multiple sites
  - Females are affected 3–5 times more frequently than males
  - Many hemangiomas undergo spontaneous involution: 90% until 7 years of age (congenital hemangiomas by 8–14 months)
- Vascular malformations – relatively common in neonates
  - Always present at birth
  - The incidence is similar in females to that in males
  - They almost never regress
- Associated syndromes
  - Sturge–Weber syndrome, associated with cerebral lesions and epilepsy
  - Kasabach–Merritt syndrome, platelet trapping leads to thrombocytopenia/bleeding
  - Klippel–Trenaunay syndrome, growth changes/high output cardiac failure

Classification of hemangiomas

- Classification of hemangiomas is given in Table 28.3

Table 28.3 Classification of hemangiomas

I	Neonatal staining (most frequently on the forehead, base of the neck – usually spontaneous involution)
II	Intradermal capillary hemangiomas ▪ Salmon patch (light pink to rust in color, usually does not change during childhood) ▪ Port wine nevus (most frequently on the posterior surface of the neck, always present at birth, pink to purple in color) ▪ Spider angioma (present at 3–5 years, a small central artery with radiating intradermal capillaries, from a few to several centimeters in diameter)
III	Juvenile hemangiomas (spontaneous involution after a rapid growth period) ▪ Strawberry mark (a pale halo with radiating telangiectasia) ▪ Strawberry capillary hemangioma (sometimes at birth, more often appears within the first 3 months, most frequently on the face, scalp, back, anterior chest, may be multiple) ▪ Capillary cavernous hemangioma (a deep subcutaneous hemangioma, most often in the neck and trunk, can be in combination with lymphatic malformation)
IV	Arterio-venous fistulas (are present at birth and become evident in childhood as pulsative tumor with warmth, palpable thrill, audible bruit, or visible pulsation) ▪ Arterial hemangioma ▪ Hemangiomatous gigantism
V	Cirsoid angioma (racemose aneurysm)

Complications

- Local complications
  - Hemorrhage
  - Infection
  - Ulceration
  - Necrosis
- General complications
  - Severe anemia in Kasabach–Merritt syndrome (combination of rapidly enlarging hemangioma and thrombocytopenia)
  - Compression of vital structures (cervicofacial hemangioma)
  - Congestive heart failure and gastrointestinal bleeding

Preoperative work-up

- Diagnosis based on history and physical examination
- Photographs (to illustrate involution)
- Coagulation studies (in cases of multiple hemangiomas or rapid growth of hemangioma)
- Ultrasonographic examination including Doppler (diagnosis of hemangiomas in internal organs)
- CT, MRI, MR angiography in cases of vascular malformations and internal hemangiomas

Therapeutic principles

- Treatment of hemangiomas has to be individualized
- The location, subcutaneous distention and size are parameters used to decide appropriate treatment
- Various options can be applied in different circumstances with varying success
  - Systemic steroids for one month (prednisolone 2 $mg \cdot kg^{-1} \cdot day^{-1}$). In case of regression therapy for another month
  - Intralesional steroid injection
  - Subcutaneous interferon α-2a 3×0^6 $U \cdot m^{-2} \cdot day^{-1}$
  - Compression therapy and sclerosant injection to reduce bulk and bleeding
  - Intravascular embolization with internal prostheses (microspheres) for arterial lesions
  - Surgical excision to debulk and decompress
  - Surgical excision with primary closure/split skin grafting/local flaps
  - Cryotherapy for superficial hemangiomas
  - Interstitial laser (Nd-YAG) under general anesthetic for large hemangiomas
  - Laser therapy to improve skin discoloration and cosmetic appearance

Conservative therapy

- Uncomplicated hemangiomas are treated conservatively (in 90% of cases there is spontaneous involution)
- Continuous compression treatment (intermittent pneumatic compression) – in hemangiomatous gigantism
- Systemic steroid therapy with prednisolone 2–4 $mg \cdot kg^{-1} \cdot day^{-1}$ for 4–6 weeks. The dose can be then decreased until involution occurs (on average, 8–10 months)
- Intralesional steroids
- Interferon α treatment (long-term therapy from 12 to 18 months)
- Embolization (in Kasabach–Merritt syndrome and for large arteriovenous malformations, sometimes as preoperative preparation)
- Sclerotherapy, radiation and chemotherapy – in individual cases
- Argon laser photocoagulation for superficial hemangiomas

Surgical therapy

- Surgical excision (only in cases of complicated hemangiomas and in arteriovenous malformations)
- Nd-YAG laser
  - Interstitial laser with a bare fiber (1000 $J \cdot cm^{-3}$)
  - Transcutaneous laser for superficial hemangiomas
- Combination of surgical resection and laser

28.3 Lymphangioma/Cystic Neck Hygroma

General considerations

- Benign hamartomatous tumor of the lymphatic system, characterized by multiple communicating lymphatic channels and cystic spaces
- May present as unilocular or multicystic masses, with a thin, often transparent, wall
- About 45% are apparent at birth, 85%–90% do not appear until the age of 24 months
- Head, neck, and oral (tongue) localization in about 75% of cases

- These tumors can grow to a large size and may be detected on prenatal scans
- Related to defective thoracic and right lymphatic duct drainage into the venous system in the neck
- More frequent on the left and sited at the angle of the jaw, extending into both triangles of the neck and ramifying through all normal neck structures
- Larynx involvement is rare
- Spontaneous regression is not characteristic

Classification

- Classification of lymphangioma/cystic neck hygroma is given in Table 28.4

Table 28.4 Classification of lymphangioma/cystic neck hygroma

Lymphangioma simplex	Small capillary-sized dilated lymphatic channels in the dermis and epidermis	Oral region, tongue
Cavernous lymphangioma	Dilated lymphatic channels, often with a fibrous covering, may extend deep into the muscles and surrounding tissue	Tongue, cheek, thorax, retroperitoneum, extremities
Cystic lymphangioma	Cystic hygroma (multiloculated cystic spaces)	Neck 75% Axillary region 20%
Lymphangiohemangioma	Combination of lymphangioma and hemangioma	

Complications

- Infections
- Bleeding into the lymphangioma
- Respiratory obstructions
- Dysphagia (in large submandibular cystic hygromas)
- Partial intestinal obstructions (in large intra-abdominal lymphangiomas)

Signs

- Multiloculated, cystic, diffuse mass – readily transilluminable
- May cause respiratory distress and problems with feeding
- Spreads to the floor of the mouth, the tongue, the thoracic inlet, and axilla
- Infection leads to inflammation and increased tension
- Bleeding may lead to rapid enlargement and pain
- Can extend to the base of the skull with risks of neurological problems

Preoperative work-up

- Antenatal diagnosis
- Accurate physical examination (compressible, and cystic tumor)
- Ultrasonographic examination
- Chest X-ray examination (in cases of intrathoracic process)
- CT
- MRI scan helps to delineate the spread and displacement of anatomical structures
- Respiratory function (blood gases, oxygen saturation monitoring)
- Direct flexible laryngobronchoscopy
- Antibiotics advised

Conservative therapy

- Uncomplicated cases may be observed initially – though growth with the child is likely
- Percutaneous sclerotherapy of large cysts leads to swelling, pain, and fever but ultimately to shrinkage
  - OKT 3
  - Bleomycin
  - 33% glucose solution

Surgical therapy

- Surgical excision at 2–6 months of age
- Complete resection rarely possible
- Staged resection (necessary in about 30% of cases)
- Draining of the cysts by partial resection

Operation

- Carefully position the patient with their neck extended (Fig. 28.1)
- Make a transverse incision in the skin crease
- Exploration/excision requires great care to avoid nerve (facial or hypoglossal) or vascular damage (thyroid vessels)
- Limited or partial excision is often the only possibility
- Interstitial laser (Nd-YAG)

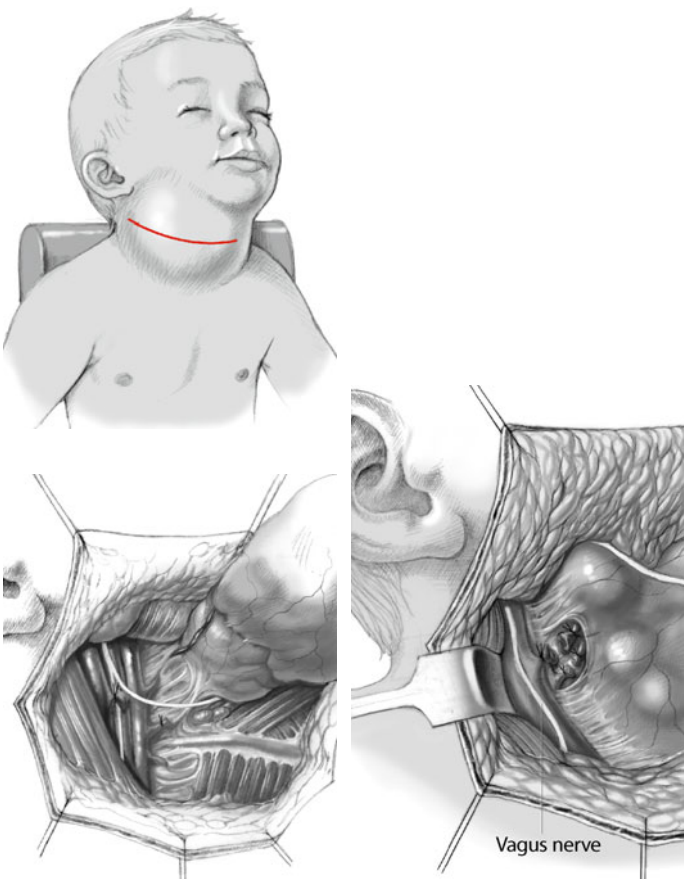


Fig. 28.1 Operative steps: cystic neck hygroma

Complications

- Infection
- Fistula formation
- Nerves and vascular injuries
- Recurrence (approximately 10%)
- Cosmetic deformity

Prognosis

- Surgical mortality 2%–6%
- Multiple procedures required for success

28.4 Lymphoma

General considerations

- Malignant disease of the lymphatic system
- 60% nonHodgkin lymphomas of high malignancy
- 40% Hodgkin lymphoma
- Surgery mainly supports conservative therapy by
  - Diagnostic biopsy
  - Staging laparotomy or laparoscopy (in selected cases)
  - Port implantation for the chemotherapy

Classification

- The Ann-Arbor classification of lymphoma is given in Table 28.5

Table 28.5 The Ann-Arbor classification of lymphoma. (*Diaphragma side* Thoracic side and abdominal side, *Ln* lymph nodes)

Stage	Regions of involved Ln	Extralymphatic organs	Spleen involved	Diaphragma side
I	One	Or one	No	Same side
II	2 or more	Plus one	No	Same side
III	More than 2	Plus one	Yes	Both sides
IV	Diffuse	More than 1	Yes	Both sides
I–IV A	Without general signs			
I–IV B	With general signs (see below)			

Non-Hodgkin Lymphoma (Kiel classification)

- Low-grade malignancy
- High-grade malignancy

Signs

- Hodgkin lymphoma
  - Indolent cervical lymph node packages
  - Hepatosplenomegaly
  - Fever with sweating especially at night
  - Weight reduction >10%
- Non-Hodgkin lymphoma
  - Hard mediastinal lymph node packages

Preoperative work-up

- Blood examination with differentiation
- Bone marrow puncture
- Chest X-ray
- Ultrasonographic examination
- CT and MRI of the abdomen and thorax
- Bone scintigraphy

Operation

- Lymph node biopsy for histologic classification
- Elective laparotomy or laparoscopy for staging with lymph node biopsies

Prognosis

- Hodgkin lymphoma: stage I, II 90%; stage III 70% 5-year remission rate
- Non-Hodgkin lymphoma: about 90% survival after 5 years

28.5 Neuroblastoma

General considerations

- The most frequent extracranial tumor in childhood (8%–10% of childhood malignancies)
- Incidence ranges from 6 to 10 cases per million per year
- Median age of diagnosis is approximately 2 years
- Embryologically originates from neural crest structures
  - Adrenal medulla
  - Sympathetic ganglia
- Neuroblastoma is an embryonic neoplasm arising from neuroblasts of any existing sympathetic ganglia. It may be in the abdomen, the thorax or neck, very rarely in the central nervous system. It is located most frequently in the adrenal gland, or in the thoracic or abdominal sympathetic ganglia extending along the vertebral column
- 60% in abdomen and pelvis, 33% in the chest, 5% in the neck
- At the time of diagnosis most children already have metastases (lymph nodes, liver, bone, bone marrow) and are classified as stage 4
- Spontaneous regression is seen in a certain number of infants (stage 4-S) and maturation during treatment toward less malignant histological variants in older children has been observed
- The recommendations in this chapter are general. It is advisable to be familiar with the protocol currently used for these patients by the

oncology team in your hospital. Help your oncologist to stay within the frame of requirements of the protocol/study

Classification

- Neuroblastoma classification is made using the International Neuroblastoma Staging System (Table 28.6) and the Brodeur risk system (Table 28.7)

Table 28.6 International Neuroblastoma Staging System (INSS)

Stage	Definition
1	Localized tumor confined to the area of origin Complete gross excision with or without microscopic residual disease
2 a	Unilateral tumor with negative lymph nodes Incomplete gross excision
2 b	Unilateral tumor with positive ipsilateral regional lymph nodes Complete or incomplete gross excision
3	Tumor infiltrating across the midline, with or without regional lymph node involvement or Unilateral tumor with contralateral regional lymph node involvement or Midline tumor with bilateral lymph node involvement
4	Dissemination of the tumor to bone, bone marrow, liver, distant lymph nodes and/or other organs (except as defined in stage 4-S)
4-S	Localized primary tumor as defined for stage 1 or 2 with dissemination limited to liver, skin, and/or bone marrow

Table 28.7 Classification (Brodeur risk grouping). (*DI* Diploid index, *LOH* loss of heterozygosity, *TRK* receptor tyrosine kinase)

Risk	Characteristics	Cure rate
Low	▪ INSS 1, 2 or 4-S ▪ Age usually <1 year ▪ No N-myc amplification ▪ Hyperdiploid/triploid with DI>1.25 ▪ No 1p loss of heterozygosity (LOH) ▪ high TRK expression	> 90%
Intermediate	▪ INSS 3 or 4 ▪ Age usually >1 year ▪ No N-myc amplification ▪ Near diploid or tetraploid ▪ Low TRK expression ▪ 1p allelic loss/other structural change	> 25%
High	▪ INSS 3 or 4 ▪ Age 1–5 years ▪ N-myc amplified ▪ Near diploid or tetraploid ▪ 1p LOH ▪ Low TRK expression	<5%

Signs

- Depend on location (wherever sympathetic nervous tissue may be found) and the behavior ("aggressiveness") of the tumor
- Palpable, hard, and fixed abdominal mass (abdominal presence)
- Respiratory distress may in theory be caused by a thoracic neuroblastoma, but far more frequently by an enlarged liver due to metastatic spread in infants staged 4-S
- Neurological signs are related to the tumor's location near/within the nervous lines and plexi or if the tumor penetrates the intervertebral foramina and invades the vertebral canal (Dumbbell neuroblastoma) producing paraparesis or paraplegia. Ataxia and opsomyoclonus are also described
- Upper thoracic or lower cervical neuroblastoma may produce Horner's syndrome

- Hypertension due to elevated level of catecholamines or compression of the renal artery
- Anorexia
- Fever (the last two signs are usually an indication of metastatic spread)
- Painless subcutaneous nodules or lymphadenopathy unresponsive to antibiotics (classical signs of dissemination)
- Enlarged liver in stage 4-S
- Skin – subcutaneous metastases = blue nodules ("blueberry muffin baby") in stage 4-S
- Painful metastases to bones
- "Raccoon eyes" or "Panda eyes" resulting from metastatic spread to the orbits
- Diarrhea with dehydration and hypokalemia due to secretion of vasoactive intestinal peptide (VIP) by the tumor

Differential diagnosis

- In the abdomen
  - Other intra-abdominal (mainly retroperitoneal) tumors and lesions
  - Nephroblastoma or other renal masses
  - Adrenocortical carcinoma
  - Metastases to the para-aortic lymph nodes at the level of the renal vessels (from germinal tumors of gonads)
  - Lymphoma
  - Teratoma
- In the thorax
  - Teratoma
  - Congenital malformations
  - Lymphoma
- In the neck
  - Congenital malformations
  - Soft tissue sarcoma
  - Neck cysts
  - Enlarged lymph nodes

Preoperative work-up

- Imaging of the primary tumor
  - Ultrasonographic examination
  - CT and/or MRI
  - Rarely arteriography or cavography
- Imaging of sites of possible metastasis
  - Ultrasonographic examination of the abdomen
  - CT of the abdomen
  - Chest X-ray and/or chest CT
  - Bone scintigraphy
  - X-ray/CT/MRI of those sites that are painful or show evidence of isotope accumulation
- MIBG scintigraphy to assess the skeletal and extra-skeletal extent of disease (MIBG is fixed specifically by neuroblasts)
- 24-h urine collection to assess the excretion of catecholamines and their breakdown products vanillylmandelic acid (VMA), homovanillic acid (HVA), dopamine, 3,4-dihydroxyphenylalanine (DOPA), adrenaline and noradrenaline. In the majority of patients these levels are high; metabolites may also serve as markers for follow-up
- Serum neuron-specific enolase (NSE) and serum ferritin
- Hematological and metabolic tests including differential blood count, C-reactive protein (CRP), electrolytes, creatinine, uric acid, liver enzymes
- Bone marrow aspirates and trephine – from different sites
- Molecular biology examinations
  - N-myc proto-oncogene can be detected early with monoclonal antibodies, high amplification correlates with poor prognosis
  - NSE is a nonspecific biochemical marker whose level is high in neuroblastoma
  - Lactate dehydrogenase (LDH) and ferritin levels are commonly high

Therapeutic principles

Therapy should be risk-adapted (for risk grouping see Table 28.7) and is usually conducted according to ongoing protocol/studies

- Low-risk neuroblastoma is usually treated with surgical excision alone (stage 1) or combined with short postoperative chemotherapy (stage 2a, b and 3). Treatment of 4-S tumors remains controversial: some authors advocate resection of the primary tumor; others, minimal chemotherapy or liver irradiation if the respiratory distress is significant
- Some ongoing studies recommend the "wait and see" policy in very young patients with low-stage, low-risk neuroblastoma, as spontaneous regression may occur
- Intermediate-risk neuroblastoma treatment consists of primary or secondary surgery, chemotherapy and radiotherapy
- High-risk neuroblastoma surgery is usually secondary. An aggressive chemotherapy, radiotherapy, and megachemotherapy followed by bone marrow transplantation or blood stem cell rescue are frequently necessary. Intraoperative radiotherapy, according to the Heidelberg Team's experience and treatment with MIBG according to Amsterdam Team's experience, may offer a chance to eradicate residual disease that classically is not resectable. Therapy with retinoids and cytokines is showing promise
- Dumbbell neuroblastoma with paraplegia or paraparesis usually responds very well to chemotherapy. Surgical decompression of the spinal cord may then be avoided. However, if the neurological signs progress, despite chemotherapy, decompression through laminotomy rather than laminectomy should be considered to prevent incurable neurological impairments

A rough guideline is shown in Table 28.8.

Table 28.8 A rough guide to the treatment of neuroblastoma

Stage	Preoperative	Operation	Postoperative
1	No therapy	Resection	No therapy
2a	No therapy	Resection	< 1 year: no therapy Others: chemotherapy/radiation
2b	No therapy	Resection	Chemotherapy/radiation
3	Chemotherapy	Complete resection if possible Intraoperative radiation performed in some centers	Chemotherapy/radiation
4	Chemotherapy	Complete resection if possible Intraoperative radiation performed in some centers	Chemotherapy/radiation Autologous bone marrow transplant
4-S	No treatment, unless for physiological reasons (e.g., respiratory distress), in which case minimal chemotherapy and/or radiotherapy may be indicated; consider surgery to release abdominal pressure and possible primary resection in the long term		

Operation: biopsy

- The applied method should be related to tumor location, the patient's condition, and pathology/molecular biology requirements
  - Laparotomy or thoracotomy and classical open biopsy
  - Laparoscopy or thoracoscopy with biopsy
  - CT/USG-guided true-cut
  - Fine needle aspiration
- Note that puncture biopsies are not very useful, as they may not provide enough tissue for the number of tests necessary, including molecular biology tests

Operation

- Classical adrenal neuroblastoma, if not invading the central vessels, is usually easily resectable. More invasive tumors, surrounding central vessels, mesenteric artery, and/or celiac trunk, are extremely difficult and complete resection is rarely possible

- Resections must be carried out through an extensive, transverse approach, with special care taken not to damage vital vascular structure as well as the renal pedicles on at least one side (Fig. 28.2)
- In order to better distinguish post-chemotherapy scars from foci of vital tissue, some authors use detection following the administration of MIBG. Tsuchida, proposed a very extensive resection of the tumor, lymph nodes and retroperitoneal tissue in cases of so-called central abdominal neuroblastoma with unfavorable factors of prognosis [Sawada T, Tsuchida Y, Voute PA, Brodeur GM (eds) *Neuroblastoma*, 2nd revised edition. Amsterdam: Elsevier, 2000]
- Intra-operative radiotherapy seems to improve local-regional control with quite acceptable risk

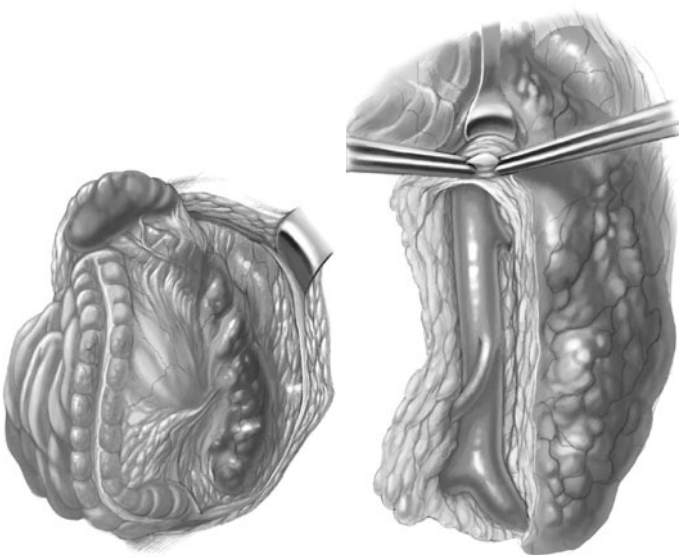


Fig. 28.2 Operative steps: neuroblastoma

Complications

- Hemorrhage
- Renal ischemia
- Intestinal ischemia
- Prolonged ileus, bowel obstruction, intussusception
- Chylous ascites
- Vascular injuries with consequent morbidity

Postoperative care

- Monitoring of blood pressure, central venous pressure, serum glucose, and urine output for at least 48 h
- For perirenal location Doppler ultrasonography is used to evaluate kidney vascularization at 24 h and 48 h
- Oncological follow-up should be performed regularly, no less frequently than every 3 months, even for prolonged follow-up. It should be discussed and planned together with the pediatric oncologist responsible for the patient's nonsurgical treatment
- Total parenteral nutrition considered in rare cases

Prognosis

- Depends on a number of factors: age, stage, location, and variant of histology, but mainly on the molecular biology of the particular tumor
- Prognostic molecular biology factors
- N-myc amplification
  - Fewer than three copies – favorable prognosis
  - Between 3 and 10 copies – unclear impact
  - More than 10 copies – unfavorable
- DNA ploidy
  - Hyperdiploid (triploid, quadriploid) correlates with good response to chemotherapy, low stages and good prognosis
  - Diploid karyotype correlates with poor response to chemotherapy and poor prognosis

- TRK (**t**yrosine **k**inase **r**eceptor) gene
  - Highly expressed – good prognosis
  - Not expressed – poor prognosis and lack of ability to maturate
- Loss of heterozygosity (LOH) on chromosome 1 (1p LOH) – poor prognosis
- High level of NSE and serum ferritin – poor outcome
- Histological classifications according to Shimada or Joshi, also on the evidence of prognostic value
- A number of other factors have been and are currently being studied; the above-mentioned factors have confirmed value and application in clinical practice
- See also the risk groups described in Table 28.7
- Screening of VMA in the urine is regularly done in Japan; however, this does not seem to influence either the prognosis of neuroblastoma significantly or the rate of low-stage cases picked-up by this method

28.6 Sacrococcygeal Teratoma

General considerations

- A congenital tumor containing derivatives of the three embryonic germ layers, arising at the tip of the coccyx
- Incidence 1 per 35,000–40,000 live births
- Unexplained female preponderance of 3:1
- Partly solid, partly cystic tumor
- Expansion behind the rectum
- More than 90% of the tumors at birth are benign
- After 6 months 50% are malignant
- Neural deficit not usual
- May be associated with hydrops

Classification

- The classification of sacrococcygeal teratoma according to the surgical section of the American Academy of Pediatrics is given in Table 28.9)

Table 28.9 Classification of sacrococcygeal teratoma according to the surgical section of the American Academy of Pediatrics

Stage	Appearance	Extension	Incidence (%)
I	Exterior		47
II	Dumbbell	Exterior plus intrapelvic extension	34
III	Dumbbell	Exterior plus intrapelvic extension plus intra-abdominal parts	9
IV	Sessile or purely intra-abdominal (often delayed diagnosis)		10

Differential diagnosis

- Myelomeningocele
- Benign ganglioneuroma
- Rhabdomyosarcoma
- Schwannoma

Signs

- Intrauterine heart insufficiency due to size of tumor and blood volume overload
- Symmetrical sacrococcygeal tumor
- Anus displaced ventrally
- Palpable at rectal examination
- May rupture and/or bleed

Preoperative work-up

- Rectal examination
- Ultrasound, often prenatally diagnosed
- X-ray (bones, teeth = teratoma)
- Contrast study of the rectum to determine compression
- α-Fetoprotein (also for recurrence control)
- MRI

Operation

- The operative steps are illustrated in Fig. 28.3
- In the first days of life, as an in toto extirpation

- As an emergency in cases with perforation, bleeding, or high risk of infection
- Incision at the level of the tumor and dorsal exposure
- Coccygectomy and control of the middle sacral vessels
- Dissection of the tumor and exposure of the proximal rectum
- Reconstruction of the pelvic floor
- Skin closure

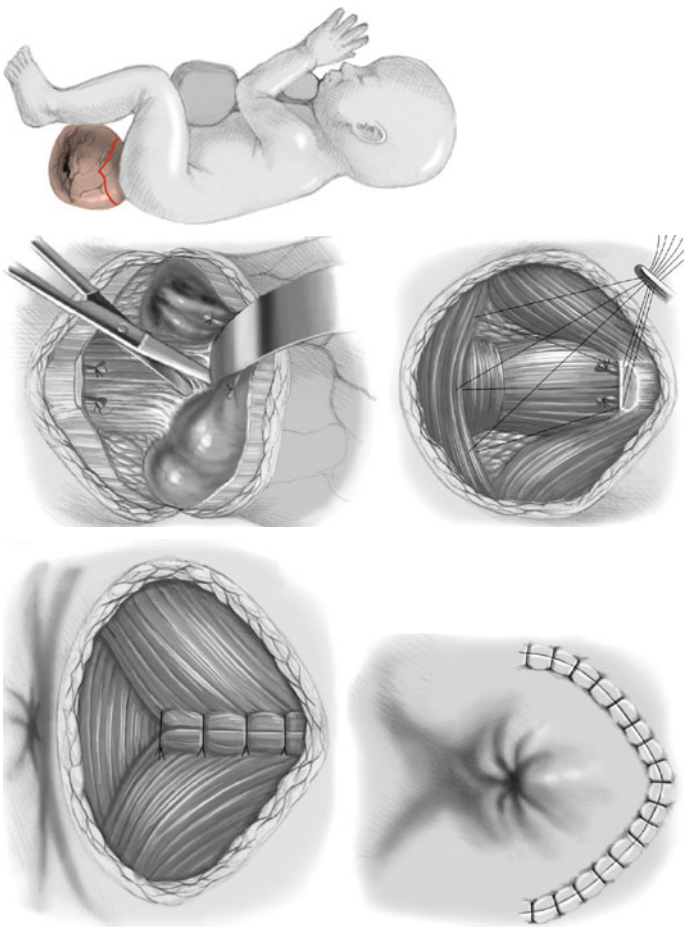


Fig. 28.3 Operative steps: sacrococcygeal teratoma

Complications

- Functional bowel problems
- Urinary incontinence or neurogenic bladder
- Recurrence if coccyx is not removed or there is malignancy at initial excision

Postoperative care

- Follow-up at monthly intervals for the first year, at 3-month intervals for at least 3 years, then annually including ultrasonography of the urinary tract
- Serial monitoring of α-fetoprotein to indicate recurrence of malignancy

Prognosis

- Excepting complications, see above, the outlook is good
- Recurrent tumors are approached surgically, however extensive disease may benefit from preoperative chemotherapy

28.7 Lung Tumors

General considerations

- In childhood the most common lung tumors are metastases of typical childhood tumors such as Wilms' tumor or neuroblastoma

Lung tumors

- **P**leuro**p**ulmonary **b**lastoma (PPB) is a rare and highly aggressive malignant tumor of the lung associated with **c**ongenital **c**ystic **a**denomatoid **m**alformation (CCAM). Metastases are usually found in the central nervous system
  - Arises from the lung, from the pleura or from both sites
  - There seems to be a constitutional and heritable predisposition to other dysplastic and neoplastic diseases (cystic nephroma, pulmonary cysts) in 25% of these patients
  - Bilateral PPB has been described
  - Poor prognosis

- Embryonal rhabdomyosarcoma is very rare: 35% arise from pre-existing pulmonary cystic malformations (CCAM)
- Bronchial carcinoid
- Bronchial mucoepidermoid tumors
- Inflammatory pseudo-tumor (synonyms: plasma cell granuloma, histiocytoma, fibrous xanthoma)
  - Suspected etiology: uncontrolled reaction to tissue damage or chronic inflammation
  - 50% of patients below the age of 16
  - 40% have no signs
  - Histology: spindle cells, myofibroblasts, granulomatous inflammation, lymphoid hyperplasia, fibrosis
  - Invasive growth (vascular invasion, bone destruction) recurrence and metastases can occur

Signs

- 40% have no specific signs
- Cough
- Fever
- Wheezing
- Fatigue
- Lower respiratory tract infection

Preoperative work-up

- Chest X-ray a.p. and profile
- CT scan
- MRI

Operation

- Needle biopsy may lead to an unclear diagnosis
- Surgical resection via thoracotomy or thoracoscopically (Fig. 28.4; see Chap. 16)

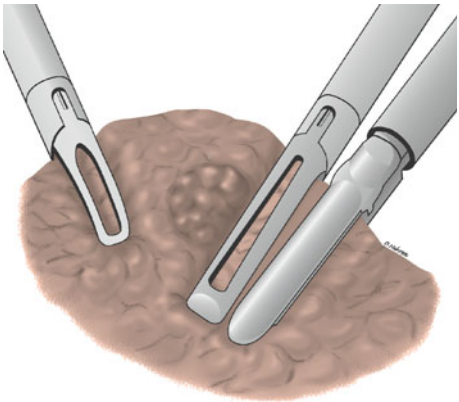


Fig. 28.4 Metastasis resection of a peripheral lung metastasis

28.8 Parasitic Lung Tumors

General considerations

- Vesicular tumor representing the development of *Echinococcus granulosus* in the lung
- Additional, hepatic localization is present in more than 50% of pulmonary cases
- A high incidence is revealed in 8- to 12-year-old children, but cases in children 4–5 years old have been described in endemic areas such as the Mediterranean shores of Europe
- The infestation occurs via the digestive tract (see Sect. 27.5)
- Aerial infestation is a controversial hypothesis (pulmonary hydatid cysts with no hepatic association)

Classification

- Noninfected hydatid cyst, as a perfectly circular, contoured formation
- Infected hydatid cyst, with the aspect of a pneumocyst

Signs

- In many cases there are no signs, and these cases are discovered incidentally
- Hemoptysis due to the rupture of small pericystic vessels
- General signs of hydatid intoxication associated with urticarial and dyspeptic disorders that may be present
- Thoracic pain, intercostal neuralgia due to pleural irritation
- Dyspnea is encountered in seldom cases of giant cysts or in multiple pulmonary cysts
- Nonproductive irritative cough
- Patients with complicated pulmonary hydatid cysts may develop shiver, fever, and dyspnea
- Hydatid cysts may rupture and the contents are eliminated by pseudovomiting with a high risk of aspiration
- If the content floods the pleura, it leads to pneumothorax

Preoperative work-up

- Blood tests demonstrating eosinophilia, not specific
- Cassoni test
- Weinberg test
- Chest X-ray
- CT
- Scintigraphy

Operation

- The only effective treatment is surgery, especially for large and complicated cysts
- The aim is to eliminate the cyst and obliterate the residual cavity
- Thoracotomy
- Isolation of the area of intervention with dressings
- Cyst puncture and 20% NaCl irrigation
- Elimination of the hydatid membrane
- Suture of open bronchi

- Drainage of the pleural cavity
- Preserving surgery is preferred in more than 90% of cases
- Radical surgery is considered in complicated cysts

Complications

- Residual cavity remains
- Bronchiectasis
- Recurrence

28.9 Mediastinal Tumors

General considerations

- Rare tumors in the mediastinum (see Table 28.10)

Table 28.10 Tumors in the mediastinum

Anatomical space	Contents	Lesions
Anterior mediastinum	Thymus	▪ Thymoma is a rare lesion in children occasionally associated with myasthenia gravis ▪ Thymomas need aggressive resection due to limited response to radio- and chemotherapy ▪ Teratomas with mixed cystic and solid components
Middle mediastinum	Heart Trachea/bronchi Great vessels Esophagus Lymphatics	▪ Masses are likely to be caused by the lymphoid tissue located around the lung hilus ▪ Non-Hodgkin lymphomas ▪ Hodgkin disease
Posterior mediastinum	Spine Lymphatics Sympathetic chain	▪ Masses are most commonly benign ganglioneuromas in older children and teenagers, frequently asymptomatic ▪ Benign neurofibromas (dumbbell tumor, Morbus Recklinghausen) ▪ Malignant neuroblastomas originating from the sympathetic chain

Signs

- Respiratory distress due to compression of the airways
- Cough, hoarseness (compression of the laryngeal nerve)
- Wheezing
- Recurrent respiratory infections
- Atelectasis
- Hemoptysis
- Dysphagia (compression of the esophagus)

Preoperative work-up

- Chest X-ray
- CT (to detect calcifications)
- MRI (spinal involvement)
- CT-guided puncture of the mass (histology)
- Bronchoscopy with transbronchial needle aspiration (cytology)
- Thoracoscopy as a minimally invasive procedure or thoracotomy to enable larger biopsy samples to be taken
- Tumor markers: α-fetoprotein and β-human chorionic gonadotrophin (β-hCG) (teratomas)
- Bone marrow puncture in lymphoma cases

Surgical therapy

- Biopsy for diagnostic reasons
- Resection

Operation

- During induction of anesthesia ventilation problems may occur (risk of respiratory collapse in relation to the size of the mediastinal mass)
- Median sternotomy for lesions in the anterior mediastinum
- Lateral thoracotomy to reach the middle and posterior mediastinum
- Pay attention to the phrenic nerve, the vagus nerve, and the recurrent laryngeal nerve
- Minimally invasive resections, such as biopsy or complete resection of thymoma, lymphatic nodes, and neurogenic tumors, are feasible

Postoperative care

- Mediastinal tube drainage

Prognosis

- Depends on the stage of the disease

28.10 Malignant Liver Tumors

General considerations

- The two main liver tumors in childhood are hepatoblastoma and hepatocellular carcinoma
- Hepatoblastoma
  - Highly malignant
  - Mostly in children under 5 years old
- Hepatocellular carcinoma
  - Less frequent than hepatoblastoma
  - Most often associated with an underlying liver disease (inborn error of metabolism, liver cirrhosis of any origin)
  - Most often in children more than 5 years old

Signs

- Palpable mass
- Increased abdominal volume
- Appetite loss, poor general condition
- Pain due to capsule distension

Classification

- The most useful classification defined in the SIOPEL studies is PRETEXT (PRE-Treatment EXTent of disease system; Table 28.11), which allows the surgeon to predict the surgical resectability at presentation and before the operation

Table 28.11 PRETEXT classification of malignant liver tumors

Stage	Liver sectors involved	Adjoining free liver sectors	Resectability
I	1	3	Left lobectomy, left or right hemihepatectomy
II	1 or 2	2	Left or right hemihepatectomy
III	2 or 3	Not 2	Extended left or right hemihepatectomy
IV	4		Unresectable (liver transplantation)

Preoperative work-up

- Serum α-fetoprotein is nearly always elevated, with a higher value in hepatoblastoma than in hepatocellular carcinoma
- Thrombocytosis in hepatoblastoma
- Doppler ultrasonography
- CT/MRI/PetScan
- Angiography is rarely needed and is to be avoided in small children

Therapy principles

- Hepatoblastoma is highly sensitive to chemotherapy (cisplatin, carboplatin, adriamycin)
- Hepatocellular carcinoma is much less chemosensitive
- Surgical resection should always be preceded by chemotherapy in order to reduce the volume of the mass and to make it less friable during surgical resection. The decreased volume also enables better anatomical definition of the tumor's location, thereby allowing for a more differentiated resection
- In both cases, complete surgical resection is a prerequisite for cure

Postoperative care

- Depends on the extent of the liver resection
- Intensive care

- In cases of extensive resection, hyperbilirubinemia occurs but will disappear spontaneously as soon as the liver regenerates
- Supplement with albumin and vitamin K

Prognosis

- With modern techniques in the hands of experienced surgeons the operation's mortality should be minimal
- The main surgical complication is bleeding
- Hepatoblastoma: with combined chemotherapy and radical surgical resection, a disease-free 5- to 10-year survival of around 70% should be obtained, depending on the stage of the tumor at presentation and at the time of surgery, the response to chemotherapy, and the completeness of surgical resection
- Hepatocellular carcinoma: poorer prognosis with a survival rate at 5 years of around 30%, despite chemotherapy

Anatomy of the liver

- Liver anatomy is illustrated in Fig. 28.5

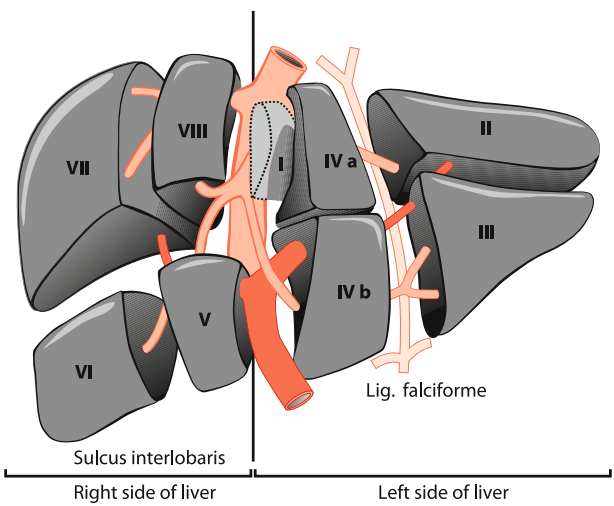


Fig. 28.5 Liver anatomy

Liver resections

- The techniques of liver resection are illustrated in Fig. 28.6 and detailed in Table 28.12

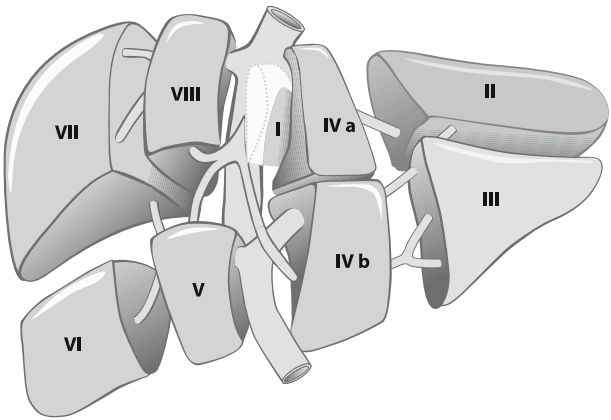


Fig. 28.6 Liver resections. (*II* Segmentectomy, *V* + *VI* + *VII* + *VIII* right hemihepatectomy, *I* + *IVb* + *V* + *VI* + *VII* + *VIII* extended right hemihepatectomy)

Table 28.12 Liver resection

Resection	Segments
Right hemihepatectomy	V, VI, VII, VIII
Extended right hemihepatectomy	I, IVb, V, VI, VII, VIII
Left lateral segmentectomy	II, III
Left hemihepatectomy	I, II, III, IVa,b
Left extended hepatectomy	I, II, III, IVa,b, V, VIII

Surgical technique

- Make a bilateral subcostal incision; in older children, extend the upper midline to the xiphoid
- Mobilize the liver completely with division of the round ligament and the right or the left triangular ligament
- Isolate the inferior vena cava, below and above the liver
- Encircle the liver pedicle (should a Pringle maneuver be required to control bleeding)
- Total vascular occlusion is rarely needed in children

Operation: right hemihepatectomy

- The operative steps are illustrated in Fig. 28.7
- Isolate and divide the right hepatic duct, the right hepatic artery and the right portal vein
- Isolate and divide the extrahepatic portion of the right hepatic vein
- Mobilize the liver to the left and divide the accessory hepatic veins between the right lobe, the right portion of segment I, and the vena cava
- Incise the liver capsule with cautery on the right side of the median hepatic vein (localized by intraoperative echography); the median hepatic vein can be localized by a line drawn from the left aspect of the suprahepatic vena cava to the fundus of the gallbladder
- Divide the liver parenchyma. A Kelly fracture can be used in combination with Cusa. The fine structures can be coagulated; biliary structures and major crossing vessels should be suture-ligated
- The rough surface of the liver should be dry. Hemostasis can be completed with an argon-beam coagulator and biological glue
- Suspend the remnant left hemi-liver in order to avoid postoperative twist causing hepatic vein obstruction
- Drain the right subphrenic space

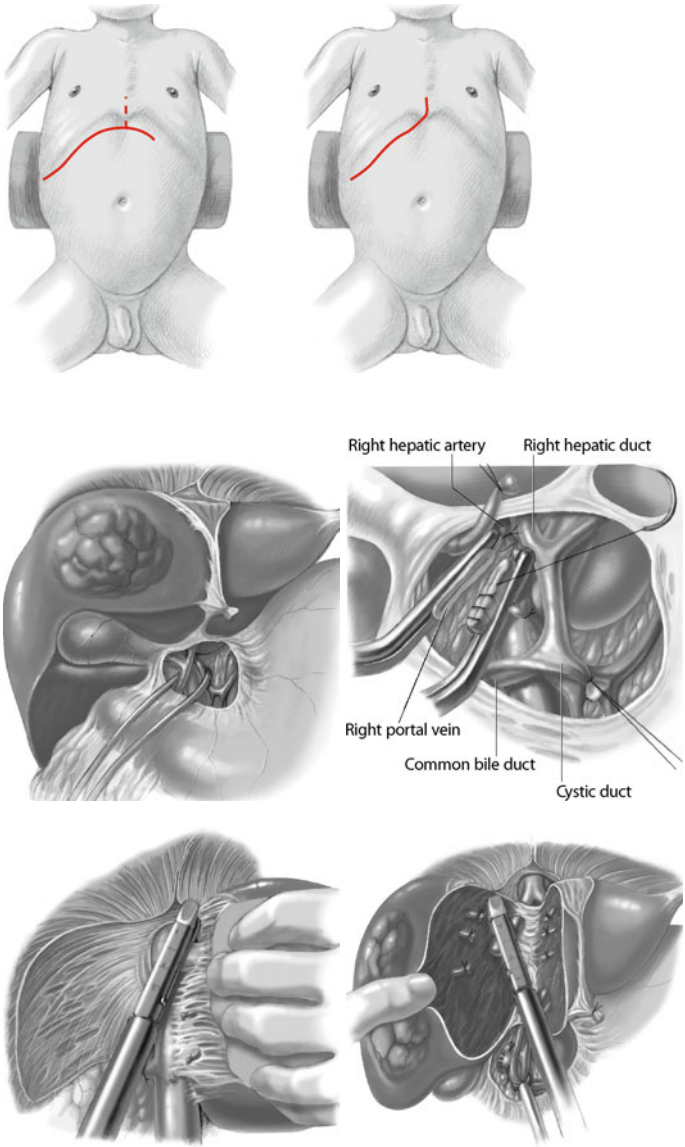


Fig. 28.7 Right/extended hemihepatectomy

Operation: extended right hepatectomy (right trisegmentectomy)

- Divide the right hepatic duct, the right hepatic artery, and the right portal vein
- Divide the branches of the left portal pedicle, branching to segment IV, located on the right side of the umbilical scissure
- The right hepatic vein should be divided as for a right hemihepatectomy; the median hepatic vein is divided transparenchymally

Operation: left lateral segmentectomy (left lobectomy)

- Resection of segments II and III
- The left hepatic duct, the left hepatic artery, and the left portal vein should be divided on the left side of the umbilical scissure, preserving the elements from the left pedicle branching for segment IV
- Isolate and divide the left hepatic vein

Operation: left hemihepatectomy

- Resect the whole right hemi-liver with segment IV
- Division of the right hepatic duct, the right hepatic artery and the right portal vein
- Division of the branches of the left portal pedicle, branching to segment IV, located on the right side of the umbilical scissure
- The right hepatic vein should be divided as for a right hepatectomy, the median hepatic vein is divided transperenchymally
- Incise the liver capsule on the upper surface of the liver on the right side of the falciform ligament and of the umbilical scissure

Operation: extended left hepatectomy (left trisegmentectomy)

- Resect the full left hemi-liver with the anterior sector of the right liver. Only the posterior sector (segments VI and VII) of the right hemi-liver (behind the right hepatic vein) should be preserved
- This is the most difficult type of liver resection
- The left liver pedicle should be divided

- Divide the portal vein, the hepatic artery, and the bile duct of the anterior sector of the right lobe of the liver via a trans-hilar or a supra-hilar approach
- The parenchymal transection plane lies in a frontal position, in front of the right hepatic vein

28.11 Nephroblastoma

General considerations

- Nephroblastoma, also called Wilms' tumor, is the most frequent intra-abdominal solid tumor in childhood
- It is an embryonal neoplasm originating from metanephric blastema and histologically has three components, which are
  - Blastemal
  - Epithelial
  - Stromal
- The majority of European patients have been or are included in multicenter trials, mainly the SIOP Nephroblastoma Trial and Study, and all receive the following
  - two-drug preoperative chemotherapy for 4 weeks after an imaging-based diagnosis in children older than 6 months
  - Surgery
  - Primary nephrectomy is recommended only in infants <6 months of age and in emergencies
- In contrast, the NWTSG (National Wilms' Tumor Study) and currently COG (Children's Oncology Group) recommend primary surgery in all cases, except those where the nephroblastoma is considered large and clearly not resectable
- The discussion between representatives of both studies is prolonged and has not yet been concluded
- This chapter is oriented according to the SIOP recommendations

Epidemiology

- 90% of cases affect children before the 8th year of life with a peak at 3–4 years
- On average, nephroblastoma affects 7 per 1,000,000 children per year. This rate varies in different populations: in the Chinese the incidence is as low as 2.5 per 1,000,000; in black Americans the incidence is high, at 10.9 per 1,000,000
- It is slightly more frequent in girls than in boys

Etiology and genetics

- The two-hit Knudson's pathway (1972) seems partly confirmed, however the etiology is probably more complex than this
- 90% occur in otherwise healthy children. In 10% of cases there are associated malformations [overgrowth syndromes as Beckwith–Wiedemann, isolated hemi-hypertrophy, Perlman, Sotos, Simpson–Golabi–Behemel, or non-overgrowth syndromes such as aniridia, WAGR (which stands for **W**ilms' tumour with **a**niridia, **g**enital abnormalities and mental **r**etardation), Denys–Drash and Bloom]. Also hypospadias, cryptorchidism, and renal fusion occur at an increased rate in these patients
- Genetic alterations in nephroblastoma concern 11p13 (gene WT1), 11p15 (gene WT2), changes in 11p and, interestingly, LOH of 16q and 1p36 – suggesting poor outcome but not related to anaplasia

Milestones of research

- Milestones of research are noted in Table 28.13

Table 28.13 Milestones of research into nephroblastoma

1814	T.F. Rance	First descriptions
1879	Osler	
1894	Döderlein and Birch-Hirschfeld	
1899	Max Wilms	
1900	First successes after surgical treatment	
1915	Friedlander, Heinmann – first application of radiotherapy	
1930s	Ladd and Gross – modification of surgical technique	
1950s	Farber – introduction of chemotherapy	
1969	First large multicenter study: National Wilms' Tumor Study 1	
1971	First international multicentre study SIOP-1 International Society of Pediatric Oncology Study	

Classification

- The revised SIOP working classification of renal tumors of childhood (2001) is shown in Table 28.14

Table 28.14 Revised SIOP working classification of renal tumors of childhood (2001)

For pre-treated cases		
1	Low risk	▪ Mesoblastic nephroma[a] ▪ Cystic partially differentiated nephroblastoma ▪ Completely necrotic nephroblastoma
2	Intermediate risk	▪ Nephroblastoma – Epithelial type – Stromal type ▪ Mixed type ▪ Regressive type ▪ Focal anaplasia
3	High risk	▪ Nephroblastoma – Blastemal type – Diffuse anaplasia ▪ Clear cell sarcoma of the kidney[a] ▪ Rhabdoid tumor of the kidney[a]

[a] These forms are not variants of nephroblastoma but are other childhood tumors of the kidney.

Table 28.14 *(continued)* Revised SIOP working classification of renal tumors of childhood (2001)

For primary nephrectomy cases		
1	Low risk	▪ Mesoblastic nephroma[a] ▪ Cystic partially differentiated nephroblastoma
2	Intermediate risk	▪ Non anaplastic nephroblastoma and its variants ▪ Nephroblastoma – focal anaplasia
3	High risk	▪ Nephroblastoma – diffuse anaplasia ▪ Clear cell sarcoma of the kidney[a] ▪ Rhabdoid tumor of the kidney[a]

[a] These forms are not variants of nephroblastoma but are other childhood tumors of the kidney.

Stages according to SIOP classification

- **Stage I**
  - The tumor is limited to the kidney or surrounded by a fibrous pseudocapsule if outside the normal contours of the kidney
  - The renal capsule or pseudocapsule may be infiltrated by tumor but the tumor does not reach the outer surface and it can be completely resected (resection margins "clear")
  - The tumor may be protruding ("bulging") into the pelvic system and "dipping" into the ureters (but it does not infiltrate their walls)
  - The vessels of the renal sinus are not involved
  - Intrarenal vessel involvement may be present
  - The presence of a necrotic tumor or chemotherapy-induced changes in the renal sinus and/or outside of the kidney should not be regarded as a reason for upgrading the stage of the tumor
- **Stage II**
  - The tumor extends beyond the kidney or penetrates through the renal capsule and/or fibrous pseudocapsule into perirenal fat but can be completely resected (resection margins "clear")
  - Tumor infiltrates the renal sinus and/or invades blood and lymphatic vessels outside the renal parenchyma but it can be completely resected

- Tumor infiltrates adjacent organs or vena cava but can be completely resected

- **Stage III**
  - Incomplete excision of the tumor, which extends beyond resection margins (gross or microscopical tumor remnants)
  - Any abdominal lymph nodes involved
  - Tumor ruptures before the operation or intraoperatively (irrespective of other criteria for staging)
  - Tumor is found on, or penetrates, the peritoneal surface
  - Tumor thrombi present at resection margins of vessels or ureters, it is transected or removed piecemeal by the surgeon
  - Tumor has been surgically biopsied (wedge biopsy) prior to preoperative chemotherapy or surgery
  - The presence of necrotic tumors or chemotherapy-induced changes in a lymph node or at the resection margins is regarded as proof of a previous tumor and therefore the tumor is assigned to stage III
- **Stage IV**
  - Hematogeneous metastases (lung, liver, bone, brain, etc.) or lymph node metastases outside the abdomino-pelvic region
- **Stage V**
  - Bilateral renal tumors at diagnosis. Each side should be substaged according to the above classifications

Signs

- The signs of nephroblastoma are listed in Table 28.15

Table 28.15 Signs of nephroblastoma

Classical	Painless mass in the upper abdomen
Unstable	▪ Hematuria ▪ Varicocele ▪ Anemia or erythrocytosis ▪ Abdominal pain in cases of extensive mass or rupture
Accidental	Small asymptomatic tumor discovered upon imaging carried out for other reasons or at screening for the predisposing syndromes

Differential diagnosis

- Other intra-abdominal (mainly retroperitoneal) tumors and lesions
  - Adrenal or paravertebral neuroblastoma
  - Adrenocortical carcinoma
  - Lymphoma of the kidney (rare)
  - Metastases in the para-aortic lymph nodes at the level of renal vessels (germinal tumors of gonads)
  - Nephroblastomatosis
  - Teratomas
  - Benign tumors of kidney, i.e., cystic lesions, hydronephrosis, xanthogranulomatous nephritis

Preoperative work-up

- All imaging results at presentation and after chemotherapy should be carefully assessed prior to an operation
- Three-step practical guide to diagnosis (according to the SIOP 2001 protocol) is given in Table 28.16

Table 28.16 Three-step practical guide to diagnosis of nephroblastoma (according to the SIOP 2001 protocol)

Stage	Investigation	Reasons
1	24-h urine collection whilst on a standard diet	▪ Exclude neuroblastoma ▪ Evaluate renal function
	Ultrasound	▪ Make three-dimensional measurements ▪ Establish if this is an intrarenal process ▪ Investigate the opposite kidney ▪ Is the tumor cystic, solid or both? ▪ Search for other abnormalities in the abdomen, intravascular extension, etc. ▪ Exclude hepatic metastases
	Plain chest X-ray a.p.	▪ Exclude pulmonary metastases

Table 28.16 *(continued)* Three-step practical guide to diagnosis of nephroblastoma (according to the SIOP 2001 protocol)

Stage	Investigation	Reasons
2	Abdominal CT scan	▪ Confirm intrarenal lesion with certain characteristics, relations to other structures ▪ Lymph nodes, invasion of vessels, other organs and structures ▪ Also useful for measurements
	Thoracic CT scan if any doubt on chest X-ray	Exclude or confirm metastases
3	Needle biopsy (see below)	Verify the tumor by histology

Needle biopsy recommendations

- **Indication**
  - Unusual clinical presentation
    - Age >5–6 years
    - Urinary infection
    - Septicemia
    - Psoas inflammation
  - Unusual imaging findings
    - Calcification
    - Voluminous adenopathies
    - Renal parenchyma not visible
    - Almost totally extrarenal process
- **Contraindications**
  - Age <6 months
  - Suspicion of rupture or hemorrhage
  - Needle biopsy is unlikely to be of benefit in pure cystic structures with no solid component. Immediate surgery to establish diagnosis is recommended in such cases

- **Procedure**
  - Test blood coagulation
  - Perform under general anesthesia
  - Biopsy from posterior
  - Ultrasound-guided biopsy preferred
  - Tissue handling by experienced pathologist
  - Discuss the coaxial needle technique with local radiologist
  - Use fine needle or thin true-cut biopsy needles

Operation strategy

- The principles of nephrectomy for pediatric malignancy were published by Gross in 1953: a wide transverse transabdominal incision and transperitoneal approach with early ligation of the renal vessels
- Preoperative chemotherapy as demonstrated by SIOP studies 2, 5, 6, 9 and 93-01 makes nephrectomy easier and less hazardous. Furthermore, metastases may disappear or become resectable, and the vascular extent may regress. This approach is recommended by SIOP Nephroblastoma Trial and Study Committee
- During the operation an oncologist must be available
- An emergency operation may be necessary, however it is usually possible to follow most of the recommendations
- Immediate surgery indications
  - If the tumor ruptures and bleeds before the planned time of operation and conservative management is ineffective
- Postoperative treatment is risk-adopted and depends on both the variant of pathology and the stage established at surgery

Operation: nephrectomy

- The operative steps are illustrated in Fig. 28.8
- Make a long transverse transabdominal incision, extended to the side of the tumor
- Meticulously inspect the abdomen prior to tumor resection looking for
  - Metastases in the liver
  - Lymph nodes (see below)

- Sample lymph nodes and examine histologically; this is imperative for accurate staging and subsequent treatment, even if tissue is not macroscopically suspicious
  - Hilar and para-aortic lymph nodes at the origin of the renal artery (regional nodes)
  - Extraregional nodes in the mesentery, the contralateral renal hilum, the aortic bifurcation and celiac trunk, the splenic hilum, and peritoneum if enlarged or suspicious
  - Radical lymph node dissection does not enhance survival and therefore is not part of surgical therapy
- Every suspicious lesion should be excised (if resectable) or biopsied (if unresectable)
- An extensive Kocher maneuver of the duodenum is a convenient approach to the renal vessels for a large tumor, whether on the right or left. An approach via the peritoneum lateral to the colon is also acceptable
- Ligate the renal vessels early. The renal artery should be ligated first, in order to avoid swelling of the tumor which increases of its fragility and leads to the possibility of dissemination via perforating perinephric veins
- The tumor should be removed along with the adipose capsule and, if possible, with all invaded surrounding structures
- Resect the ureter as close to the bladder as possible
- Leave the adrenal gland in situ if a safe resection margin between the tumor and the gland can be guaranteed
- Heroic and mutilating resections such as pancreatectomy are not recommended as these tumors are both chemo- and radio-sensitive

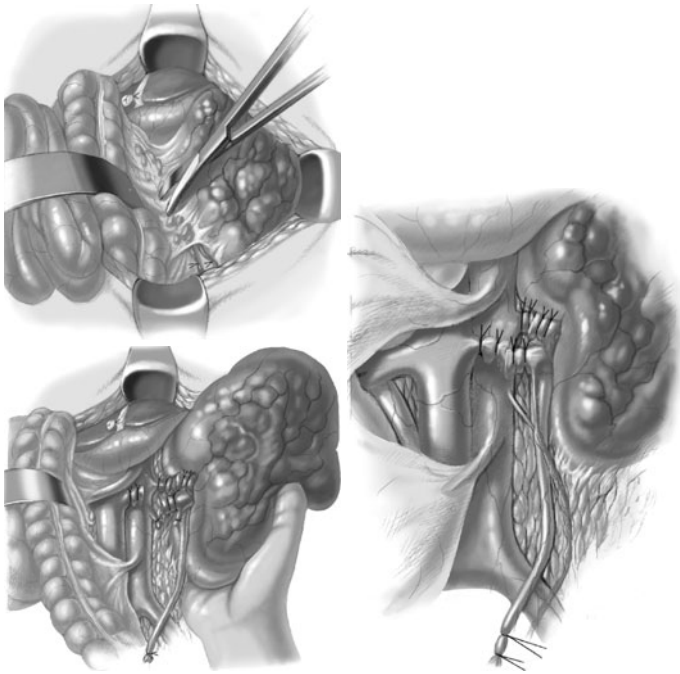


Fig. 28.8 Operative steps: nephrectomy

Operation: cava thrombus

- Although the intravascular extent of the tumor is usually apparent at preoperative imaging, the vena cava and renal vein should be carefully examined during the operation
- If a thrombus is found, it should be removed (Fig. 28.9)
  - A short thrombus in the renal vein may be resected together with the vein
  - A thrombus extending to the infra-hepatic vena cava should be removed through a cavotomy, after occluding the contralateral renal vein and cava above and below the thrombus. The venotomy is then closed either directly or with a patch

- A longer thrombus (intra-hepatic, supra-hepatic, or right atrial) may require the assistance of a vascular or cardiac surgeon and cardio-pulmonary by-pass

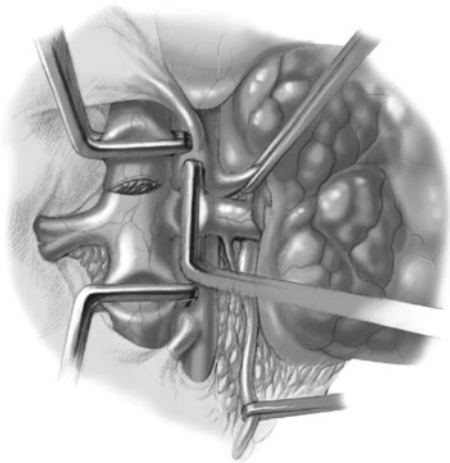


Fig. 28.9 Vena cava thrombus extraction

Operation: metastases resection (Stage IV patients)

- Lung metastases should be excised as soon after nephrectomy as the patient's condition permits, usually within 14 days. Bilateral resectable lung metastases should be excised via either two thoracotomies or one sternotomy, depending on surgical choice and anatomy. Wedge resections can frequently be radical. If wedge resection cannot achieve complete excision then segmentectomy or lobectomy is indicated, however pneumonectomy is never justified
- A similar approach for extrapulmonary metastases is recommended, especially for the second most frequent site, namely the liver
- Metastases outside the lung or liver should be excised completely, provided the operation can be done without mutilation or loss of vital organs

- It is not recommended to operate on metastases that have progressed during preoperative chemotherapy, as complete excision is rarely successful. Alternative chemotherapy and/or radiotherapy should be considered first

Operation: bilateral disease

- Bilateral cases should be treated individually by a highly experienced team
- Surgery is scheduled after tumor reduction with chemotherapy
- The aim is a bilateral partial nephrectomy, preferably in two separate operations performed 1–2 weeks apart, starting with the less involved kidney. The partial resection should be performed either in situ or extracorporally with subsequent autotransplantation
- Complete nephrectomy on one side with partial nephrectomy on the opposite side is acceptable, providing enough functional renal tissue can be preserved
- The options for radiotherapy as a local treatment are limited after partial nephrectomy. But there are examples indicating that low-dose radiotherapy (10 Gy) and chemotherapy may result in long-term remission even after incomplete excision
- If bilateral nephrectomy is performed, transplantation should be planned preferably after 2 years of disease-free survival
- In cases of accidental diagnosis of bilateral tumors during operation in a previously untreated patient, biopsies should be taken from both tumors, preferably with the True-cut needle, and the patient treated with chemotherapy. If the lesion is very small, the biopsy should be excisional

Operation: partial nephrectomy

- Partial nephrectomy may also ensure local control in an unilateral Wilms' tumor, but the advantages and risks have to be precisely evaluated in each individual case
- Contralateral urological and nephrological disorders and genetic syndromes with an increased risk of a Wilms' tumor compared with the risk of a hyperperfusion nephropathy in the remaining kidney are important criteria when considering this option

- We do not recommend partial nephrectomy in a classical unilateral nephroblastoma that is not related with the above disorders
- The probability of curing the patient after recurrence is usually not higher than 40%

Postoperative treatment

Postoperative treatment as recommended by SIOP (2001) is given in Table 28.17.

Table 28.17 Risk-related postoperative therapy (SIOP 2001)

Risk	Stage	Treatment (postoperative)
Low risk	I	No further treatment
	II, III	Two-drug chemotherapy
Intermediate risk	I[a]	Two-drug chemotherapy
	II[b], III[b]	Two-drug vs. three-drug (randomly assigned) chemotherapy and radiotherapy for stages III
High risk	I	Three-drug chemotherapy
	II, III	High-risk four-drug chemotherapy and radiotherapy

[a] GPOH protocol (German participants of the study) treats patients at intermediate risk (excluding epithelial and stromal predominant) and a preoperative tumor volume >500 ml as stage II with DOX.

[b] GPOH protocol treats patients at intermediate risk (excluding epithelial and stromal predominant) and a preoperative tumor volume >500 ml according to the high-risk protocol.

Prognosis

- Over 85% of patients are long-term survivors
- Patients with Stage I disease are nearly all curable
- Prognosis for those with higher stage disease is strongly dependent on the pathology variant
  - Those with diffuse anaplasia and blastemal predominance persisting despite pre-treatment are at high risk
- Stage IV and V patients have encouraging survival rates and should always be treated with curative intention

- Patients who relapse still have a chance of being cured, especially if relapses occur at late follow-up and the primary treatment involved the two-drug regimen only, with no radiotherapy
- LOH 16q1p seems to be a newly discovered risk factor

28.12 Ovarian Tumors

General considerations

- The ovaries present a variety of tumors due to hormonal influence or congenital malformations
- Most ovarian tumors are benign, especially the cystic ones

Classification

- Germinal tumors in children can be classified according to the TNM–SFOP system (Table 28.18)

Table 28.18 Germinal tumor classification in children according to the TNM-SFOP system

Stage	Description
I	Tumor <5 cm confined to the organ of origin
II	Tumor >5 cm
IIIA	Regional lymph node involvement regardless of tumor size
IIIB	Local progression, peritoneal extension (e.g., implants) and/or malignant cells in the peritoneal liquid, regardless of tumor size
IV	Any tumor, any regional lymph nodes, presence of distant metastases

Preoperative work-up

- α-Fetoprotein (AFP) level
- β-hCG level
- CA 125
- Ultrasonography
- CT
- Chest X-ray

Types of ovarian tumor

- **Immature teratoma**
  - It is doubtful whether foci of clearly malignant tissue exist (e.g., embryonal carcinoma, yolk sac tumor)
  - The measurement of AFP and β-hCG is mandatory prior to surgery, after resection and at follow-up
  - Surgical treatment is usually sufficient, but oncological evaluation and further follow-up are mandatory
  - Chemotherapy has its place in non-resectable tumors
- **Mature teratoma**
  - Require surgical excision (unilateral adnexectomy), which can also be performed laparoscopically
  - In cases of unresectable disease, chemotherapy is rarely effective. However, if implants are mature, usually they do not progress and may only require observation
- **Yolk sac tumor and embryonal carcinoma**
  - Require surgical resection only in stage I disease (= primarily completely resectable and AFP levels are <15,000 $ng \cdot ml^{-1}$)
  - If staged over I and/or AFP >15,000 $ng \cdot ml^{-1}$ chemotherapy is also required
  - Operation is a unilateral adnexectomy with a small biopsy taken from the contralateral ovary
  - In cases of high-stage disease, secondary surgery is performed when tumor markers are normalized; the aim is to resect the residual mass
- **Dysgerminoma**
  - Female phenotype and XY karyotype may predispose to the development of dysgerminoma
  - As the gonads in these patients are not functioning, bilateral adnexectomy should be considered since it is not mutilating
  - After complete resection of stage I disease, complementary, moderate-dose (approx. 20 Gy) radiotherapy to the inguino-lumbo-aortic and supraclavicular lymph nodes should be considered

- Those with higher stages of disease should receive chemotherapy with/without radiotherapy
- Bilateral adnexectomy is only carried out in cases of dysgerminoma "in situ," supplemented with chemotherapy if dysgerminoma penetrates the gonadal capsule

- **Folliculoma**
  - The treatment should be based upon surgery
  - The role of chemotherapy is uncertain, however it is recommended in higher stages of disease and incompletely resected tumors
- **Ovarian carcinoma**
  - Not a typical pediatric tumor, however it should be taken into account especially in adolescents
  - CA 125 is its serum marker
  - The adult gyneco-oncology rules should be applied
  - In general, primary surgery is important if complete resection is achieved. However, patients with advanced stages are not counted as lost if they respond to chemotherapy, which can be either a complementary treatment or both neo-adjuvant and complementary. In such cases secondary surgery should be considered

Prognosis

- Prognosis is favorable in approx. 70% of patients

28.13 Testicular Tumors

General considerations

- Pathologically similar of ovarian tumors
- Teratomas, yolk sac tumors, and embryonal carcinomas are typical non-seminomatous germinal testicular tumors
- Seminoma is not a pediatric malignancy; adult oncology rules should be applied upon its occasional diagnosis in children. In general, surgery, radiotherapy, and chemotherapy may be useful

- Similar diagnostic work-up as for ovarian tumors should be implemented
- Consider whether the condition is an emergency torsion of a healthy or diseased gonad or its appendices, epididymitis, or a simple hydrocele
- If sonography clearly excludes testicular tumor and clearly describes the origin of the disease, diagnostic work-up should not include radiological examinations or marker assessment
- In cases of testicular tumor, regional lymph nodes should also be assessed. The first station nodes are at the level of the renal vessels, which may have been altered by previous scrotal surgery so that inguinal nodes may also be first station nodes

Operation

- The operative steps are illustrated in Fig. 28.10
- The approach is inguinal; the resection is always an orchidectomy with high ligation of the spermatic cord
- Lymph nodes should not be dissected primarily
- If a scrotal approach has already been performed, hemiscrotectomy should follow confirmation of the malignant character of the testicular tumor
- In cases of a high-stage tumor, secondary surgery is performed after tumor markers have been normalized, with the aim of resecting the residual mass
- In non-secreting tumors, second-look surgery aims either to resect the residual mass or to verify the remission

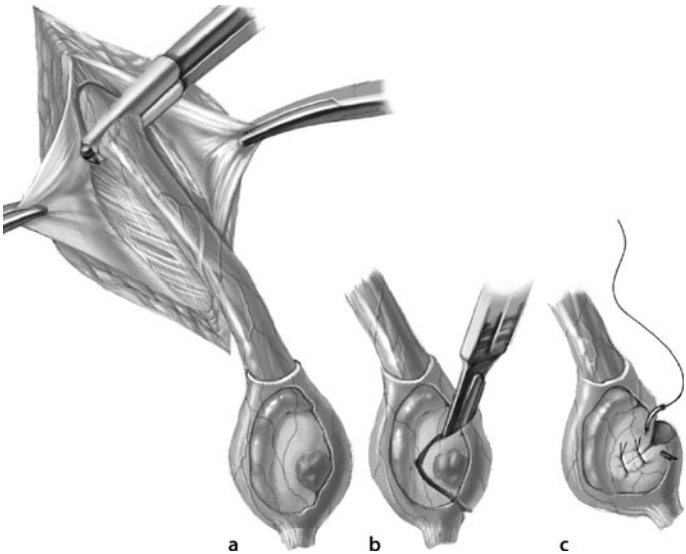


Fig. 28.10a–c Operative steps: testicular tumor resection

28.14 Soft Tissue Sarcomas

General considerations

- Undifferentiated mesenchymatous cells (muscles, connective tissue, fat, vessels)
- Morbidity is 8.4 per 1,000,000 live births
- 6% of the malignant tumors are in children
- The most frequent tumor in children is rhabdomyosarcoma
- An ill-defined lump should always raise suspicion of a soft tissue tumor with infiltrative growth
- Non-capsulated tumor, but, at times, with a pseudocapsule

Classification

- Soft tissue sarcoma classification is given in Table 28.19

Table 28.19 Soft tissue sarcoma classification

Origin	Benign	Malignant
Striated muscle	Rhabdomyoma	Rhabdomyosarcoma
Smooth muscle	Leiomyoma	Leiomyosarcoma
Connective tissue	Fibroma Fibromatoses Desmoid	Fibrosarcoma Malignant mesenchymoma
Synovial tissue	Synovioma	Synovialsarcoma
Histiocyts	Histiocytofibroma	Malignant histiocytoma
Fat	Lipoma/lipoblastoma	Liposarcoma
Blood vessels	Hemangioma	Angiosarcoma Hemangiopericytoma Juvenile angiofibroma
Lymphatic vessels	Lymphangioma	Lymphangiosarcoma

28.15 Rhabdomyosarcoma

General considerations

- Originates in the myoblasts and can appear at any site in the body
- It is one of the more frequently found solid tumors of childhood, with an incidence of between 8% and 13% of all solid tumors
- Metastases appear mainly in the lung, lymph nodes, bones, and liver
- Pediatric surgeons contribute by taking biopsy samples (either fine needle aspiration cytology or surgical biopsy) that enable the staging of disease and thus determination of further treatment, which is multidisciplinary

Classification

- Classification of rhabdomyosarcoma is given in Table 28.20, and its staging in Table 28.21

Table 28.20 Classification of rhabdomyosarcoma

Histology	Localization
Embryonal rhabdomyosarcoma (mostly botryoid variety)	Head, orbit, nasopharynx, middle-ear Neck Retroperitoneum Genito-urinary system
Alveolar rhabdomyosarcoma	Mainly in extremities
Pleomorphic rhabdomyosarcoma (extremely rare in children)	Skeletal muscles
Mixed rhabdomyosarcoma	See above

Table 28.21 Intergrouped rhabdomyosarcoma study's stages

I	Localized tumor Resection without microscopic residue
II	Invasion of neighboring structures, regional lymph nodes involved or microscopic residue
III	Incomplete resection or macroscopic residue
IV	Metastases

Signs

- Palpable tumor
- Local pain
- Polypoid mass in vagina or aural canal
- Ptosis and exophthalmos
- Hematuria
- Dysuria

Preoperative work-up

The following tests help to determine the stage correctly:

- Clinical examination
- Fine needle biopsy and/or surgical biopsy
- X-ray of the involved area and of the chest (p.a., profile)
- Ultrasonography

- Genitourinary system i.v. pyelogram
- Cystoscopy
- Liver and bone scintigraphy
- MRI and CT scan, in areas of difficult access or when lymph nodes are involved
- Blood count, C-reactive protein, liver function tests, creatinine, bone marrow aspirate
- Urine analyses
- Pre-chemotherapy for all tumors not manageable by excisional biopsy (with adequate safety margins)

Operation strategy

- The operative strategies are given in Table 28.22

Table 28.22 Operative strategy for rhabdomyosarcoma

Stage	Preoperative chemotherapy	Operation	Chemotherapy	Radiotherapy
I		+		
II		+	+	+
III	+	+ Second look	+	+
IV	+	+	+	+

Operation: biopsy

- Primarily it is necessary to obtain enough material for differential diagnoses and to define the histology
- Start with fine needle biopsy and then, if negative or doubtful, use incision or excisional biopsy (depending on size and location)
- Eventual biopsy of regional lymph nodes
- Eventual biopsy of metastases

Operation

- The excision must be wide
- If the localization is in the extremities or in the urogenital system, regional lymph nodes should also be removed (or at least sampled)
- If limits of excision prove positive, re-excision should be performed, especially on the limbs

Postoperative care

- Chemotherapy
- Radiotherapy

Prognosis

- Prognoses will depend on staging and histology
- Overall relapse-free cure is 60% at 3 years. Eventually, amputation will be required
- Particularly favorable in vaginal botryoides (80%–90%)

28.16 Ewing's Sarcoma

General information

- Tumor of mesenchymal tissues of the bone marrow
- It is the second most frequent bone tumor in children
- More frequent sites are pelvis and diaphyses of long bones
- Differential diagnoses: osteomyelitis, metastases, histiocytosis X, leukemia

Classification

- Classification is given in Table 28.23

Table 28.23 Classification of Ewing's sarcoma

I	Intra-osseous unilocular tumor
II	Extra-osseous unilocular tumor
III	Multilocular tumor (limited to the affected bone)
IV	Metastatic

Signs

- Local pain at the site of swelling
- Fever and pseudo-inflammatory signs
- Pathological fractures (late sign)
- Non-specific tumor signs

Preoperative work-up

- Blood count, electrolytes, Ca, P, alkaline phosphatase, lactate dehydrogenase (LDH), urine
- Fine needle biopsy or surgical biopsy
- Local X-ray (showing osteolyses and sub-periosteal reaction – "onion like")
- Chest X-ray
- Bone scintigraphy
- MRI
- CT scan
- Angiography

Operation

- Surgery, non-mutilating, whenever possible, with radical excision
- Surgery of localized metastases (namely lung), as for osteosarcoma

Postoperative care

- Polychemotherapy
- Radiotherapy

Prognosis

- Depends on staging
- Survival after 5 years 50%–75%

28.17 Osteosarcoma

General information

- Neoplasm of mesenchymal cells that form bone and osteoid
- It is the most frequent malignant bone tumor in children
- Localized mainly close to the knee joint (mostly lower femur), sometimes in the upper humerus as well
- Appears mainly after the age of 10 years

Signs

- Persistent and slowly progressing local pain
- Swelling
- Pathological fractures (late sign)

Preoperative work-up

- Blood count, electrolytes, Ca, P, alkaline phosphatase, LDH, creatinine, urine
- Bone marrow aspirate
- Local X-ray (showing osteolyses, bone formation and Codman's triangle) and chest X-ray
- Bone scintigraphy
- MRI
- CT-scan
- Preoperative chemotherapy

Operation

- En bloc resection
- Placement of the definitive endoprostheses (preferably of the growing type)

- Amputation/disarticulation if unavoidable
- Proceed with adaptation of a prosthesis
- Surgery for local metastases (namely in the lung)

Postoperative care

- Chemotherapy
- Physiotherapy/rehabilitation

Prognosis

- 80% cure rate at 5 years

29 Urogenital Tract

29.1 General Considerations

Pediatric urology specialists care for children with congenital conditions and acquired diseases. Specifically this includes affections of the kidneys, ureters, bladder, urethra, and the internal and external genital organs for both boys and girls from birth to early adulthood.

- Disorders of the urinary tract system are found in 0.5%–5% of pregnancies
- Congenital anomalies of the kidney and urinary tract constitute approximately 20%–30% of all anomalies identified in the prenatal period
- As not all newborns with diseases detected antenatally need long-term medical care, a postnatal clinical examination with ultrasonography should be carried out at a pediatric center specializing in urology on days 3–7, in order to inform the management team which additional diagnostic and therapeutic measures need to be taken

The most common problems in pediatric urology include

- Urinary tract infection
- Phimosis and paraphimosis
- Minor malformations of the penis (e.g., chordee)
- Hypospadias
- Undescended testicle (cryptorchidism)
- Ureteral obstruction
- Ureteral reflux
- Urolithiasis (bladder and kidney stones)

- Urethral obstruction
- Bladder control problems such as enuresis and daytime urinary incontinence
- Neurogenic bladder
- Tumors and cancers of the kidneys
- Ovarian cysts and tumors
- Repair of genitourinary trauma
- Genitourinary malformations and birth defects
  - Prune belly syndrome
  - Cloacal exstrophy
  - Bladder exstrophy
  - Epispadias
  - Intersex conditions
  - Ambiguous genitalia
- Signs of urogenital problems occur in addition to non-specific problems with urination
- Impaired growth
- Serum electrolyte imbalance
- Late or incorrect treatment results in chronic renal failure with the necessity for
  - Dialysis
  - Kidney transplantation

29.2 Organ Anatomy

Urogenital tract organ anatomy is shown in Table 29.1.

Table 29.1 Urogenital tract organ anatomy

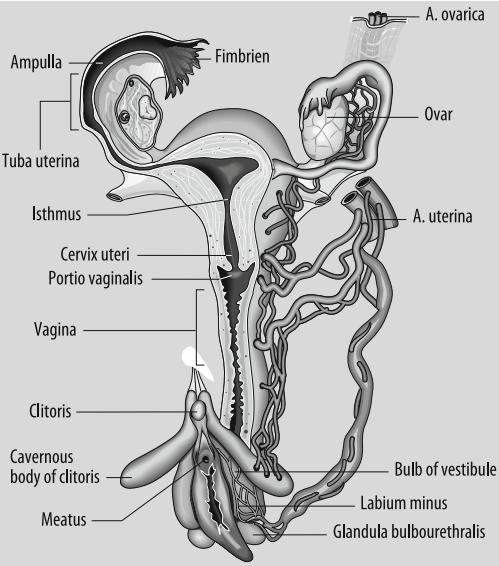
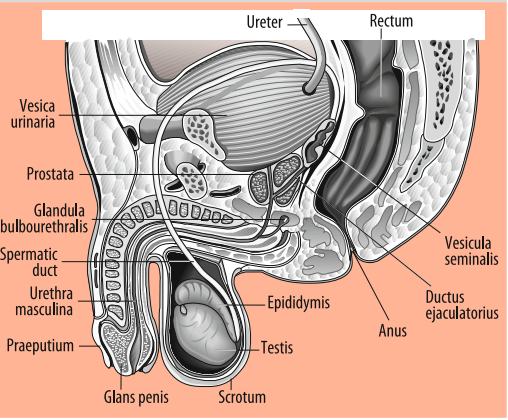
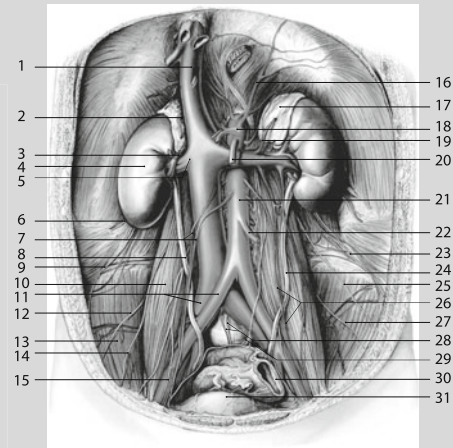
Organ systems	Organs	Anatomy
Female genital organs	Vagina Uterus Ovaries	
Male genital organs	Penis Testicles	

Table 29.1 (*continued*) Urogenital tract organ anatomy

Organ systems	Organs	Anatomy
Upper urinary tract	Kidneys and ureters	
Lower urinary tract	Bladder and urethra	

1. inferior vena cava 2. right suprarenal vein 3. right renal artery 4. right kidney 5. right renal vein 6. subcostal muscle 7. right testicular artery and vein 8. right ureter 9. iliohypogastric nerve 10. psoas femoral cutaneous nerve 11. common ilial artery and vein 12. ilioinguinal nerve 13. iliac muscle 14. lateral femoral cutaneous nerve 15. genitofemoral nerve 16. phrenic artery 17. left suprarenal gland 18. coelic trunc 19. left suprarenal vein 20. superior mesenteric artery 21. abdominal aorta 22. inferior mesenteric artery 23. iliohypogastric nerve 24. left ureter 25. ilioinguinal nerve 26. left testicular artery and vein 27. lateral femonral cutaneous nerve 28. median sacral artery and vein 29. superior rectal plexus 30. sigmoid colon 31. urinary bladder

29.3 Vascular Anatomy

- The aorta and iliac arteries form the vascular supply for most of the organs of the urogenital system (Fig. 29.1)
- Good compensatory mechanisms are in place in the event of a thrombosis, embolization, injury or iatrogenic lesion

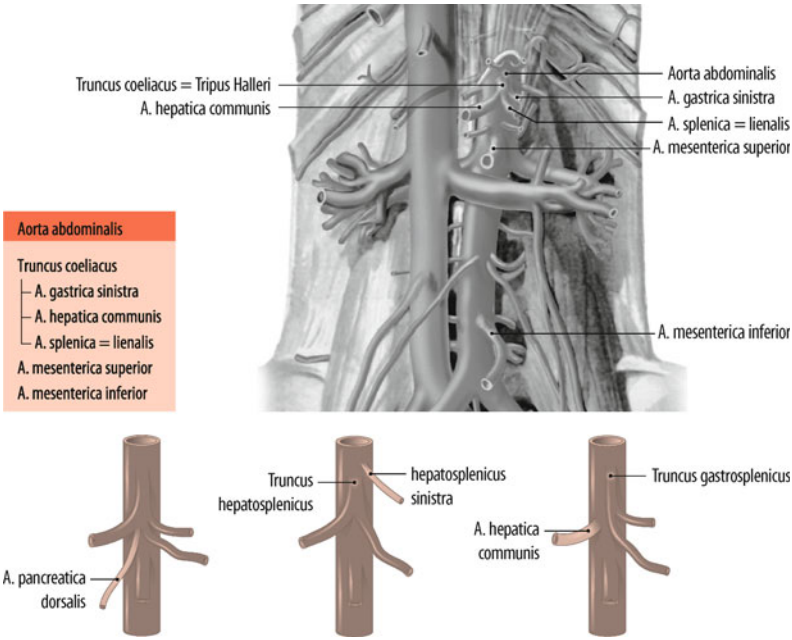


Fig. 29.1 Vascular anatomy of the abdominal vessels

Urinary tract

- Renal arteries arise from the aorta at the level of the second lumbar vertebral body
- The left renal artery runs along the tail of the pancreas and the right renal artery along the head of the pancreas behind the inferior vena cava to the kidney. Both are overlaid by the venous renal vessels
- However, variations are common, as during embryonic development the renal segments produce a number of arteries, which do not all merge to form a single common renal artery

- The right artery and the left renal vein are very short and require careful handling during surgery
- The blood vessels of the ureter come segmentally from the kidney, the retroperitoneal connective tissue, and the testicular or ovarian and vesical arteries

Male genital system

- The testicular artery arises on both sides from the aorta below the renal artery. It crosses the psoas muscle, the ureters, and the external iliac artery, and then goes with the spermatic cord through the inguinal canal
- The testicular veins on the right side follow the same course as the artery. On the left side they cross behind the sigmoid colon and lead to the renal vein at right angles
- The vascularization of the penis is separate

Female genital system

- The ovaric artery arises on both sides from the aorta under the renal artery. It crosses the psoas muscle, the ureters, and the external iliac artery supplying the ovary along the upper border of the latum ligament. An anastomotic arcade with the uterine artery is usually present
- The vascularization of the uterus and vagina is separate

29.4 Development of the Genital Organs in Puberty

- The anatomy of the genital organs changes during childhood and can be influenced by endocrinological disorders
- The aspect of the external genital organs should be classified according to the Tanner system (Table 29.2)

Table 29.2 Developement of genital organs in puberty

Parameter	Mean (years)	SD
Girls		
Pubic hair growth starts	10.4	1.2
Breast development (thelarche) begins	10.9	1.2
Menarche	13.4	1.1
Full pubic hair growth	14.0	1.3
Full breast development	14.0	1.2
Delay between thelarche and menarche	2.2	1.1
Boys		
Genital development starts	11.2	1.5
Pubic hair growth begins	12.2	1.5
Full pubic hair growth	14.9	1.0
Mature male genitals	14.7	1.1
Testicular volume >3 ml	11.8	0.9
Full testicular volume	15.3	1.2
Delay between genital development and mature male genitals	3.5	1.1

29.5 Pathophysiology

- The urogenital system normally functions as a coordinated unit
- Disorders of one organ may be responsible for dysfunction or disease of the whole system (Fig. 29.2)
- The complex course of the ureter means that there are many places where an obstruction may occur, namely on the psoas muscle along the tips of the ribs, at the crossing behind the testicular vessels in the middle and then at the iliac vessels entering the pelvis
- Critical points for obstruction or functional problems include:
  - Ureteropelvic junction
  - Ureteral orifices into the bladder
  - The area between the prostatic and the membranous parts of the urethra

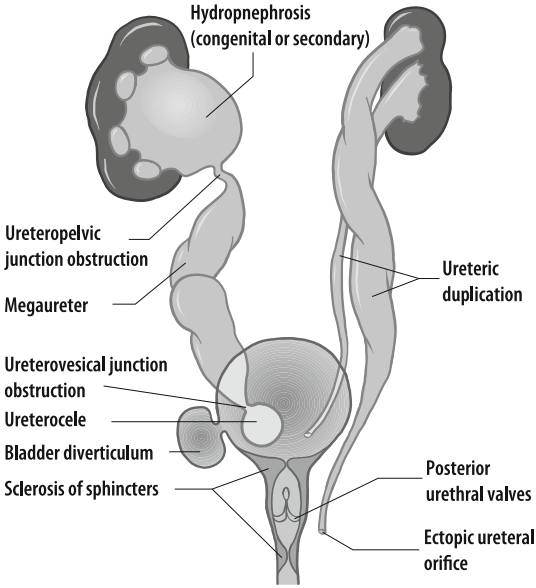


Fig. 29.2 Congenital anomalies of the urogenital tract

29.6 Principles of Pediatric Urology

- If one urinary tract anomaly is present, a second anomaly is likely
- The wide range of urogenital disorders and diseases, in terms of presentation, the patient's age, damage to renal function, and other functional problems, make it necessary for these patients to be cared for by specialized pediatric urologists and pediatric nephrologists teams, preferably in a specialist center
- Urinary tract infection and obstruction are common and dangerous problems in childhood
- The first aim is to protect renal function
- In the first year of life the vesicoureteral reflux and a very high pressure during micturition are physiological, and its coordination by the nervous system is still developing
- For this reason, bladder surgery should, if possible, be avoided during the first year of life

- One option is a cutaneous ureterostomy or suprapubic vesical drainage, if necessary
- For most problems there are guidelines for parents available from appropriate pediatric and urological associations
- Functional problems often lead to social and psychological problems

30 Penis

30.1 Phimosis/Paraphimosis

General considerations

- The prepuce is generally not retractable in the newborn; however, it can be retracted completely in 20% of boys at 6 months. The prepuce cannot be retracted in 60% of boys at 6 years (physiological phimosis)
- If forcible retraction of the prepuce is tried at this age, it causes a split along the length of the prepuce with consequent cicatrization leading to permanent preputial restriction (pathological phimosis)
- Recurrent balanitis also heals with cicatricial stenosis
- In those cases where the prepuce has healed with a stenosis, forced retraction can lead to a (paraphimosis), with significant swelling of the prepuce and reduced blood supply to the glans

Signs

- Lacerations on the prepuce seen upon attempted retraction
- Recurrent balanitis
- Painful micturition
- Ballooning of the prepuce during micturition
- Urinary tract infections

Preoperative work-up

- Clinical examination, without forcing the retraction

Circumcision

- Circumcision may be required on medical, ethnical or religious reasons
- Circumcision may be complete or partial

Total circumcision

- Total circumcision may be performed in the newborn period using the Gomco clamp, the PlastiBell or the Morgan clamp
- After the newborn period surgical circumcision is recommended
- General surgical guidelines include complete sterile dissection, complete separation of the glandular adhesions, exclusion of hypospadias
- Bleeding is a common complication after circumcision unless meticulous hemostasis with bipolar diathermy is performed. Monopolar diathermy is contraindicated

Circumcision in the newborn period

- Circumcision is performed 2–4 hours after the last feed
- Perform a dorsal penile nerve block
- It is important to dilate the preputial orifice (or dorsal slit) to visualize the urethral orifice and exclude hypospadias
- It is advisable to mark the level of prepuce excision to ensure that the appropriate amount of skin is excised before applying the clamp
- Leave the clamp in place for 10 min, avoiding torsion of, or tension on, the penile shaft; excise the redundant skin prepuce and remove the clamp, ensuring hemostasis
- If a PlastiBell clamp is used, place a cotton tie around the clamp, excise the prepuce, break the clamp handle off, and leave the rest of the device to separate over time
- Apply antiseptic ointment to the junction between the skin and the inner epithelium of the glans

Total circumcision after the newborn period

- The operative steps are illustrated in Fig. 30.1
- Widen the prepuce with dissecting forceps
- Expose the coronal sulcus and lyse the preputial adhesions

- Reposition the prepuce and apply traction with two forceps
- Make an incision in just the outer cutaneous layer, distal to the coronal sulcus (which gleams through the skin), drawing it obliquely, below from the back and above to the front
- Mark the inner layer 2 mm from the coronal sulcus and incise dorsally
- Incise the inner layer in a semicircular shape at about 2 mm from the coronal sulcus to the frenulum
- Clamp the frenulum and excise the prepuce
- Reconstruct the frenulum with three to four stitches. Suture both crura of the glans
- Suture the inner and outer layers together taking care to avoid bleeding. To achieve this, the circumference is first subdivided into four quadrants, and then all quadrants are sutured

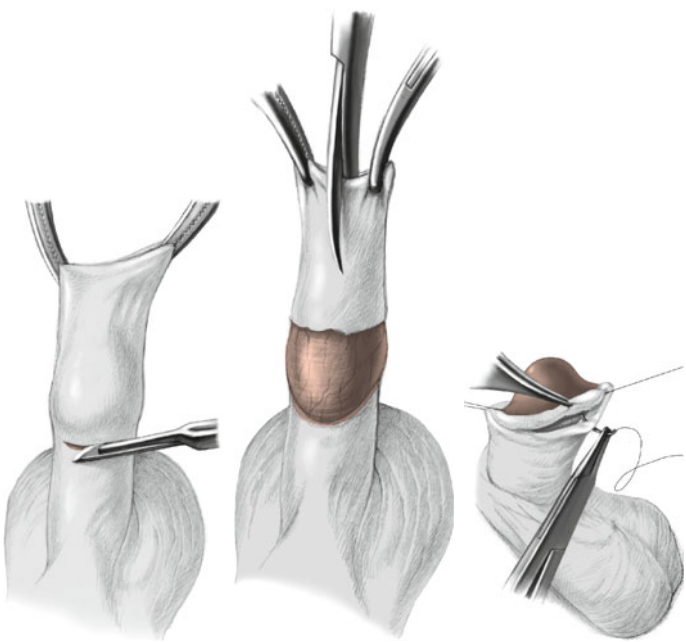


Fig. 30.1 Operative steps: circumcision

Partial circumcision

- In cases of partial circumcision, some two-thirds of the prepuce must be left to cover the glans
- The success of this operation essentially lies in the skin incision. The prepuce should be incised immediately below the stenotic ring, to ensure that the scarred prepuce is completely removed

Postoperative care

- Apply gauze medicated with local anesthetic
- Warm baths from the 4th postoperative day with cautious retraction of the remaining prepuce

Paraphimosis reposition

- Apply an anesthetic cream to the glans and wait for the ointment to act
- Grasp the prepuce with gauze and try to squeeze circumferentially around the edema in the penis' shaft
- Constant pressure should be applied for a long time
- In cases where reduction is impossible, surgical treatment is indicated
- Perform a dorsal incision along the prepuce with section of the phimotic ring
- Carry out secondary definitive surgical repair

30.2 Hypospadias

General considerations

- Hypospadias is the most frequent urogenital anomaly, occurring in 3 per 1000 live births; it presents as incomplete closure of the urethral groove
- Circumcision must be avoided and early counseling of the parents should be provided
- May be associated with ventral curvature, "chordee"
- There is a familiar tendency in 5%–10%

- It may be associated with cryptorchidism and/or urethral valve anomalies
- The penis grows less than 1 cm in the first 4 years of life: the phallus that is small at 3 months will still be small at 3 years
- Sexual identity is determined by 3 years of age. In older children the psychological burden relating to this must not be underestimated (in some cases this amounts to the sensation of being "different" from one's peers; in others, repeated operations on genitalia)
- Recent studies evaluating emotional, psychosexual, cognitive, and surgical risks identified that there is an optimal window for surgery at 3–18 months of age

Classification

- Consistent classification is necessary in order to standardize the terminology of hypospadias, and to enable improved treatment and comparison of results across centers and surgeons
- It is suggested that forms such as those in Figs. 30.2 and 30.3 are always completed at the first operation and, if needed, after follow-up examinations

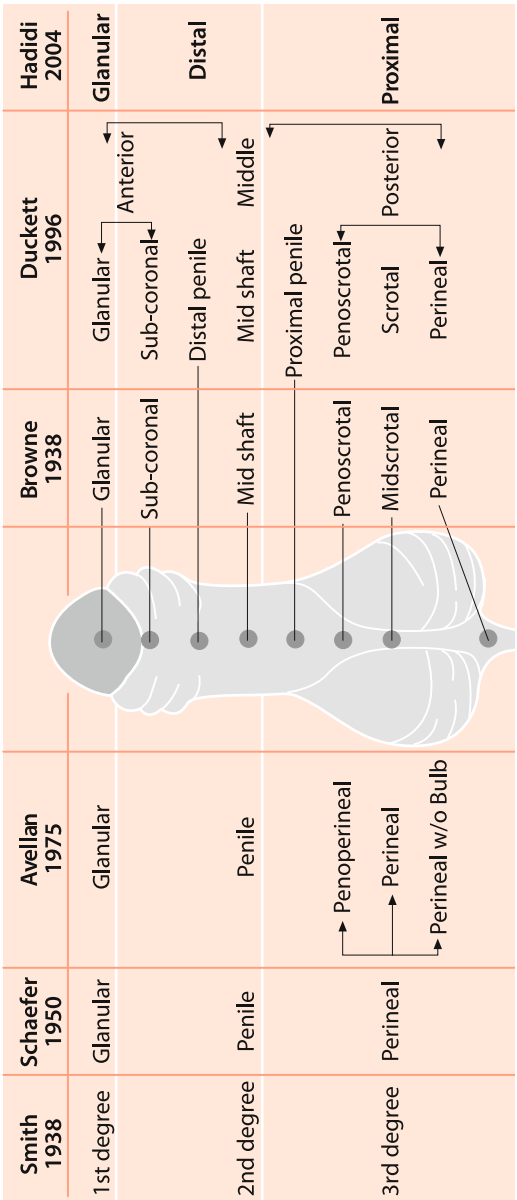


Fig. 30.2 Classification of hypospadias

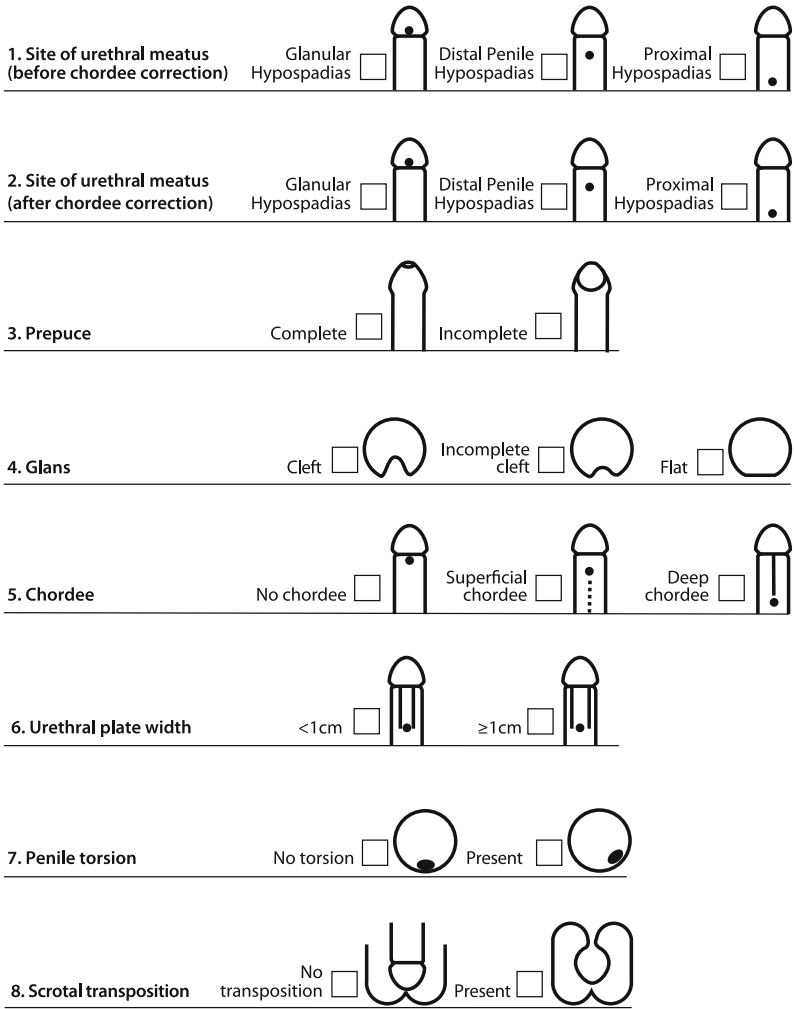


Fig. 30.3 Classification sheet for hypospadias and related abnormalities

Signs

- Hypospadias is usually asymptomatic
- Signs are usually caused by a narrow meatus and downward direction of the urine stream (voiding troubles)

Preoperative work-up

- Clinical examination, with evaluation of
  - The meatus size and location
  - Glans
  - Width of urethral plate
  - Presence of chordee
  - Prepuce
  - Penile torsion
  - Scrotal transposition and width
- Classify the form of hypospadias
- Ultrasonography of kidneys and bladder (to rule out the presence of associated congenital anomalies and to evaluate the residual volume after voiding)
- Flowmetry (voiding troubles)

Technical recommendations

- Fine surgical instruments are essential in hypospadias repair
- Optical magnification is advisable
- Operation can be performed using a tourniquet, which should be released every 40 min
- Only bipolar diathermy should be used for hemostasis
- The repair should be performed around a large catheter (at least 8 or 10 F)
- Transurethral catheter use is associated with a relatively high complication rate, and a suprapubic catheter is recommended only for proximal hypospadias repair
- For hypospadias surgery only very fine sutures must be used (6-0 or 7-0)

- Every operation should start with calibration and dilatation of the meatus
- Urethrocystoscopy is recommended to exclude associated anomalies
- An artificial erection test is performed to identify and, if appropriate, correct chordee
- More than 300 techniques and modifications have been described for hypospadias correction. The following recommendations (Table 30.1) must be regarded only as a suggestion

Table 30.1 Recommended methods of hypospadias correction

Hypospadias after chordee release		
Glandular	**Distal penile**	**Proximal**
▪ Inverted Y technique ▪ MAGPI ▪ Y-V modified Mathieu	▪ Y-V modified Mathieu ▪ Tubularized incised plate (TIP)	▪ Lateral-based flap ▪ Onlay island flap ▪ TIP ▪ Two stage repair

Operation Steps

Meatotomy

- Dilate the meatus with a bougie à boule (i.e., a bulb-tipped bougie)
- Make a longitudinal incision on the meatus of adequate length
- Suture the urethral epithelium to the glans

Chordectomy

- In cases of chordee, free the penile urethra completely from the meatus as far as the perineum
- Dissect the fibrous bands running parallel to the urethra, and resect them until the penis shaft is straightened
- Evaluate the residual curvature, preferably when the penis is erect. To induce an artificial erection, inject 0.9% saline solution into the corpora cavernosa using a 25-G butterfly needle (Gittes's maneuver) (Fig. 30.4)

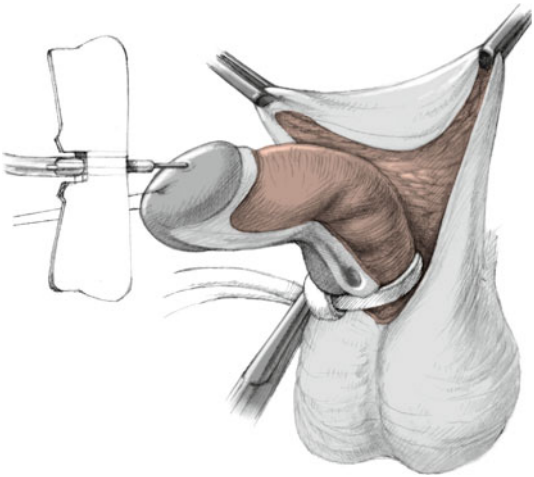


Fig. 30.4 Chordectomy: the artificial erection test

Meatal advancement of the urethra

- This technique is only suitable for patients with glandular hypospadias and a mobile urethra
- Make a circular incision in the meatus and hold it with a stay suture
- Dissect the urethra longitudinally along the shaft, if necessary as far as the penoscrotal junction
- Careful coagulation with bipolar electrocautery reduces bleeding and postoperative edema
- Then follow either of these two options:
  - Split the glans along the middle, pull the urethra to its tip, and suture
  - Incise the urethra dorsally for 2–3 mm, creating a V shape to increase its lumen (author's preferred method)
- Make a M-shaped incision on the glans tip
- Stitch the central point of the M at the deepest point of the V, to result in a wider lumen
- Suture the two glandular wings over the urethra from the top distally
- The operative steps are illustrated in Fig. 30.5

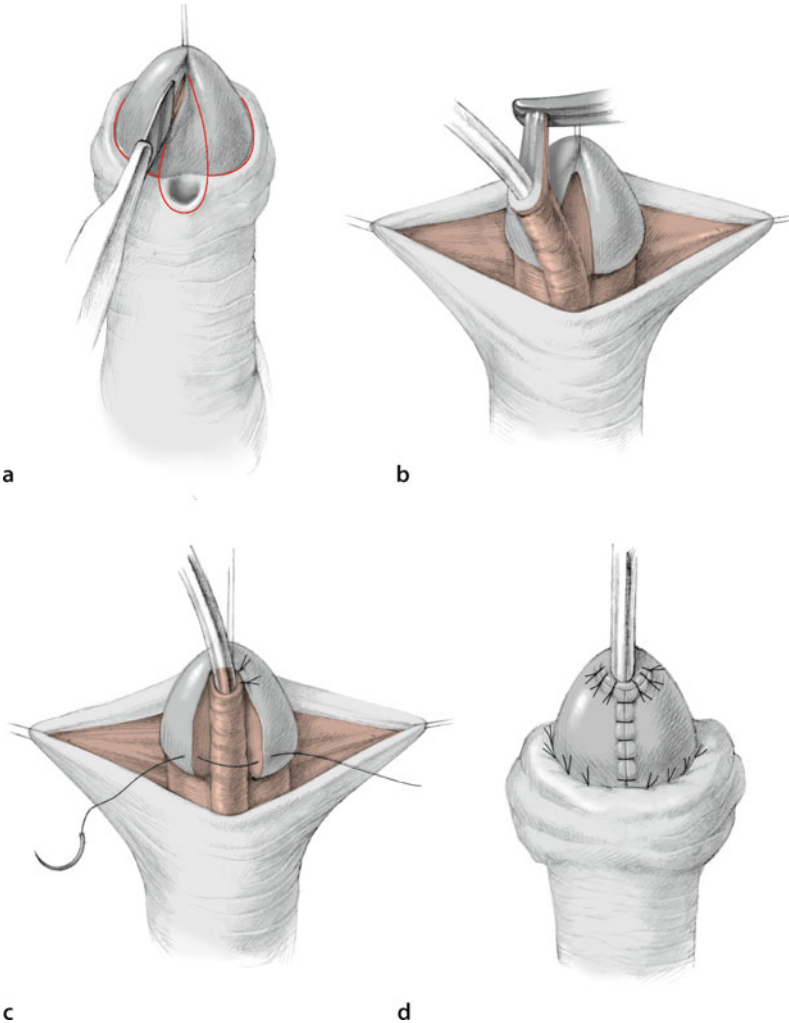


Fig. 30.5a–d Meatal advancement of the urethra

MAGPI (meatal advancement and glanuloplasty)

- Make a circular incision in the prepuce at the level of the coronal sulcus
- Make a longitudinal incision at the inner aspect of the meatus as far as the tip of the glans
- Suture this longitudinal incision transversally with single stitches
- Secure the anterior aspect of the newly created meatus with a stay suture and pull it up to the tip of the glans
- Make a sharp incision in both glanular wings in an inverted V-shape
- Suture the glanular wings in the apical to distal direction, to enclose the urethra within the glans
- Suture the outer layer of the prepuce to the coronal sulcus
- Preputial reconstruction, though possible, is not recommended at this stage
- The operative steps are illustrated in Fig. 30.6

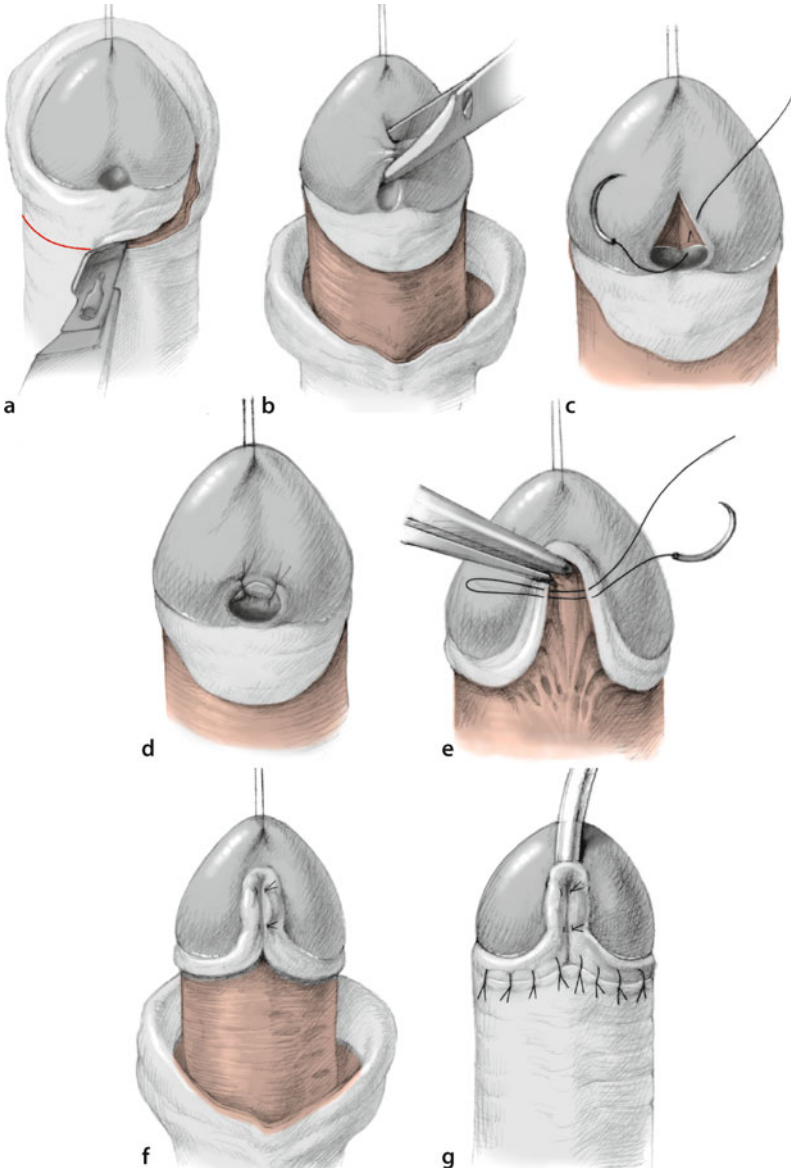


Fig. 30.6a–g MAGPI (meatal advancement and glanuloplasty)

Y-V Mathieu repair

- This is the most popular technique for distal hypospadias repair (Fig. 30.7)
- The Y-V modification avoids the drawback of the original Mathieu repair, which involves making a circular meatus that is not at the tip of the glans
- Outline A Y-shaped incision on the glans
- Close this Y-incision as a V, creating a bulge on either side, like "dog ears"
- Outline the flap so that the distance between the meatus and the proximal end of the flap is slightly greater than the distance from the meatus to the tip of glans
- Make A U-shaped incision extending from the tip of the V in the glans down to the lower end of the designed flap; this results in two glanular wings
- Mobilize the Mathieu flap, preserving its fascial blood supply
- Perform urethroplasty using continuous subcuticular polyglactin 6-0 sutures
- Fashion a protective intermediate layer, using the flap fascia or dartos fascia
- Suture both granular wings together around a neo-urethra using interrupted mattress sutures

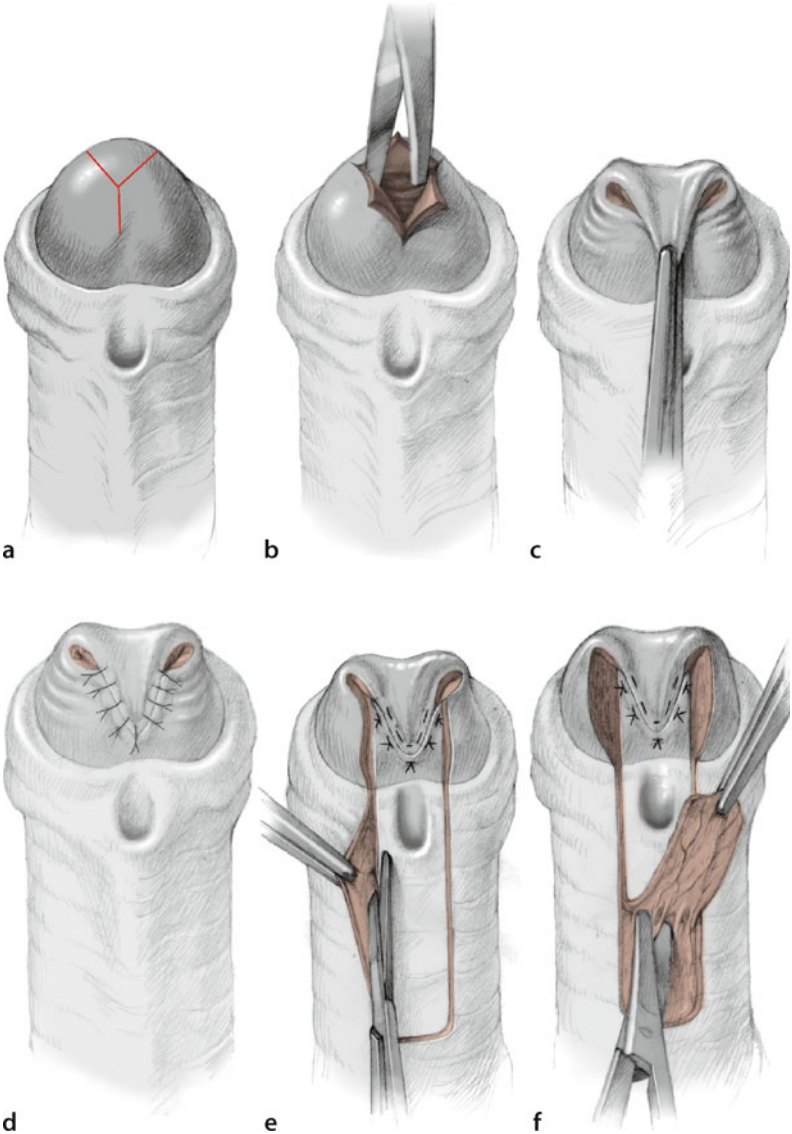


Fig. 30.7a–j Y-V Mathieu repair

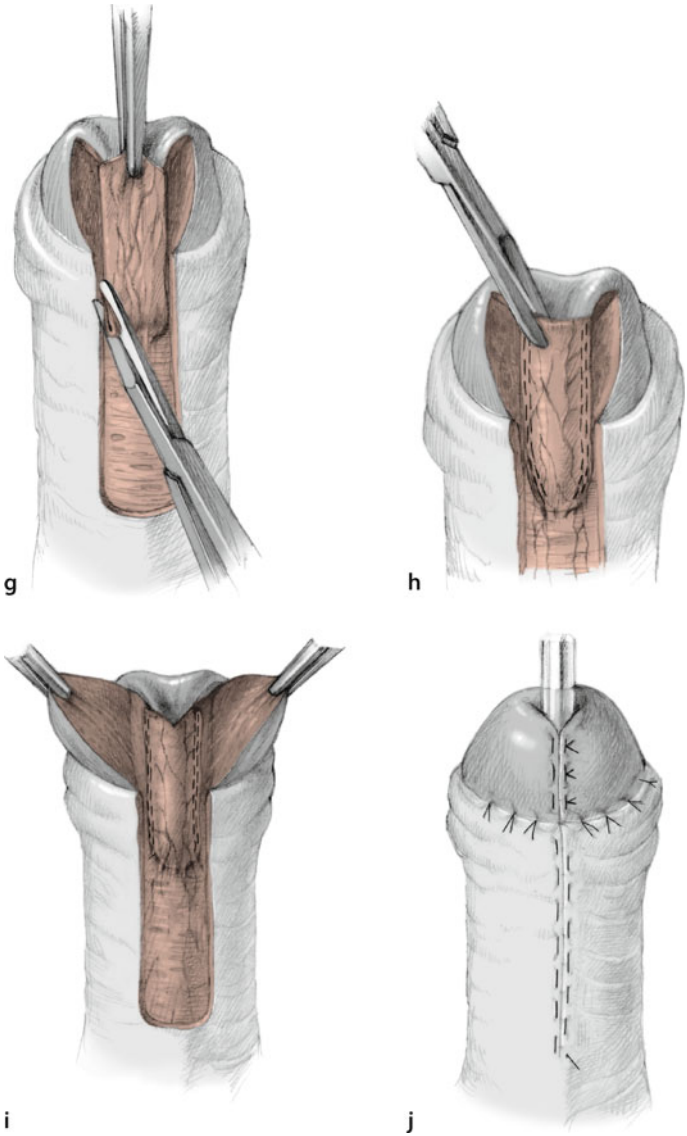


Fig. 30.7i–j *(continued)* Y-V Mathieu repair

Tubularized incised plate urethroplasty (tip urethroplasty)

- The operative steps are illustrated in Fig. 30.8
- Make a circumscribing skin incision 1–2 mm proximal to the meatus
- Deglove the penis skin to the penoscrotal junction
- Separate the urethral plate from the glans wings by parallel incisions
- Mobilize the glans wings, avoiding damage to the margins of the urethral plate
- Make a relaxing incision using scissors in the midline from within the meatus to the end of the plate
- The depth of this relaxing incision depends on the plate width and depth
- Preferably with 7-0 polyglactin, tubularize the urethra on the inserted catheter, placing the first stitch at approximately the midglans
- Complete the tubularization with a two-layer running subepithelial closure
- Develop a dartos pedicle from the dorsal shaft skin, button-hole it, and transpose it to the ventrum to additionally cover the repair
- Suture the skin edges of the tubularized glans together with the meatus
- This method has become popular because of its simplicity; however, it may be associated with meatal stenosis and fistula

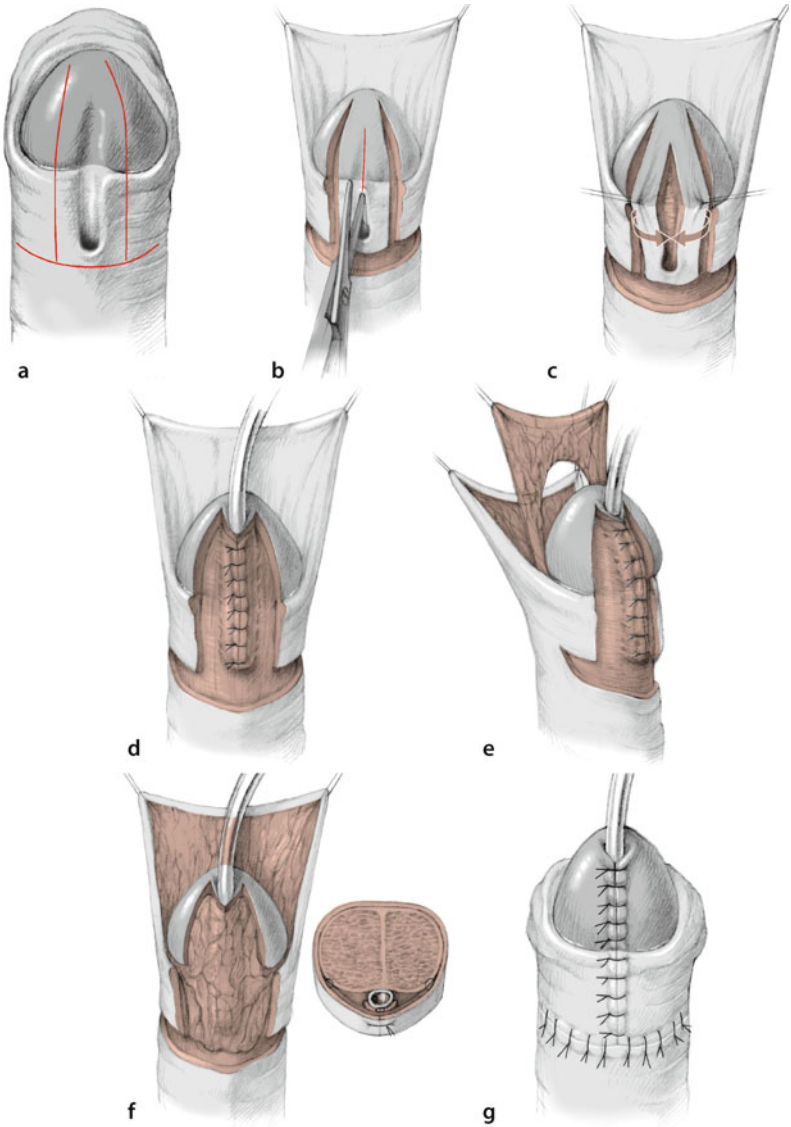


Fig. 30.8a–e Tubularized incised plate urethroplasty (tip urethroplasty)

Lateral-based flap

- The lateral-based flap may be used in all types of proximal hypospadias
- It has double blood supply and allows extensive excision of ventral chordee
- Make a deep Y-shaped incision on the glans that goes all the way down to the coronal sulcus. This permits two deep glanular wings and a wide meatus
- Outline a rectangular skin strip that extends proximally from the urethral meatus, staying at the midline in the scrotum to avoid potentially hair-bearing skin
- Extend the skin strip distally and laterally by curving towards the prepuce. This allows for formation of a very long tube
- Mobilize the flap with its pedicle through the dorsum and down to the root of the penis to avoid penile rotation
- Tubularize the flap using continuous subcuticular 6/0 polyglactin
- Cover the neourethra with a protective intermediate layer (dartos or tunica)
- Construct the neomeatus by suturing the terminal end of the neourethra to the center of the glans
- Suture the glanular wings around the neourethra using interrupted mattress sutures
- Insert a percutaneous suprapubic cystocath into the bladder for 10–14 days
- Apply a compression dressing for 6–24 h for hemostasis
- The operative steps are illustrated in Fig. 30.9

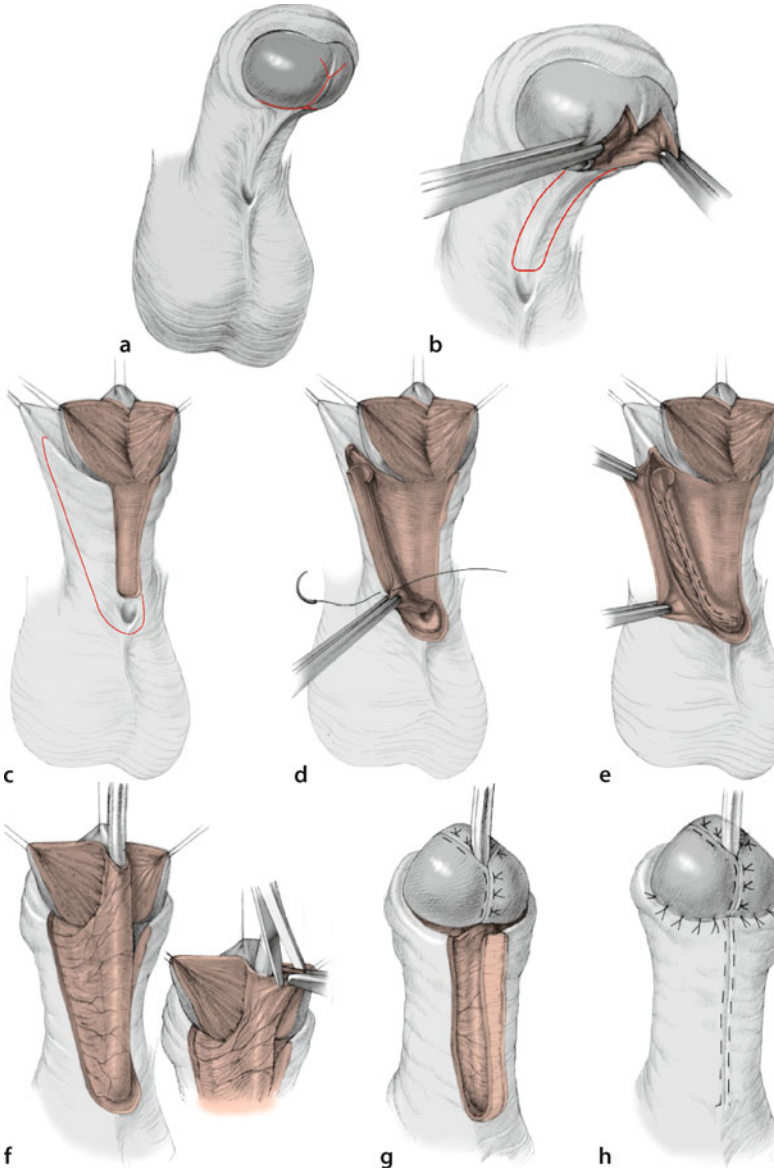


Fig. 30.9a–h Lateral-based flap

Transverse preputial island flap (Duckett operation)

- Create a neourethra utilizing the inner preputial layer and anastomose it distally with the native urethra and apically with the glans
- Make a circular incision of the meatus and of the inner preputial layer along the coronal sulcus
- Dissect the urethra and chordectomy
- Estimate the length of the urethral defect while the penis is erect
- Fix the prepuce with four holding sutures, so that its inner layer lies flat
- Once the length of the neourethra is determined, incise the inner layer accordingly
- Separate the inner layer from the outer one, taking particular care not to injure its blood supply
- Conduct the dissection of the vascular pedicle in such a way that rotation of the neourethra, in the craniocaudal direction, is possible without tension
- Create the neourethra by rolling up the inner preputial layer on a catheter as a tube and closing it with a running suture
- On the anterior aspect of the penis make a channel from the coronal sulcus beyond the frenulum, as far as the tip of the glans, through which a passage is formed
- Make an oblique anastomosis between the neourethra and the native urethra, in such a way that the suture line on the neourethra lies on the penis shaft
- Pull the neourethra through the channel previously developed up to the glans tip and fix it to the glans
- Close the skin of the penis on the shaft with a running suture
- Suture the outer layer to the inner one
- It is recommended to avoid reconstruction and removal of the prepuce until a successful result is obtained
- The operative steps are illustrated in Fig. 30.10

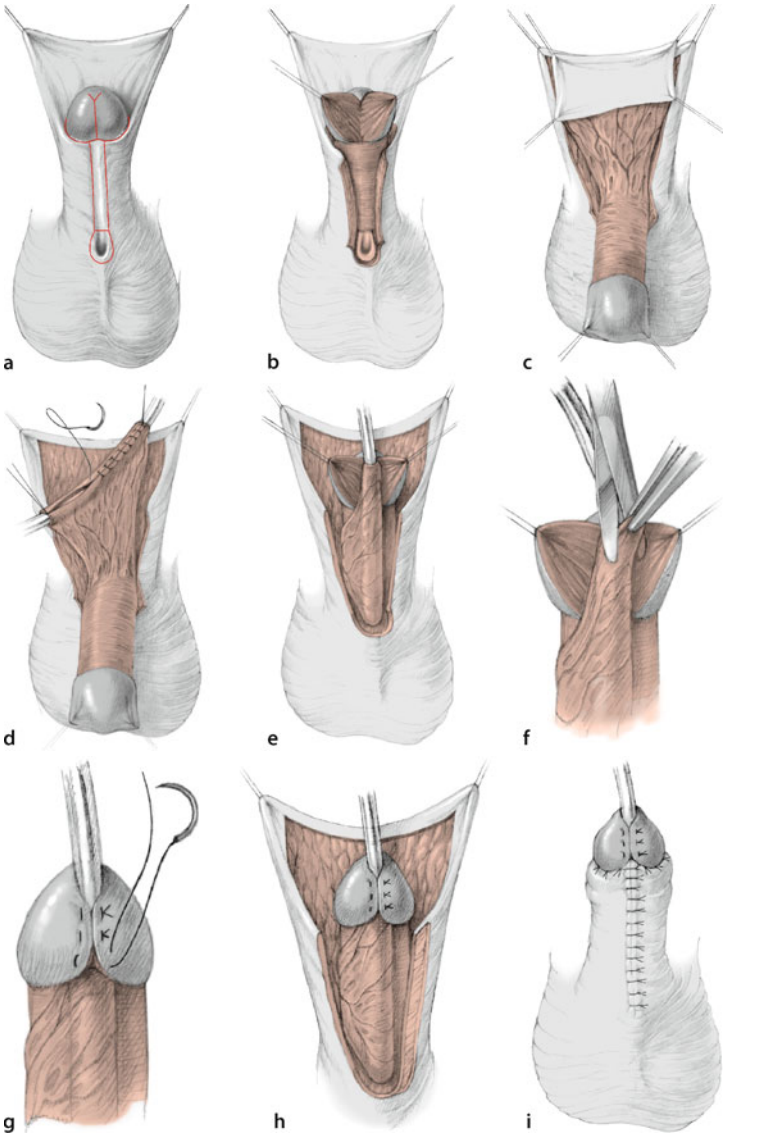


Fig. 30.10a–i Transverse preputial island flap (Duckett operation)

Onlay island flap

- Instead of creating a complete neourethra from the inner preputial layer, it is possible to complete the urethral plate with the layer itself
- Make a semicircular skin incision along the urethral plate around the meatus
- Dissect a pedunculated flap from the inner preputial layer in the same way as for the Duckett's operation (above)
- Split the glans in line with the urethral plate
- Transpose the flap on the ventral aspect of the penis shaft and suture both flap borders with the free borders of the urethral plate
- Close the skin and the glanular wings
- The operative steps are illustrated in Fig. 30.11

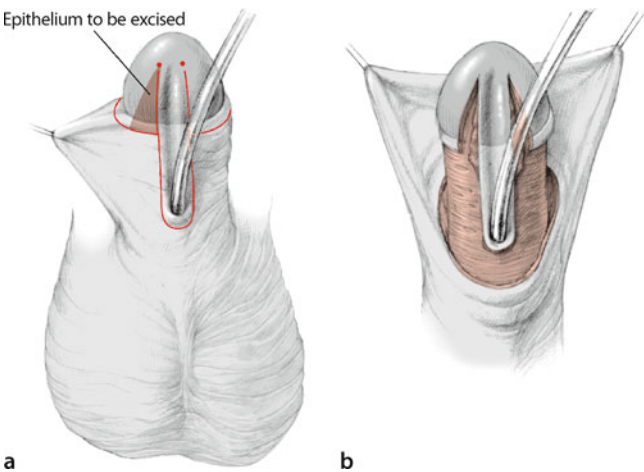


Fig. 30.11a–e Onlay island flap

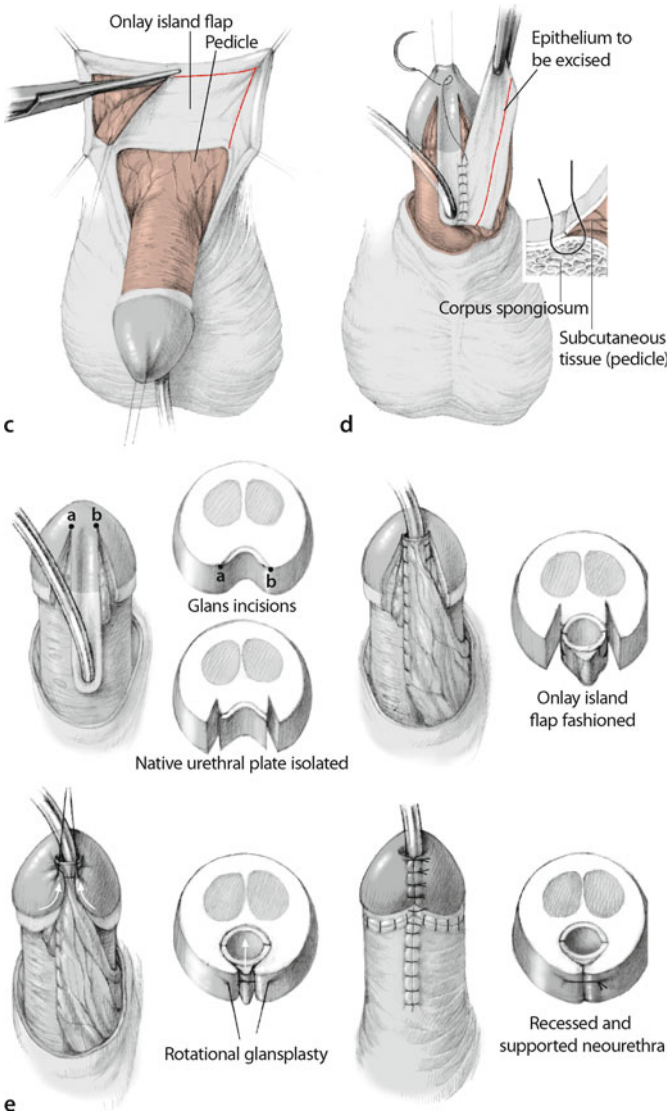


Fig. 30.11a–e *(continued)* Onlay island flap

Neourethra from buccal/bladder mucosa

- In re-do operations it is possible to resort to buccal or bladder mucosa to form a wide urethral plate as a first stage
- Reconstruct a neourethra in the second stage
- Bladder mucosa and one-stage repair using buccal mucosa are becoming less popular in complicated proximal hypospadias due to the high incidence of complications

Fistula

- Fistula is the most frequent postoperative complication
- Several factors may be responsible
  - Distal stenosis
  - Technique applied
  - Skin damage
  - Bleeding
  - Edema
  - Tension on the sutures
  - Overlapping of suture lines
- There are three important steps to the correction of urethral fistula
  - Exclusion and correction of distal stenosis
  - Wide excision of the fistula tract
  - The use of a protective intermediate layer between the urethra and skin closure

Postoperative care

- Apply a compression dressing. The author removes the dressing after 6 h
- Many surgeons leave a silastic stent or catheter for 7–10 days. The author does not leave stent inside the urethra more than 48 hours because it increases the incidence of complications
- Instead
  - Patients with distal hypospadias are allowed to pass urine through the repair within 48 hours
  - Proximal hypospadias patients are diverted with a suprapubic catheter for 12–14 days

- Oral antibiotic prophylaxis is recommended
- Follow-up twice in the first year
  - Control of micturition
  - Uroflowmetry

Complications

- Fistula formation (5%–30%)
- Meatal stenosis
- Anastomotic urethral stenosis

Prognosis

- Good for distal and middle hypospadias, in the hands of a fairly skilled surgeon
- Good for the proximal hypospadias, in the hands of an expert

31 Testicles

31.1 Cryptorchidism

General considerations

- The term cryptorchidism defines a situation in which the testis is neither visible nor palpable in the scrotum (kryptòs = hidden + òrchis = testis)
- In clinical usage it denotes a synonym of undescended testis, although not all "cryptorchid" testes are "undescended"
- Overall incidence
  - Premature babies up to 20%
  - Full-term neonates 4%–5%
  - 3-month-old infants 1%–2%
  - 1-year-olds 0.3%
- In a great number of cases where testicular descent is delayed secondary ascension may occur
- The undescended testis is often associated with an open vaginal process or an inguinal hernia which hinders the normal descent
- Consequences of cryptorchidism are infertility, increased risk of testicular torsion, and increased risk of malignant degeneration (about eightfold increase)

Classification

- The classification of cryptorchidism is given in Table 31.1
- The different positions in which the testis may be found are shown in Fig. 31.1

Table 31.1 The classification of cryptorchidism

Type	Description		Incidence (%)
Hypoplastic or absent testis	Testis never develops (agenesis) or disappeared after an intrauterine torsion (anorchia)		3–5
Undescended testis = Testicular retention	Testis whose descent stopped before reaching the scrotum		
	Abdominal retention	Intra-abdominal testis, above the internal inguinal ring	5
	High inguinal retention	Testis at the level or just below the inner inguinal ring	15–20
	Low inguinal retention	Testis at the level or above the outer inguinal ring	45–50
	Gliding testis	The testis can be brought as far as the upper scrotum, but retracts immediately once released	
Ectopic testis (ek = outside tòpos = site)	Testis whose descent took a wrong direction	Perineal Crossed (contralateral hemi-scrotum) Penis root Crural	2
Retractile testis	Descended testis changing its position	In the warm the testis is in the scrotum while in the cold it is in the inguinal canal	30
Secondary cryptorchidism	Testis no longer palpable in the scrotum after a normal descent or following surgery (e.g., inguinal hernia)		

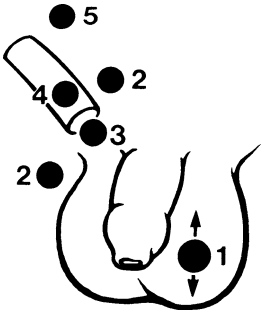


Fig. 31.1 Position of the testis. [*1* Retractile testis, *2* ectopic testis, *3* gliding testis, *4* inguinal testis (low or high), *5* abdominal testis (true retention)]

Embryology

- Descent of the testis occurs by two different mechanisms, one morphologic and the other hormonal
- The key to the morphologic mechanism is the gubernaculum testis, which in the early embryonic period joins, in form of a band, the testis with the inguinal region. As the embryo develops, the testis is pulled downwards. The vaginal process and the inguinal canal, through which the testis descends into the scrotum, are formed with migration of the gubernaculum in the scrotum
- The hormonal mechanism is not yet fully understood. It probably involves Müllerian inhibiting substance (MIS), testosterone and calcitonin gene-related peptide (CGRP). These substances stimulate the genitofemoral nerve, which regulates the migration of the gubernaculum
- When one or both mechanisms fail, undescended testis occurs

Signs

- The main sign relates to the scrotum which, the parents notice, is empty

Preoperative work-up

- Clinical examination is performed with the patient in a supine position. Squeeze the inguinal canal with one hand while trying with the other one to grasp the testis and bring it down into the scrotum
- Ultrasonography if an abdominal testis is suspected

- MRI if the testis cannot be found by clinical or ultrasonographic investigation
- Diagnostic laparoscopy, to determine the site and aspect of the testis, with the possibility of carrying out further therapeutic procedures in the same operation
- Chromosomal and hormonal examinations if intersexuality is suspected

Treatment

- The treatment options for cryptorchidism are given in Table 31.2
- Testicular descent should occur by the end of the second year of life
- During and after hormonal therapy regular controls should be performed, as relapses occur in up to 80% of cases. In light of its possible side-effects (induced apoptosis), a second conservative attempt is not warranted and surgical correction is indicated
- The indication for the operative intervention is given on diagnosis. The smaller the child, the more frequent the relapse and the testicular atrophy

Table 31.2 Treatment options for crytorchidism

Treatment	Details	Success rate (%)
hCG therapy: stimulation of Leydig cells, with incremental testosterone	1 year: 500 IU i.m. once a week for 5 weeks 2–6 years: 500 IU i.m. twice a week for 5 weeks >6 years: 1000 IU i.m. twice a week for 5 weeks	20–25
GnRH therapy: stimulation of LH and FSH, no incremental testosterone	Two intranasal puffs (0.2 mg) 3 times a day for 4 weeks	15–20
Combined GnRH–hCG therapy	GnRH therapy as above, followed by 1500 IU hCG once a week for 3 weeks	25–30
Primary operation	▪ Undescended testis with associated hernia ▪ Ectopic testis ▪ Secondary cryptorchidism ▪ Undescended testis in the puberty	95

Operations

Shoemaker's orchidopexy

- The operative steps are illustrated in Fig. 31.2
- Make an inguinal incision in the lower abdominal fold and open Scarpa's fascia
- Search for the testis, which can lie in the epifascial space
- Dissect the aponeurosis of the internal oblique muscle and dissect the gubernaculum
- Resect the gubernaculum and expose the external inguinal ring
- Make an incision in the fascia and dissect the spermatic cord as far as the internal inguinal ring
- Make an incision in the cremasteric muscle along the spermatic cord, looking for the remnant of the vaginal process or an inguinal hernia leading to the abdominal cavity
- Position a holding suture in the sheath testis/periorchium
- Dissect the vas deferens and the pampiniform plexus from the vaginal process or inguinal hernia, performing a ligation at the base
- Expose an open vaginal process in cases where there is one
- Dissect the spermatic vessels and the vas deferens up to the retroperitoneum, as far as is needed for orchidopexy
- Sharply resect the thin lateral spermatic fascia that hinders straightening of the spermatic cord
- Introduce a finger into the scrotum and stretch the hemiscrotum of the affected side with the thumb outside pulling downwards
- Make a skin incision on the scrotum and expose the dartos tunica
- Develop a pouch between the scrotal skin and the dartos tunica with scissors
- Open the dartos tunica and gently draw the testis through it down to the pouch
- Check the position of the spermatic cord along the inguinal canal (vas must be medial, vessels lateral)
- Narrow the opening in the dartos with resorbable sutures
- Place the testis in the pouch

- Close the scrotal incision with subcuticular absorbable suture (Vicryl rapid 5-0)
- Make a layered closure of the inguinal incision, in the same way as for an inguinal hernia

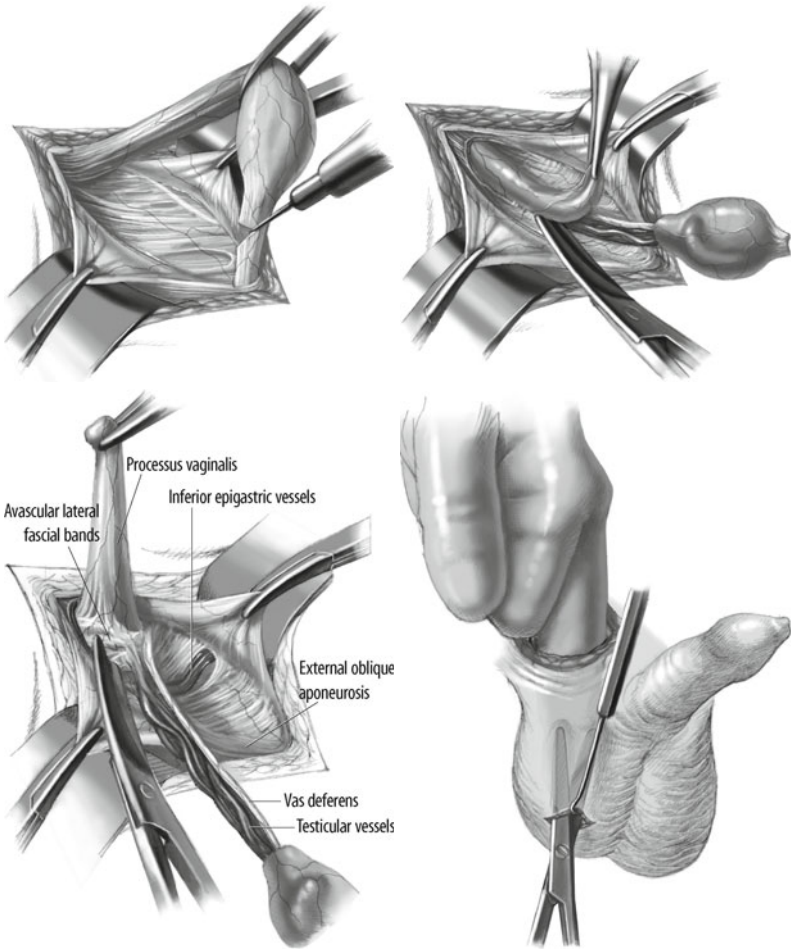


Fig. 31.2 Operative steps: orchidopexy

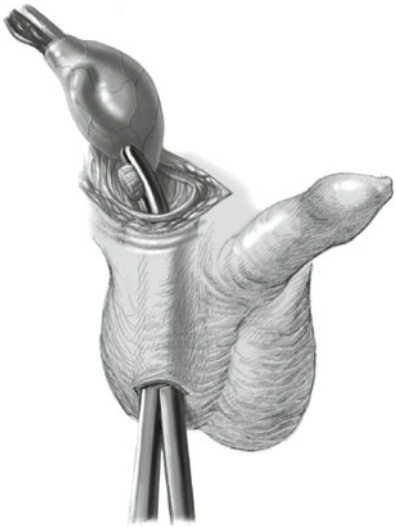


Fig. 31.2 *(continued)* Operative steps: orchidopexy

Orchidopexy

- Reserved for neonates and infants, when the dartos and the scrotal skin cannot be split apart
- Dissect the testis as above, although there is no need to prepare a scrotal pouch
- Place a suture in the tunica albuginea and another through the testicular parenchyma, without tying them (resorbable material)
- Secure all four ends of the sutures to a straight needle and pass it through the scrotal skin. The testis is thus brought to lie near the inner side of the dartos
- Transfix a soft gauze with two of the sutures and bring it near the scrotal skin. Then tie these with the two other sutures

Complementary techniques

- When the length of spermatic cord is too short to reach the scrotum, it is possible to run it under the epigastric vessels, a route that is about 0.5 cm shorter
- Open the transversal fascia and expose the epigastric vessels
- Under run the epigastric vessels with the vas and the testicular vessels, taking care not to twist them

Laparoscopy

- An abdominal testis can be dissected laparoscopically by liberating the vessels and the vas
- The testis can then be brought down to the internal inguinal ring
- Thereafter, orchidopexy can be completed through an inguinal approach in the same operation or in a second operation
- An atrophic testicular remnant can be removed laparoscopically

Fowler–Stephens procedure

- The testicular vessels are the main limiting factor for an orchidopexy
- The principle behind this procedure is that the testis is additionally vascularized by collateral vessels from the gubernaculum
- In the first operation, inspect the testis and the vessels laparoscopically and at the same time dissect the testicular vessels without impairing their vascularization from the gubernaculums
- In a second operation, evaluate the testis laparoscopically. The testis can then be placed in the scrotum via the same inguinal approach

Postoperative care

- Change dressing every 2 days
- Strict bed rest for 2–3 days

31.2 Testicular Torsion

General considerations

- This should be the first suspicion in cases of acute scrotum in childhood

- The disease has two different peaks of incidence: perinatally and between 10 and 25 years of age
- The risk of testicular torsion is increased in cases of undescended testis or trauma
- Differential diagnosis: orchitis, incarcerated inguinal hernia, hydrocele, epididymitis, idiopathic scrotal edema, and testicular appendages torsion

Table 31.3 Classification of testicular torsion

Intravaginal	Only the testis and the spermatic cord are twisted, with no involvement of the tunica vaginalis	Adolescents
Extravaginal	The entire tunica vaginalis is twisted and, as a consequence, also the spermatic cord	Neonates
Mesorchial	Torsion between the testis and the epididymis	Very rare

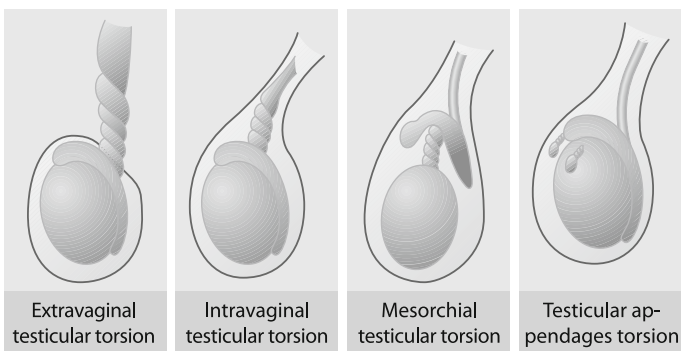


Fig. 31.3 Classification of testicular torsion

Classification

- Testicular torsion is classified as intravaginal, extravaginal or mesorchial (Table 31.3), as illustrated in Fig. 31.3

Signs

- Sudden severe testicular pain
- Swelling of the testis
- Pain upon palpation of the scrotum
- Redness of the scrotum

Preoperative work-up

- Examination of the cremasteric reflex
  - In extravaginal torsion it cannot be elicited
  - In intravaginal torsion it can be scantily elicited
  - In hydatid torsion it can be elicited
- Testicular Doppler ultrasonography examination
- Testicular radioisotope imaging is the most reliable examination, it is however not available immediately and thus is not always practicable
- If in doubt, an operative inspection of the testis is inevitable as prompt clarification differentiating between infection and torsion gives a far better prognosis

Operation

- Make a transverse scrotal incision or inguinal incision
- Open the tunica vaginalis
- Perform external dislocation of the testis
- Regardless of the anatomical finding (extravaginal or intravaginal torsion) the testis is detorsed
- Pack the testis in a warm and wet gauze and leave it to rest for 5–10 min
- Testicular recovery is evaluated on color and ability to bleed from a small incision in the tunica albuginea
- If necrotic, the testis must be removed
- If the testis recovers, pass two stitches through the parenchyma and secure them to the tunica vaginalis
- An orchidopexy on the contralateral side is to be recommended. A point of dispute is whether this additional step should be performed during the same anesthetic or in a second operation 6 weeks later
- Make a layered closure of the scrotum

Postoperative care

- Uplift the testis until the edema reduces
- Change the dressing daily
- Antibiotic therapy, depending on the degree of necrosis
- Second look if testicle becomes necrotic

Prognosis

- The sooner the surgical intervention for the proper indication the better the prognosis

31.3 Testicular Appendages Torsion

General considerations

- The hydatids are appendices between the testis and epididymis, which are remnants of the obliterated Müllerian ducts
- Differential diagnosis: orchitis, incarcerated inguinal hernia, hydrocele, epididymitis, idiopathic scrotal edema

Signs

- Sudden severe testicular pain
- Swelling of the scrotum
- Pain upon palpation of the scrotum
- Redness of the scrotum
- Blue dot sign

Preoperative work-up

- Examination of the cremasteric reflex, which can be elicited
- Testicular Doppler ultrasonography examination to determine that the testicular parenchyma is vascularized
- If in doubt an operative inspection of the testis is inevitable as prompt clarification differentiating between torsion of the testis and torsion of the appendages gives a far better prognosis

Operation

- Gain access as described above for testicular torsion
- Remove the hydatid
- Make a layered closure of the scrotum

Therapy

- In some cases (symptoms >24–48 h) conservative therapy may be justified
- The diagnosis must be definite, with full vascularization of the testis and a swollen hydatid
- Antibiotics as therapy

31.4 Varicocele

General considerations

- Distention of the veins of the pampiniform plexus
- Affects preferentially the left side, because on this side the testicular vein drains into the renal vein at right angles, while on the opposite side the vein drains into the inferior vena cava
- The varicosity of the plexus raises the temperature in the scrotum, causing testicular damage
- Varicocele develops through insufficiency of the venous valves

Table 31.4 Classification of varicocele

First degree	Palpable veins, increased filling after Valsalva maneuver
Second degree	Varicocele evident without induction
Third degree	Veins apparent in the scrotum ("bag full of worms")

- A neoplastic compression of the testicular venous outflow should be ruled out, particularly in right-sided varicocele

Classification

- Varicocele is classified as first, second or third degree (Table 31.4)

Signs/symptoms

- Aesthetically annoying
- Occasionally painful
- Thrombosis exceedingly rare
- Alteration in fertility in the long run
- Decreased testicular volume on the affected side

Preoperative work-up

- Clinical examination with the patient in a supine position; raise the scrotum in order to empty the varices, then press on the spermatic cord to empty of remaining blood. When the patient stands, the veins fill again
- Ultrasonography of the spermatic cord with Valsalva maneuver
- Ultrasonography of the abdomen to exclude neoplasma
- Ultrasonography of the kidneys to exclude renal vein stenosis
- Phlebography only in exceptional cases

Conventional Bernardi operation

- Extra-abdominal procedure (retroperitoneal approach)
- Make a skin incision medial to the rectus fascia
- Expose the abdominal musculature and split the muscles in the direction of the muscle fibers
- Exposure and medial dislocation of the peritoneal sac and retroperitoneal dissection to reach the testicular vessels
- The testicular artery is accompanied by two veins
- Identify the vessels and resect the veins over a length of about 2 cm
- Leave the artery untouched

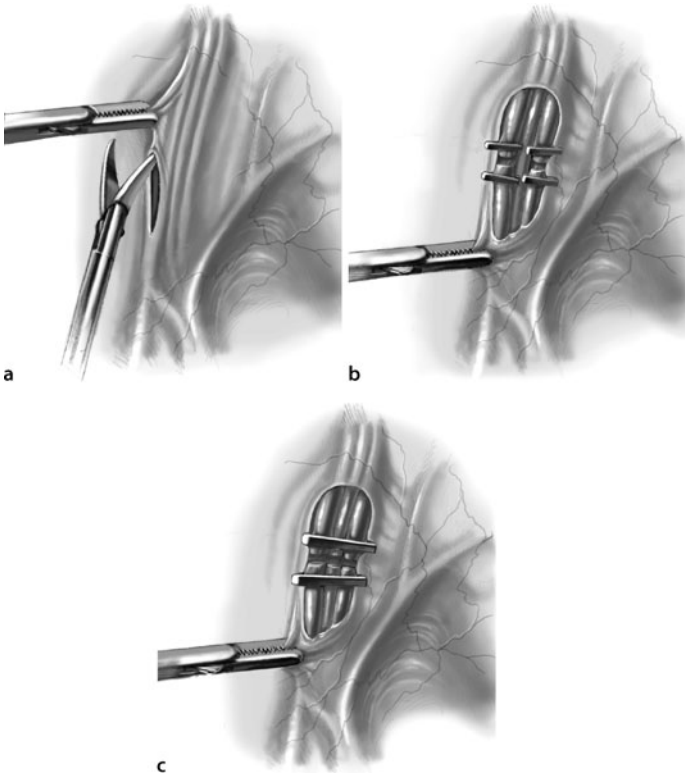


Fig. 31.4a–c Laparoscopic varicocelectomy after Bernardi or Palomo

Conventional Palomo operation

- The operative steps for the Bernardi and Palomo approaches are shown in Fig. 31.4
- Same approach as the Bernadi operation
- Resect all vessels, including the testicular artery, for a length of about 2 cm

Laparoscopic ligature and laser coagulation

- Both operations described above are feasible laparoscopically and retroperitoneoscopically
- In this modification the operation is performed using a transperitoneal procedure, a fact that has no negative effect
- Due to magnification, the vessels can be identified more easily. The veins can be selectively ligated or coagulated with electrocoagulation or laser
- Another advantage is the possibility of selectively coagulating the venous outflow in the iliac vessels, thereby reducing the risk of a relapse

Sclerotherapy

- Angiographic or open injection of substances to induce sclerosis (Fig. 31.5)
- Only perform on adolescent boys, as a Valsalva maneuver is necessary
- Perform the procedure under regional infiltration anesthesia
- If the open approach is chosen, make a small incision over the spermatic cord

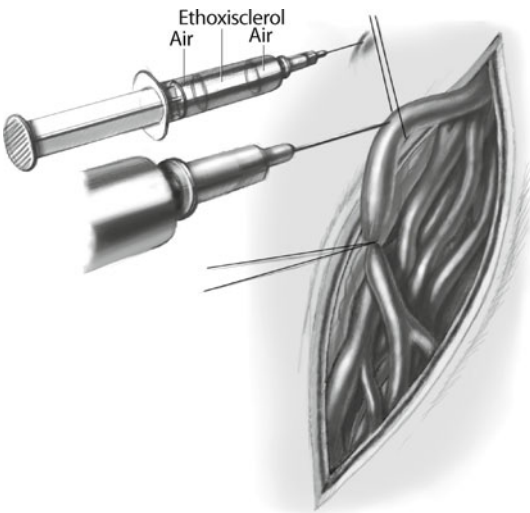


Fig. 31.5 Sclerotherapy by injection of ethoxysclerol

- Isolate the veins of the pampiniform plexus over a distance of 1.5 cm
- Choose one of the veins and ligate it distally, placing a second suture proximally
- Puncture the vein and inject saline. It should flow without any pressure
- Under fluoroscopy, inject contrast medium checking that this vein does not drain into the pelvic veins; if so, ligate this vein and puncture the next
- When the appropriate vein is found ask the patient to perform a Valsalva maneuver as the medium is injected, beginning with 1 ml air and ending with 1 ml air

32 Vagina and External Female Genitalia

32.1 Labial Synechiae

General considerations

- Labial synechiae can develop because of estrogen deficiency in the presence of recurrent inflammation

Signs

- Occasional finding during examination
- Seldom problems in micturition with recurrent infections

Treatment

- Usually no treatment is required, since spontaneous labia separation can be expected
- Application of an estrogen cream can accelerate this process

Operation

- Indicated in cases where conservative therapy has failed
- Split the thin membrane in the medial line
- To keep the labia open use a tampon soaked with estrogen cream

32.2 Hymeneal Atresia

General information

- Hymeneal atresia remains undiscovered until puberty, when menstruation starts
- Differential diagnosis: abscess of the Douglas' pouch, vaginal atresia

Signs

- Hydrometrocolpos or hematometrocolpos (filling with secretions or blood in the vagina and/or the uterus)
- Palpable and painful mass in the lower abdomen
- Protrusion of the hymen outside the vulva
- Problems in micturition (cranial dislocation of the bladder)
- Constipation (dislocation of the rectum)

Preoperative work-up

- Clinical examination
- Abdominal ultrasonography
- Renal ultrasonography
- MRI if necessary

Operation

- Open the hymen
- Seldom, in cases of vaginal malformation: laparotomy with opening of the vagina and perforation of the septum

33 Ovarian Cysts

General considerations

- Ovarian cysts are common especially in adolescent girls
- They correlate with abdominal complaints or are detected incidentally during abdominal imaging
- Occasionally, ovarian cysts and tumors may be considerably large, even weighing over 3–4 kg
- It is important to differentiate correctly between benign and malignant tumors, and non-neoplastic cysts, as each category has specific treatment

Classification

- Follicular cysts are physiological and do not require treatment
- Luteal cysts are physiological and do not require treatment
- Luteal cysts may occasionally reach an abnormally large size and require repeated ultrasonographic follow-up
- Large, simple multi- and unilocular cysts may require treatment if >6 cm in diameter

Signs

- Abdominal mass visible from the outside
- Acute pain in the lower abdomen, especially during activities
- Trouble with micturition (cranial dislocation of the bladder)
- Constipation (dislocation of the rectum)

Preoperative work-up

- Assessment of α-fetoprotein, β-hCG and CA-125 (in adolescents)
- Ultrasonographic examination of the abdomen
- USG-CT-based imaging of the abdominal cavity
- Chest X-ray possibly supplemented with CT if malignancy is suspected
- In some cases endocrine assessment as indicated by the endocrinologist
- Simple ovarian cysts with negative markers, clearly seen on good-quality ultrasonography, do not require CT diagnostics
- Regular ultrasonographic examinations including Doppler ultrasonography to exclude torsion of the ovary seen as:
  - Rapid changes in size (usually a increase)
  - Changes (on Doppler ultrasonography) indicating hypovascularization of the ovary

Treatment

- If tumor markers are negative a more conservative approach can be applied in the sense of a "wait and see" tactic if the cyst size does not exceed 6 cm in diameter, as assessed by regular ultrasonographic examinations
- If tumor markers are positive an interdisciplinary approach with the oncologists is absolutely necessary

Operation

- Signs of torsion are indication for emergency surgery
- If detorsion is successful, the ovary is not necrotic and apparently benign, an ovary-sparing operation can be performed instead of unilateral adnexectomy (excision of the cysts, or even just a fenestration)
- This therapy also applies to large non-neoplastic cysts shown by regular observations to be non-regressing
- Surgery may be performed in the classical way, or laparoscopically
- Consulting an endocrinologist is recommended for the above-listed conditions

34 Kidney and Bladder

34.1 Urinary Tract Infection

General considerations

- Recurrent urinary tract infections are the most common diagnosis made for referrals concerning the genitourinary tract in infancy and childhood
- They are commonly associated with renal tract malformations including vesicoureteral reflux
- The main sign is voiding dysfunction
- All urinary tract infections require further examination

Signs

- Dysuria
- Fever
- Dystrophy (in newborns)
- Failure to thrive (in infants and toddlers)
- Jaundice (in young infants)
- Vomiting
- Pollakisuria
- Hematuria
- Enuresis
- Diarrhea
- Weak urinary stream (in boys)
- Abdominal pain

Diagnostic measures

- Physical examination (palpable kidney, bladder, urinary stream)
- Mid-stream urine culture or suprapubic urine with antibiogram
- Infection parameters
- Laboratory examinations
- Ultrasonography
- Miction cystourethrography (MCU) after the acute infection has receded
- Isotope studies
- Urodynamic examination (cystometry, uroflowmetry):
- Intravenous pyelogram (in selected cases)
- MRI urography (in selected cases)

Therapy strategies

- Further careful investigations to detect the cause of infection are vital
- Primary infection: parenteral antibiotic therapy (guided by urine culture and sensitivity)
- Oral antibiotics after the child has been afebrile for 48 h and maintained for 10–14 days
- Recurrent infection: prophylaxis >3 months, perineal hygiene, treatment of constipation and encopresis
- Voiding program for the infrequent voiders including pelvic training

Complications

- Pyelonephritis
- Segmental renal parenchymal scars
- Reduction of renal function mainly in vesicoureteral reflux

34.2 Vesicoureteral Reflux

General considerations

- Retrograde flow of urine from the bladder into the ureters and kidneys
- Short intramural course of the ureter in the bladder wall with consequent valve mechanism failure (Fig. 34.1)

- Absence of longitudinal muscles in the ureter and of the vesicoureteral valve mechanism (primary reflux)
- Due to meatus stenosis, urethral valves, strictures, neurogenic bladder, etc., a pressure increase leads to bladder distention and consequently impaired function of the vesicoureteral valve mechanism. Additionally, associated anomalies, i.e., ureterocele, para-ureteric diverticulum, ureter duplex, may lead to reflux (secondary reflux)
- Spontaneous resolution of reflux is possible in up to 80% of cases in the first year of life

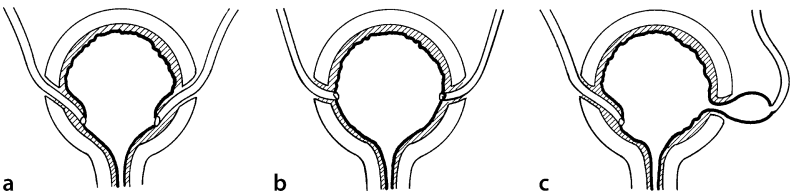


Fig. 34.1a–c Normal anatomy and pathology of the ureter. **a** Normal anatomy with adequate length of the intramural ureter. **b** Pathologic anatomy with short length of the intramural ureter. **c** Hutch diverticulum

Classification

- Vesicoureteral reflux is graded from I to V (Table 34.1, Fig. 34.2)

Table 34.1 Classification of vesicoureteral reflux

Grade	Radiographic correlate
I	Contrast filling of an undilated ureter
II	Contrast filling of the ureter and pelvis without dilatation
III	Contrast filling of the ureter, pelvic and calyceal system with mild dilatation
IV	Contrast filling of the ureter, pelvic and calyceal system with dilatation and atrophy of the renal parenchyma
V	Massive hydronephrosis with tortuosity of the ureters occasionally intrarenal reflux

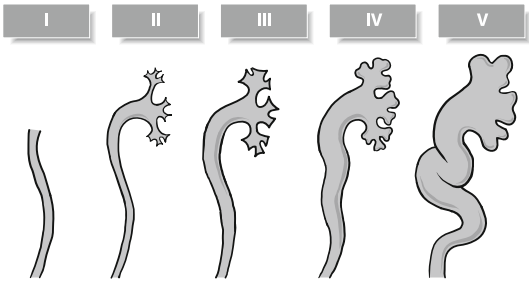


Fig. 34.2 International classification of radiographic grading of vesicoureteral reflux

Signs

- Recurrent urinary tract infection
- Pyelonephritis
- Growth problems
- Enuresis

Preoperative work-up

- Ultrasonographic examination to evaluate the renal size, pelvic and calyceal dilatation, and parenchymal thickness
- Ultrasound to measure the volume of residual urine in the bladder after voiding
- Voiding cystourethrography
- Urine status
- Uroflowmetry
- Bladder manometry (neurovesical dysfunction)
- Isotope ^{99m}Tc-scintigraphy
- Cystoscopy

Indications for intervention

- Grades II–V with relapse after conservative therapy
- Grades IV–V with an open procedure

Conservative therapy

- Prophylactic antibiotic therapy to minimize infection for 12–36 months mainly in cases of recurrent infection
- Continuous follow-up documenting eventual progression (ultrasonography, ^{99m}Tc-DMSA, cystourethrography with UTI) throughout long-term antibiotic prophylaxis
- During this time the vesicoureteral valve function can mature
- Pelvic training

Surgical therapy

- Indicated following unsuccessful conservative therapy over a long time period
- Grade II–V refluxes can be indications for endoscopic treatment or a primary surgical approach
- Contraindicated in patients with renal insufficiency and infravesicular urine retention

Operation: Cohen procedure

- The operative steps are shown in Fig. 34.3
- Pfannenstiel skin incision
- Dissect the bladder and open in the midline
- Expose the trigonum and the ostium
- Catheterize the ureter
- Make a circumscriptive incision around the ostium and make an intramural dissection of the ureter
- Establish a transverse submucosal tunnel to the contralateral side of the trigonum
- Pull the ureter through the trigonum and reimplant it on the contralateral side
- Site a urethral catheter and close the bladder

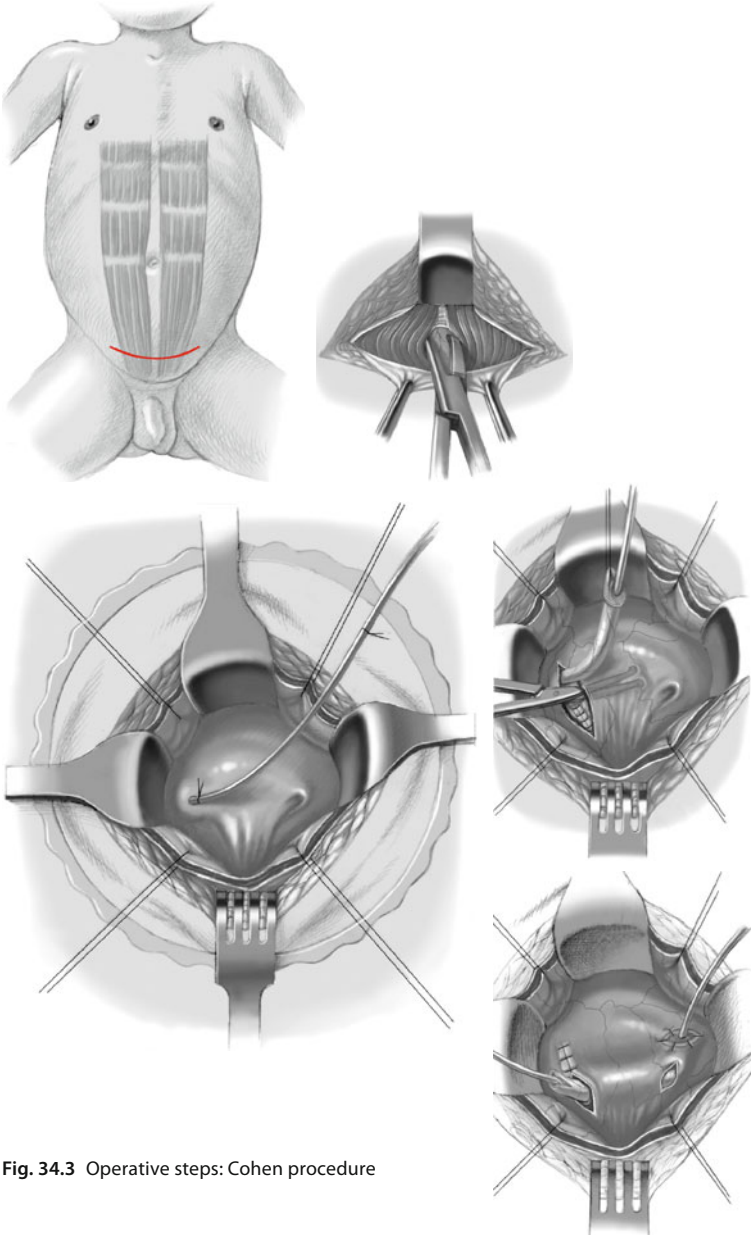


Fig. 34.3 Operative steps: Cohen procedure

Operation: Politano–Leadbetter procedure

- Make a Pfannenstiel skin incision
- Dissect the bladder and open in the midline
- Dissect the ureter and pull it out through the bladder wall
- Create a new bladder opening several centimeters above the original ostium
- Form a long vertical submucosal tunnel, from the new opening to the original ostium
- Pull the ureter through the new opening in the tunnel down to the original ostium
- Reimplant the ureter in its original position ipsilaterally

Operation: Lich–Gregoir procedure

- The bladder is not opened during this procedure
- Dissect the ureter at the entrance point to the bladder
- Dissect the bladder muscles (detrusor) without opening the bladder mucosa in order to create a tunnel facilitating the ureter
- Then suture the dissected bladder muscles above the ureter, creating a tunnel
- This lengthens the distance between the entry point and the ostium

Postoperative care

- Antibiotics for a period of 3 months (sterile urine)
- Transurethral or suprapubic catheter is removed on day 3–7
- Remove the ureteral stent on days 7–10

Endoscopic procedure

- Insert a sharp-edged needle beneath the ureteral orifice under endoscopic control (Fig. 34.4)
- Inject a compound (e.g., Teflon, collagen, Macroplastique, Deflux) to produce a depot
- This depot increases the intramural ureter length at the bladder entrance and changes the direction of the ostium
- The lumen of the ostium is narrowed

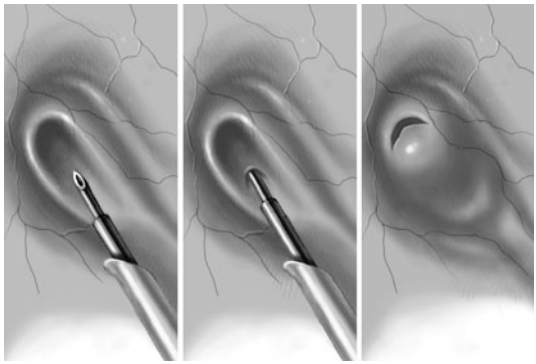


Fig. 34.4 Endoscopic treatment of vesicoureteral reflux

Prognosis

- Reflux can be treated successfully conservatively, endoscopically or surgically, but the kidney damage may progress

34.3 Ureterovesical Junction Obstruction/Megaureter

General considerations

- Narrowing of the ureter at the level of the bladder junction results in urine retention and the formation of a megaureter and hydronephrosis
- This narrowing can be either structural (fibrosis) or functional (congenitally incompetent uretero-vesical function)
- Secondary uretero-vesical obstruction may be caused by posterior urethral valves, tumor, inflammation, ureterocele or be as a consequence of surgical procedures
- Primary megaureter without obstruction and a low-grade vesicoureteral reflux normally resolve spontaneously

Signs

- Recurrent urinary infection with the associated symptoms

Preoperative work-up

- Urine bacteriology
- Ultrasonography (hydroureter and hydronephrosis)
- Voiding cystourethrography – to exclude reflux and primary bladder pathology
- Isotope (DMSA and MAG3) studies: to investigate renal function and to support the presence or absence of obstruction
- MRI urography
- Cystoscopy – in selected cases

Conservative therapy

- Cystoscopy
- Temporary splitting of the dilated ureter with a double pig-tail catheter until the megaureter recovers
- Antibiotic therapy
- Spontaneous resolution of the obstruction may occur

Surgical therapy

- Recurrent infections and severe ostium stenosis with functional or organic obstruction are indications for operation
- Uretero-vesical junction obstruction with severe loss of renal function is an indication for:
  - Percutaneous transrenal drainage
  - Temporary uretero-cutaneostomy
  - Nephroureterectomy in cases of a functionless kidney

Operation: cutaneous ureterostomy

- In the first year of life a cutaneous ureterostomy may lead to recovery of the ureter and the kidney enabling successful reconstructive surgery later
- Extravesical dissection of the ureter
- See Sect. 34.6

Operation: remodeling

- Dissect the obstructing megaureter from the insertion into the bladder either by opening the bladder or using the extravesical approach
- Excise the obstructing segment
- Reimplanting the dilated megaureter may lead to technical problems
- Tapering the distal part of the ureter along its length reduces the size of the lumen and allows easier reimplantation
- Reimplant the ureter applying either the Cohen or the Politano–Leadbetter procedure
- See Sect. 34.2

Complications

- Vesicoureteral reflux in cases of insufficient tunnel length
- Obstruction due to excessive tapering or ischemic contracture

Postoperative care

- Measure urine output and blood pressure
- Ultrasonography to detect obstruction
- Voiding cystourethrography after 6 months
- Isotope studies of kidney function and to detect obstruction

Prognosis

- Depends on the severity of the megaureter and extent of renal tissue damage
- In 90% of cases with appropriately timed operative treatment the results are satisfactory

34.4 Ureteric Duplication

General considerations

- Frequent malformation of the urinary tract with an incidence of 8 in 1000 children
- 40% are bilateral
- 60%–70% in females

- Findings are often incidental
- The upper pole ureter enters the bladder adjacent or distal to the lower pole ureter and is often infravesical (Weigert–Meyer law)
- In cases of partial duplication, the urine can pass down one limb of the duplication and then up the other side of the Y (yo-yo phenomenon)
- There is an increased incidence of infection due to both reflux and obstruction
- Reflux in the lower pole ureter is common; however, reflux may occur in the upper pole ureter or in both ureters

Classification

- The classification of ureteric duplication is given in Table 34.2 and Fig. 34.5
- Both ectopic ureter and ureteroceles are frequently associated with duplicated upper urinary tracts

Table 34.2 Classification of ureteric duplication

▪ Complete ureteral duplication	▪ Both ureters drain independently into the bladder
▪ Partial ureteral duplication (ureter fissus)	▪ The two ureters join proximally to a common insertion into the bladder
▪ Intramural connection of the ureters	▪ Both ureters join within the bladder wall and are drained with one ostium

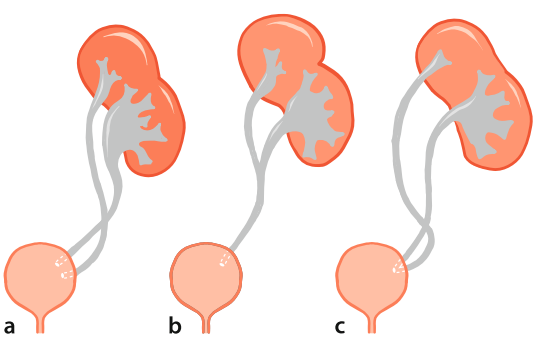


Fig. 34.5a–c Duplicated ureter systems. **a** Crossing of ureters with separate orifices. **b** Ureter fissus. **c** Intramural unification of ureters with conjoint orifice

Signs

- Recurrent urinary infection
- Abdominal pain
- Pyelonephritis
- Enuresis

Preoperative work-up

- Urine culture
- Ultrasonography
- MRI urography
- Voiding cystourethrography
- Cystoscopy (number of ostia, the upper moiety ureter always terminates distal to the lower moiety ureter, ureterocele)
- Isotope study (scintigraphy – DMSA)
- Intravenous pyelography in selected cases

Indication for intervention

- Reflux: following unsuccessful conservative therapy
- Obstruction
- Ureterocele
- Ectopic ureter (bladder neck, urethra, vagina, vestibule)
- Yo-yo phenomenon leading to stasis and ureter dilatation
- Non-functioning upper moiety with recurrent UTI

Operation

- The operative steps are illustrated in Fig. 34.6
- Complete duplication: distal and proximal ureters share a common blood supply therefore reimplantation involves mobilization and reimplantation of the common sheathes, as in reflux surgery
- Partial duplication: en-bloc reimplantation as a single ureter
- Ureterocele: endoscopic incision and later antireflux surgery or heminephro-ureterectomy if necessary, or primary heminephroureterectomy without opening the bladder
- Obstruction: mobilization, resection and reimplantation of the distal end of the common sheath in an antireflux fashion

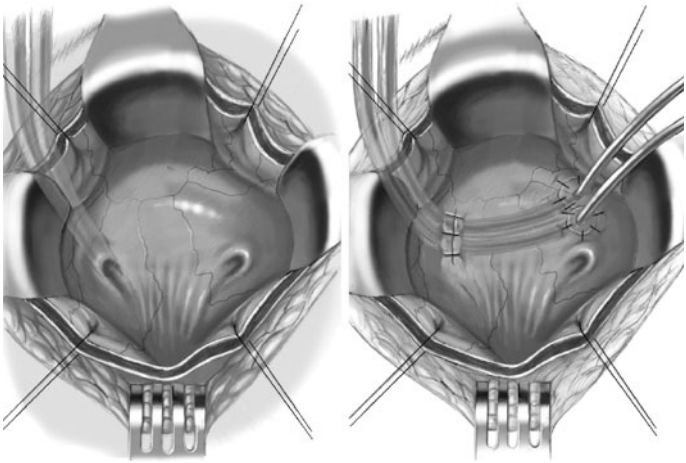


Fig. 34.6 Operative steps: ureteric duplication

Operation: heminephroureterectomy

- The operative steps are illustrated in Fig. 34.7
- Make a small transverse flank incision
- Expose the kidney with the dysplastic upper pole and the double ureter
- Dissect the renal hilus and expose the vessels and ureters
- Ligate the artery supplying the upper pole
- Resect the upper renal pole with the proximal part of its ureter
- Suture the parenchyma and capsule
- Make a separate small inguinal incision to remove the distal part of the double ureter without opening the bladder
- This operation is also feasible by a transperitoneal laparoscopic or retroperitoneal minimally invasive approach

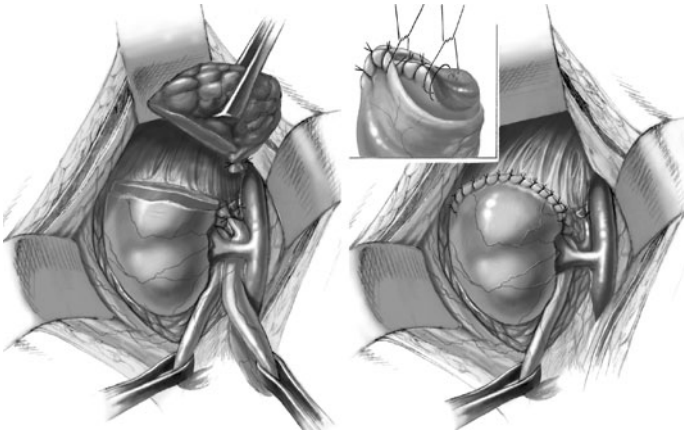


Fig. 34.7 Operative steps: heminephroureterectomy

Postoperative

- The upper pole bed should be drained
- Bladder catheter is recommended for a few days

Prognosis

- Depends largely on the type of malformation and the degree of renal damage

34.5 Ureteropelvic Junction Obstruction/Hydronephrosis

General considerations

- Inadequate drainage of urine from the renal pelvis into the upper ureter, resulting in hydrostatic distention of the renal pelvis and calyces
- Increased intrarenal urine stasis and pressure inducing repeated infections and finally resulting in progressive kidney damage

- Most cases of ureteropelvic junction obstruction are probably caused by failure of recanalization
- Other possible causes are aberrant vessels, high insertion of ureter, ureteral valves, polyps, kinking of the ureter and stones
- Incidence 1:1000–2000 live births
- Male predominance (ratio 2:1)
- A dilated renal pelvis may resolve without surgical intervention, especially in early infancy

Signs

- Most hydronephrotic kidneys are detected antenatally
- Palpable mass in the abdomen as a result of hydronephrosis
- Hematuria (after minor trauma or exercise)
- Urinary tract infection
- Renal hypertension
- Cyclic or acute abdominal pain associated with nausea and vomiting
- Many of these children are initially seen by a gastroenterologist
- Associated malformations [VATER/VACTERL syndrome (syndrome with a wide range of anatomical births defects, i.e., vertebral, anal, cardiac, tracheo-esophageal, renal and limb abnormalities), multicystic kidneys, reflux]
- Nephrolithiasis (long-standing obstruction and infection)

Preoperative work-up

- Antenatal and postnatal ultrasonographic examination
- Urine examination (including urine culture)
- 24-h creatinine clearance
- Isotope studies (diuretic renogram DTPA or MAG3) to distinguish between obstructing and nonobstructing hydronephrosis
- Voiding cystourethrography (rule out vesicoureteral reflux and subvesical obstruction)
- Intravenous pyelography (only in selected cases)

Indication for intervention

- Decreased function on renography of the hydronephrotic kidney with fixed obstruction
- Thinning of renal parenchyma due to increasing dilatation of the calyces
- Recurrent urinary tract infections
- Repeated hematuria
- Percutaneous drainage for sepsis is required preoperatively
- Nephrectomy is rarely indicated (dysplastic kidney, nonfunctioning hydronephrotic kidney) mainly in infants

Operation: pyeloplasty

- Anterior extraperitoneal (or posterior lumbotomy) approach is preferred
- Make a transverse incision from the edge of the rectus to the top of the 12th rib
- Enter the retroperitoneum through an incision that splits the muscle
- Kidney may be left in situ
- Mobilize the renal pelvis and upper ureter
- Divide the ureter obliquely just below the obstructive segment
- Resect the narrow pyeloureteric junction
- Taper the renal pelvis, if necessary
- Form an anastomosis between the incised ureter and pelvis
- Implant a transrenal nephrostomy tube or a ureteric stent, in some cases both
- A minimally invasive approach, either transabdominally or retroperitoneally, is feasible
- The operative steps are illustrated in Fig. 34.8

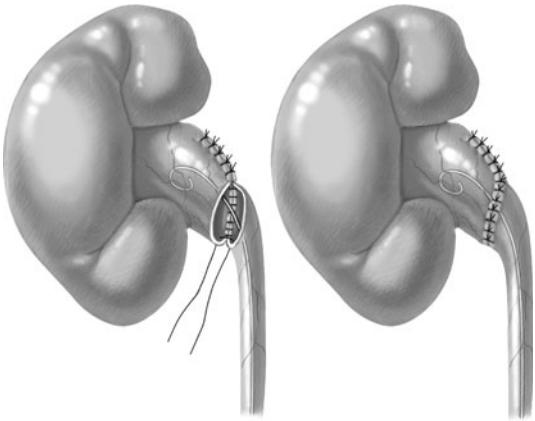


Fig. 34.8 Operative steps: pyeloplasty

Postoperative care

- Perirenal drainage for 4–7 days
- Nephrostomy drainage removal on the postoperative day 3–7
- Regular follow-up (ultrasonography, urine analysis)
- Antibiotics (in case of urinary infections)

Prognosis

- Operative success >90%

34.6 Cutaneous Ureterostomy

General considerations

- Indications for temporary or permanent cutaneous ureterostomy have been significantly reduced by the advancement of percutaneous and endoscopic techniques, lessening the need for cutaneous upper tract diversion
- An indication for ureterostomy is the need to decompress a massively dilated upper tract in neonates or infants
  - These patients are not candidates for reconstruction, because of their size or presence of urinary tract infection

  - It remains unclear whether the involved kidney functions adequately
  - Wait for functional bladder development
- Controversy exists with respect to the benefit of cutaneous ureterostomy in cases of posterior urethral valves in those patients who have persistent severe hydronephrosis, hydroureter and azotemia despite ablation of the valves or diversion by vesicostomy

Operation: proximal ureterostomy

- Make a subcostal incision
- Perform retroperitoneal dissection and expose the dilated ureter
- Dissect a ureter sling and displace it to the cutaneous level
- Pull a skin bridge through the sling and anchor it
- Suture the ureter to the skin
- Alternatively, the ureter can be divided and diverted as a single limb stoma. The distal limb in this case can be anastomosed end-to-side to the proximal limb

Operation: distal ureterostomy

- Empty the bladder using a transurethral catheter
- Make a low transverse muscle-splitting incision that allows access to the retroperitoneum
- Carefully mobilize the ureter to the level of the bladder and transect
- Preserve the periadvential and retroperitoneal blood supply of the ureter in order to avoid stoma stenosis
- Bring the ureter out either in the lateral margin of the incision or in the midline
- A V-shaped skin flap can be used to prevent stoma stenosis

Operation: Sober ureterostomy

- The main urine stream flows from the renal pelvis to the skin in the proximal limb, bypassing the anastomosis of the distal limb to the bladder
- Mobilize the ureter

- Divide the ureteric loop at a level that enables the proximal limb to comfortably lie at skin level
- Anastomose the distal limb to a corresponding incision in the proximal limb
- Suture the end of the proximal limb to the skin as a stoma

Operation: reversed Sober ureterostomy

- The main urine stream flows from the renal pelvis to the bladder through the end-to-side anastomosis of the proximal to the distal limb, bypassing the part of the distal limb that leads to the skin
- Mobilize the ureter
- Divide the ureteric loop at a level that enables the distal limb to comfortably lie at skin level
- Anastomose the proximal limb end-to-side to a corresponding incision in the distal ureter limb
- Suture the distal limb to the skin as a stoma

Postoperative care

- Strict electrolyte and fluid balance
- Antibiotics
- Remove catheter when the stoma has healed

Prognosis

- Permanent ureterostomy has a tendency to stenose and irritate the skin
- Stenosis facilitates stone formation

34.7 Posterior Urethral Valves

General considerations

- Most common life-threatening anomaly of the external genitalia in males
- Severe forms may result in oligohydramnios and are diagnosed as Potter's syndrome due to the oligohydramnios

- Frequent cause of urine retention in children
- Prestenotic dilatation of the urethra
- Consequences depend on the degree of obstruction
  - Renal dysplasia
  - Hydronephrosis
  - Hydroureter
  - Hypertrophy of the bladder muscle
  - Vesicoureteral reflux
- Differential diagnosis
  - Neurogenic bladder
  - Subvesical strictures
  - Prune-belly syndrome
- Exclude congenital urethral valves in cases of bilateral renal pathology (i.e., hydronephrosis)

Signs

- Sings are often manifested in the neonatal period or in the first 3 months of life
- Dribbling of urine
- Weak urine stream
- Enlarged bladder
- Straining on urination
- Vomiting
- Failure to thrive
- Urinary tract infection
- Enuresis in older children

Preoperative work-up

- Antenatal ultrasonography
- Urgent bladder decompression by transurethral catheterization if possible, or suprapubic catheter
- Postnatal ultrasonography
- Laboratory studies (hyperkalemia with metabolic acidosis, azotemia)
- Urine culture

- Antibiotic therapy
- Voiding cystourethrography (lateral view!)
- Urethrocystoscopy
- Intravenous pyelography (only in selected cases)

Therapy

- In small neonates transurethral, if possible, or suprapubic bladder catheter
- Change the catheter every third day in order to induce dilatation until it is possible to insert a cystoscope with a working channel for the resection
- Operation
- Cystoscopic resection of the valves (Fig. 34.9) using
  - Laser resection (bare fiber)
  - Valve fulguration
  - Cold knife
- Vesicostomy if the urethra is too small to allow endoscopic therapy
- Cutaneous ureterostomy in cases of severe ureteral dilatation and uretero-vesical junction obstruction (see Sect. 34.6)

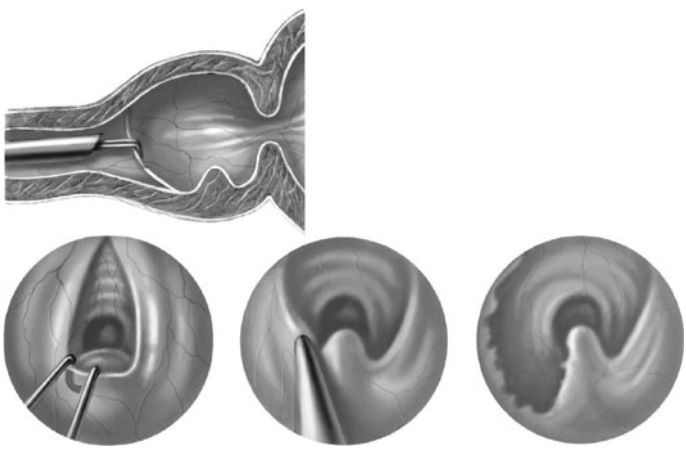


Fig. 34.9 Endoscopic resection of urethral valves

Postoperative care

- Bladder catheter for 1–2 weeks
- Antibiotic prophylaxis for 1 month (in cases of vesicoureteral reflux, 6 months)
- Sufficient fluid management in cases where the kidneys are seriously impaired
- Follow-up using voiding cystourethrography after 6 months
- If there is doubt about the remaining valves, a second cystoscopy is indicated

Prognosis

- Depends on the level of renal insufficiency
  - Better in cases with postoperative creatinine <1 $mg \cdot dl^{-1}$, worse if creatinine is >1 $mg \cdot dl^{-1}$
- Urinary tract infections
- In cases of urinary incontinence, ureteral neoimplantation and/or bladder augmentation may be required
- Kidney transplantation in severe cases

34.8 Bladder Exstrophy

General considerations

- The bladder lies open in the lower abdomen, with defects of the genitalia, rectus muscles, and separation of the pubic bones with a widened symphysis as well as an open bladder neck and urethra
- The bladder epithelium is normal at birth but liable to inflammatory changes with metaplasia and fibrosis later
- The umbilical cord inserts directly above the bladder
- Inguinal hernias commonly coexist
- Urinary incontinence is a part of this anomaly
- In males the epispadiac penis is cranially displaced as well as markedly shortened due to wide separation of the crural attachments and the prominent dorsal chordee

- In females, the clitoris is bifid, with separated labia
- Incidence: 1:30,000–40,000 live birth
- Males predominate, ratio 4:1
- Associated malformations
  - Cardiac anomalies
  - Omphalocele
  - Spinal cord malformations
- The correction should take place in centers with surgeons who are experienced in the complex management of this deformity

Preoperative work-up

- Cover the defect with a moist plastic sheet not using any gauze, which could lead to epithelium swelling making surgery difficult
- Ultrasonography to exclude other renal malformations
- Intravenous urography, if anomalies of the upper renal tract are suspected
- It is important to explain the malformation to the parents and involve them in the decision-making process

Surgical intervention

- Bladder exstrophy correction usually requires staged reconstructive procedures
  - Bladder closure is possible in the first 48 h of life (first stage)
    - Primary bladder closure
    - Repair the abdominal wall defect
    - Bladder neck reconstruction
  - Penile reconstruction (second stage)
    - Epispadias repair
- Bladder closure in the first few days can be achieved without osteotomy
- Later an osteotomy (posterior or anterior) makes the primary result look better, however the long-term results do not show any difference as the symphysis parts again later
- Epispadias correction (with penile reconstruction) at the age of 15–24 months

- Ureter implantation to prevent reflux, if necessary
- In those cases where normal urination and continence are not achieved, bladder augmentation and intermittent clean catheterization may be necessary

Operation: bladder closure

- The operative steps are illustrated in Fig. 34.10
- Catheterize both ureteral ostia
- Make a skin incision on the exstrophic bladder border, continuing to the penis and around the umbilicus
- Mobilize the bladder and identify the abdominal rectus muscle
- Identify the bladder neck and the prostatic part of the urethra
- Further dissect the corpora cavernosa and the displaced symphysis
- Place a catheter along the urethra and close the bladder in a cranio-caudal technique with drainage of the ureters
- Mobilize the connective tissue behind the symphysis in order to facilitate closure of the bladder neck
- Approximation of the symphysis with strong sutures or external fixation (with or without osteotomy)
- Skin sutures

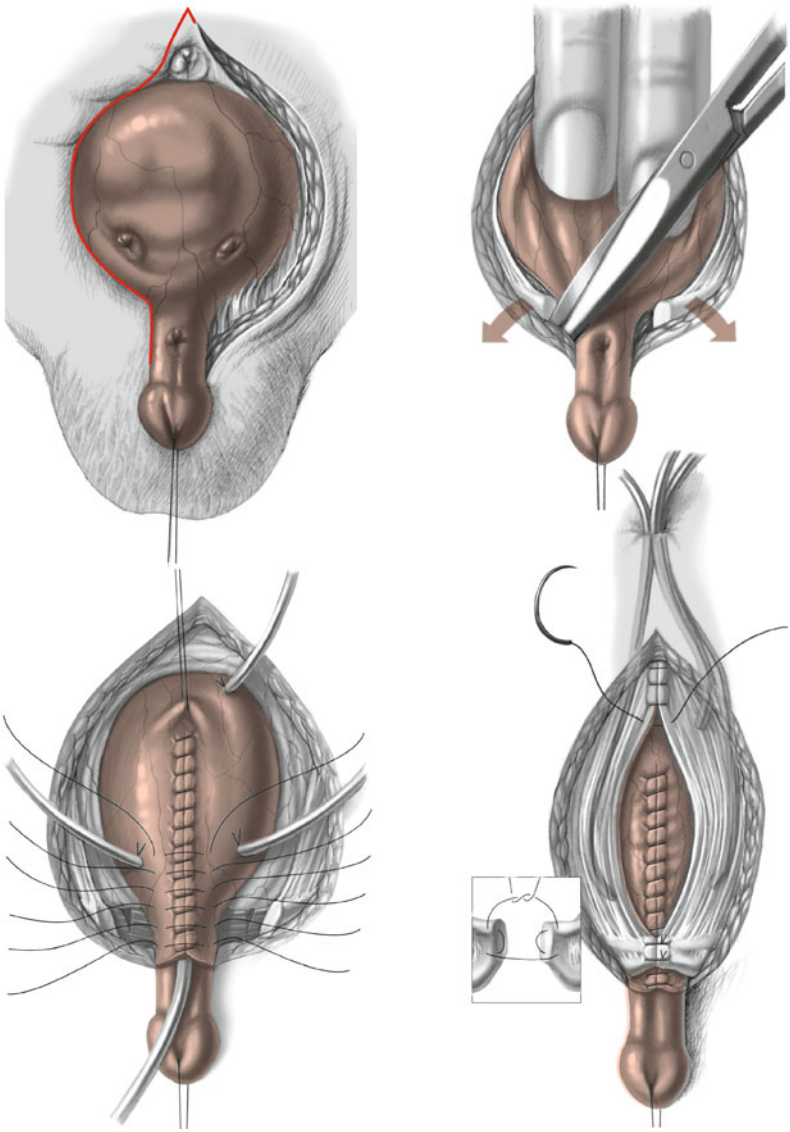


Fig. 34.10 Operative steps: primary bladder exstrophy repair

Epispadias correction

- Incision in the area of the urethra, along the length of the urethra on either side and around the penis
- Meatotomy – small longitudinal incision and transverse sutures
- Identify the corpora cavernosa and mobilize the ventral penile skin
- Identify and protect the neurovascular bundle around the corpora cavernosa
- Tubularize the urethral plate around a catheter
- Carry out chordectomy and adaptation of the corpora cavernosa over the urethra
- Close the glans
- Mobilize a vascular ventral prepuce flap, which is translocated to the dorsal penile area
- The operative steps are illustrated in Fig. 34.11

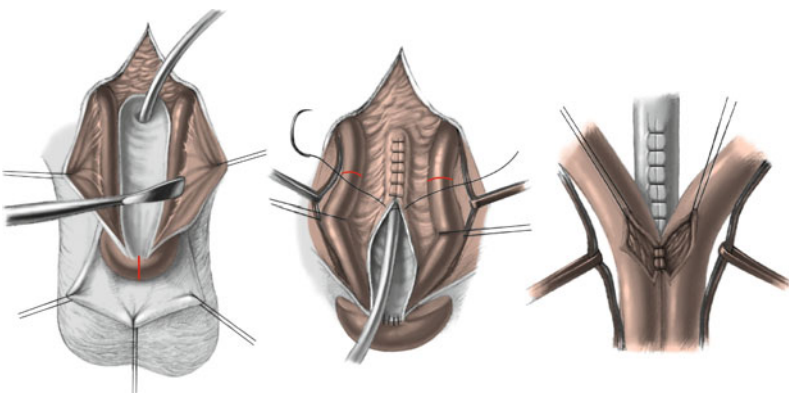


Fig. 34.11 Operative steps: epispadias repair

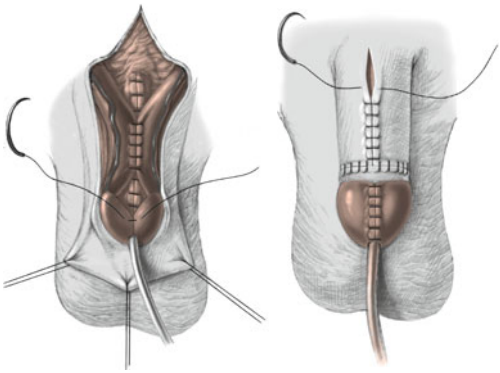


Fig. 34.11 *(continued)* Operative steps: epispadias repair

Postoperative care

- Antibiotics to prevent urinary infection
- Bandage application around the hips with inner rotation
- Ureteral stents for 2 weeks
- Drainage of the bladder to prevent bladder outlet obstruction

Prognosis

- Variable depending on the severity of the defect
- Parent cooperation and understanding are required to obtain optimal results
- Incidence of incontinence varies considerably
- Further surgical interventions (reflux, stone, hernia, urethral lengthening, bladder augmentation, continent abdominal stoma) are frequently necessary
- Hip problems occur often later in life

34.9 Bladder Augmentation

General considerations

- Bladder augmentation aims at
  - A compliant bladder reservoir with low intravesical pressure
  - Limiting bladder contractility
  - Increasing bladder capacity
- Most commonly bladder augmentation is performed utilizing the ileum

Preoperative work-up

- Clear fluid diet 2 days before surgery
- Bowel preparation with GoLitely
- Antibiotics just before surgery and then as therapy

Operation

- The operative steps are illustrated in Fig. 34.12
- Make a lower midline incision
- Bladder dissection
- Maximally expose the bladder including dissection down to the ureters on each side
- Incise and open the bladder
- Intubate the ureters and the bladder neck
- Measure the circumference of the rim on the open bladder (use a catheter along the rim)
- Bowel preparation
  - Isolate an ileal segment (usually about 20–30 cm) about 15 cm from the ileocecal valve
  - The length is equal to the open bladder circumference
  - Detubularize the ileal segment by opening it antimesenterially
- Augmentation
  - Suture the detubularized ileum along the free rims of the bladder
  - Consider creating a Mitrofanoff stoma (see Sect. 34.10)

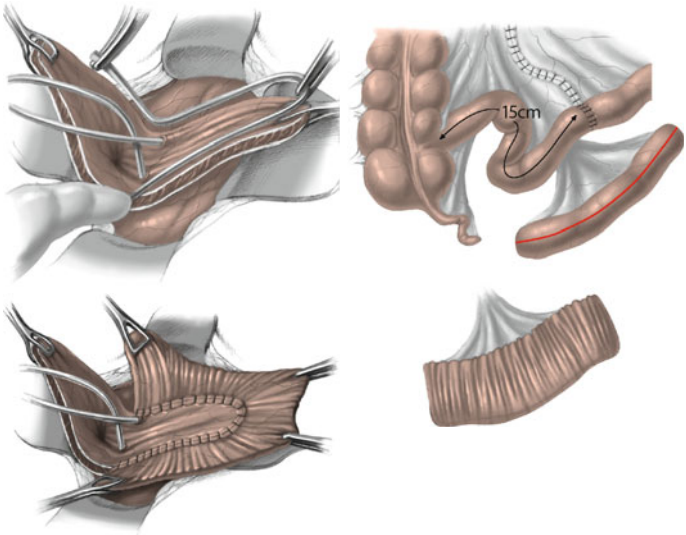


Fig. 34.12 Operative steps: bladder augmentation

Postoperative care

- The stent in the bladder remains in place for at least 10 days
- Antibiotics as a therapy
- Bladder training
- Clean intermittent catheterization (CIC) of the bladder

34.10 Mitrofanoff Procedure

General considerations

- Despite bladder augmentation to increase bladder capacity, clean intermittent self-catheterization has become a well established procedure that ensures effective bladder emptying
- Especially convenient for wheelchair patients and patients with a sensitive urethra (i.e., male exstrophic bladder)

Operation

- This operation is usually performed during bladder augmentation
- Isolate the appendix with a vascular pedicle and a small cuff of cecum
- Open the tip of the appendix
- Before suturing the bladder and the ileum together, implant the open tip of the appendix through the bladder wall or the ileum used for augmentation in an oblique submucosal tunnel 2.5–3 cm long, thus preventing reflux
- Close the bladder
- Draw the base of the appendix to the skin through a hole, preferably next to the umbilicus for best cosmetic results
- Take care to prevent kinking of the appendix and its vascular pedicle
- Spatulate the cutaneous end of the appendix in order to facilitate a triangular skin flap, preventing delayed stenosis
- Pass a catheter through the appendix and leave it in place for at least 2 weeks
- The operative steps are illustrated in Fig. 34.13

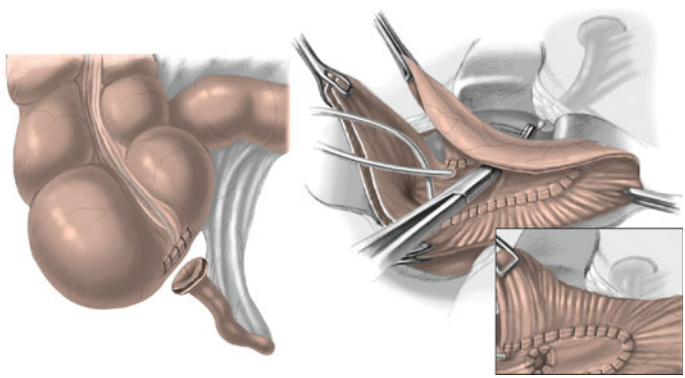


Fig. 34.13 Operative steps: Mitrovanoff procedure

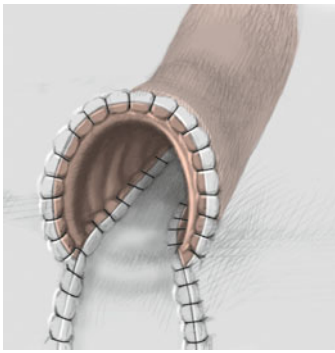


Fig. 34.13 *(continued)* Operative steps: Mitrovanoff procedure

34.11 Intersex

General considerations

- The genes on the X- and Y-chromosomes (karyotypes) are responsible for sex organ development in the embryo. The development of the Wolffian duct in males and of the Müllerian ducts in females is responsible for sex differentiation, such that as one sex form develops, the other form is suppressed
- The gonads produce the hormones required for the development of the inner and outer genitalia and determine the phenotype of an individual
- Disruption of these mechanisms is responsible for intersex-related developmental anomalies
- Intersex is a condition in which the patient has both male and female characteristics. For clinical purposes, the intersex disorders are divided into four main groups, based on gonadal histology
  - Female pseudohermaphroditism: two histologically normal ovaries are present. The karyotype is 46, XX, and the external genitalia show a variable degree of virilization. By far the most common example of this group is congenital adrenal hyperplasia
  - True hermaphroditism: both ovarian and testicular tissues are found in the same patient. The karyotype and appearance of the external genitalia are variable

  - Male pseudohermaphroditism: two histologically normal testes are present, karyotype is invariably 46, XY, and the external genitalia show either partial or complete failure of masculinization. The most frequently encountered of these disorders are various syndromes of androgen insensivity – testicular feminization
  - Gonadal dysgenesis: the gonads are histologically disordered, often being replaced partially or completely by fibrous stroma. The appearance of the external genitalia and karyotype varies, but the latter frequently shows mosaicism, with XY and XO lines
- Incidence: not uncommon
- Differential diagnosis: cloacal exstrophy, urogenital sinus anomalies, micropenis, absence of penis, bifid scrotum, scrotal and perineal hypospadias
- The sex of the child should be determined as soon as possible after birth, however after considering many parameters (length of the phallus, testis, vagina, etc.) and the wishes of the parents
- Feminizing or masculinizing genitoplasties can be carried out from the age of 3 months, and should be completed as early as possible
- Secondary procedures in older children should take in account the previously determined sex of the child

Differential diagnosis

- The differential diagnosis of intersex abnormalities is given in Table 34.3

Table 34.3 The differential diagnosis of intersex abnormalities

	Female pseudo hermaphroditism	Male pseudo hermaphroditism	True hermaphroditism	Mixed gonadal dysgenesis
Buccal smear	Positive	Negative	Positive	Negative
Karyotype	XX	XY	XX	XY
Gonads	Normal ovaries	Testes	Testis or ovary Ovotestis	Dysgenetic and streak ovaries
Steroids in urine	Positive	Negative	Positive	Negative

Classification

- The Prader classification of intersex abnormalities is shown in Table 34.4 and Fig. 34.14

Table 34.4 The Prader classification of intersex abnormalities

Type	Description
I	Clitoris hypertrophy in the presence of normal female genitalia
II	Clitoris hypertrophy, urogenital sinus, vaginal and urethral with common opening
III	Clitoris hypertrophy, narrow and deep seated urogenital sinus, vaginal and urethral opening
IV	Phallus with small urogenital opening
V	Male genitalia

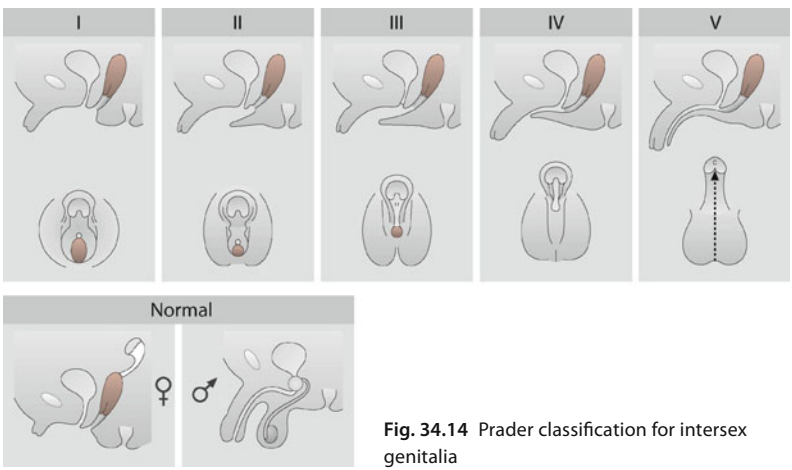


Fig. 34.14 Prader classification for intersex genitalia

Preoperative work-up

- History (hormones consumed during pregnancy, placental insufficiency, siblings with adrenogenital syndrome)
- Physical examination (gonadal symmetry, phallus size and shape, vaginal evaluation, rectal examination, palpating the uterus)
- Buccal smear for Barr bodies
- Karyotype examination
- Ultrasonography
- MRI
- Serum electrolytes
- Retrograde genitogram and urethroscopy
- Diagnostic laparoscopy/laparotomy (internal genitalia, histology)
- Extensive consultation with the parents when deciding the gender

Parameters for determining the sex of the neonate

- Determined gender female
  - If the phallus is smaller than 1.5×0.7 cm, normal development of the penis cannot be expected
  - Patients with male hermaphroditism, true hermaphroditism and female hermaphroditism (clitoris reduction plasty and vaginal reconstruction)
- Determined gender male
  - If the infant responds well to androgen therapy and an adequate length of the phallus is achieved after therapy

Principles for the surgical therapy

- Based on the principle that phenotype and genotype need not necessarily be coordinated
- The genitalia should be corrected in a way that will provide functional sexuality
- The fact that it is easier to make a vagina than a functional penis of sufficient dimensions should represent guidelines for treatment
- The surgical approach can be: feminizing or masculinizing and sex reassignment surgery, accompanied by hormonal treatment

- The timing of the surgical procedure represents a balance between the psychological advantages of early surgery and the technical limitations imposed by the small size of the structures. The trend is clearly toward earlier reconstruction
- Ambiguous genitalia constitute an important group of disorders that are not uncommon
- It is imperative that the precise cause should be identified as quickly as possible so that the appropriate sex of rearing can be assigned

Operation: male gender

- The first step involves a chordectomy and straightening of the phallus
- Correct the accompanying hypospadias
- Correct the position of the penis in relation to the scrotum. Place the scrotum below the level of the penis, since in most males with hermaphroditism it is sited above the level of the penis
- Remove the remnants of the Müllerian ducts (uterus, vagina, and ovaries)

Operation: female gender

- Many different procedures exist, however they have a single common purpose
  - To remove the testis
  - Clitoris reduction plasty
  - Introitus and vaginal reconstruction
- Total urogenital mobilization (TUM)

Postoperative care

- Antibiotic therapy
- Urethral catheter for 6–10 days (in feminizing genitoplasty, dilatation of the enlarged introitus)

Prognosis

- The results correlate with the experience of the operating team
- The patients do not always accept the sex assigned to them
- Long-term psychological management of the children and parents

34.12 Renal Malformations

General considerations

- Agenesis: absence of both (very rare) or one kidney. In the first case, the children die or develop Potter's syndrome; in the second case the prognosis is good
- Hypoplasia: renal insufficiency, if bilateral
- Polycystic kidney: bilateral autosomal-recessive disease usually with lethal outcome
- Multicystic (dysplastic) kidney: generally, unilateral malformation with cysts of various sizes held together by fibrous tissue. Normal renal tissue and reniform shape are absent, the ureter is frequently atretic
- Simple renal cysts: solitary or multiple isolated cysts that are mostly asymptomatic. Incidence of simple cysts increases with age
- Renal dystopia is failure of the normal ascent of the kidney to the lumbal region with rotation of the pyelon towards the midline
  - Pelvic dystopia with practically no ascent of the kidneys
  - Crossed renal ectopy with ascent of both kidneys on one side
  - Horseshoe kidney with joint caudal renal poles

Signs

- Parenchyma anomalies (agenesis, hypoplasia, cysts)
  - Different grades of renal insufficiency
- Renal dystopia
  - Palpable tumor in the abdomen
  - Nephrolithiasis especially in cases of obstruction
  - Urinary tract infections
  - Impaired urodynamics
  - Dystopic kidneys may be functionally insufficient

Preoperative work-up

- Urine examination
- Ultrasonography
- MRI

- Isotope scintigraphy
- Intravenous pyelography in selected cases

Operation

- Cystic renal diseases
  - Multicystic (dysplastic) kidney: nephrectomy if no resolution
  - Simple renal cyst: percutaneous puncture and injection of sclerosant into the cyst or cyst extirpation
  - Polycystic kidney: transplantation in cases of severe renal failure
- Renal dystopia
  - Signs relate to the pathology
    - Obstruction
    - Vesicoureteral reflux
    - Stone formation
    - Malignancy
- Severe functional and parenchymal reduction – nephrectomy

Prognosis

- Multicystic kidney has a good prognosis
- Solitary renal cysts have a good prognosis
- Polycystic (dysplastic) kidneys have a poor prognosis
- Dystopic kidneys have a good prognosis

34.13 Nephrolithiasis

General considerations

- Most kidney stones in childhood are associated with infection
- The most frequent pathogens are *Proteus* spp. and *Escherichia coli*
- Boys aged between 2 and 3 years are the most frequently affected
- Stones due to metabolic disease in children are not rare (enzyme disorders, renal tubular syndrome, hypercalcemic states, idiopathic calcium oxalate urolithiasis)

- Pelviureteric or ureterovesical junction obstruction may lead to stone formation
- Extracorporeal shockwave lithotripsy (ESWL) and smaller endoscopic equipment have narrowed the differences in surgical management between pediatric and adult patients with nephrolithiasis

Signs

- Colic
- Recurrent urinary infection, mainly under the age of 3 years
- Hematuria (microscopic or macroscopic), mainly in older children
- Abdominal flank pelvic pain
- Interrupted stream or dysuria (bladder or urethra stones)

Medical evaluation

- Urine analysis (pH, specific gravity, urine culture), urine sediment
- 24-h urine analysis (Ca, P, Na, K, creatinine, urea, uric acid, cystine)
- Stone analysis after passage
- Serum Ca, P, Na, K, creatinine, urea, uric acid level
- Examination of metabolic disorders
- Ultrasonography
- Abdominal X-ray
- CT scan
- Intravenous pyelography

Indication for stone removal

- Symptomatic stones, obstructive or threatening to become so
- Stones as a source of infection

Conservative treatment

- Fluid balance with control of specific gravity
- Acidification of the urine in magnesium–ammonium–phosphate stones and antibiotics
- Special diet in cases of cystine stones
- ESWL
- Percutaneous lithotripsy (PL)

Operation: percutaneous nephrolithotomy

- Rarely used in pediatric surgery
- Utilize a nephroscope or ureteroscope
- Extract with visualization
- Break larger stones using ultrasonography

Operation: open stone removal

- Rarely necessary, only when urinary calculi are not amenable to ESWL or PL
- Make an incision below the 12th rib
- Expose the kidney and the ureter
- Open the renal pelvis and extract the stone (or ureter in the case of a ureteral stone)
- Wash the entire calyx system
- Suture the pyelon or the ureter

Postoperative care

- Ureter drain for 2–5 days with an antegrade contrast X-ray before drain removal
- Antibiotic therapy as prophylaxis in cases of vesicoureteral reflux
- Urine culture once a month
- Ultrasonography

Prognosis

- Stone recurrence is rare if urine is sterile and an obstruction does not occur

34.14 Urinary Incontinence

General considerations

- Urine incontinence is a term used for unintentional loss of urine beyond toilet-training and until the 5th year of life

- Urine incontinence may have an organic or a psychological etiology
  - Hereditary
  - Abnormal day–night rhythm of antidiuretic hormone (ADH) secretion
  - Reduced bladder capacity

Classification

- Urinary incontinence can be classified as continuous, intermittent, urge, or stress (Table 34.5). Enuresis classification is shown in Table 34.6.

Table 34.5 Classification of urinary incontinence

Incontinence	Definition	Occurrence
Continuous	Constant urine leak	Almost exclusively associated with congenital malformations
Intermittent	Daytime incontinence and/or Nocturnal incontinence = enuresis	Applicable only to children over 5 years old
Urge	Urine loss by unexpected, immediate need to void	After attainment of bladder control or after the age of 5 years
Stress	Urine leakage due to increased intra-abdominal pressure (coughing)	Very rare in neurologically normal children

Note that according to the most recent nomenclature (2006) only nocturnal incontinence is defined as enuresis.

Table 34.6 Subgroups of enuresis

Enuresis	Incontinence
Primary	During the night (sleeping time), child has never been continent or has been continent for less than 6 months
Secondary	During the night (sleeping time) following a significant dry period (at least 6 months)
Monosymptomatic	Without any other symptoms of lower urinary tract and without a history of bladder dysfunction
Not monosymptomatic	With other symptoms of lower urinary tract (see "Signs")

Signs

- Daytime urinary incontinence
  - Urinary tract infections
  - Vaginal reflux
  - Underactive bladder with decreased voiding frequency (≤3 voids per day)
  - Overactive bladder (incontinence together with urge)
  - Dysfunctional voiding with habitual contraction of the urethral sphincter during voiding, probably combined with incontinence (due to recurrent urinary tract infections or an overactive bladder)
  - Constipation
  - Mechanical obstruction of the lower urinary tract
- Enuresis
  - Failure to thrive
  - Dysuria (increased/decreased voiding frequency)
  - Urgency
  - Hesitancy
  - Straining
  - Intermittency
  - Weak stream
  - Holding maneuvers
  - Incomplete emptying
  - Post-micturition dribble
  - Genital pain or pain of lower urinary tract
  - Abdominal tumor on palpation (full bladder)

Diagnostic work-up

- Detailed history
- Physical examination: genitals, gait, lumbo-sacral region, abdominal mass (constipation), lower extremities reflex
- Urine examination
- Bladder diary
- Ultrasonography as a diagnostic tool for malformations

- Ultrasonography for determination of residual urine in the bladder after voiding
- Uroflowmetry
- Video voiding cystourethrography
- Bladder manometry
- Cystometry
- Cystoscopy to exclude malformations

Treatment: daytime urinary incontinence

- Surgical treatment of underlying pathologies
- Increase the frequency of voiding by imposing timed voiding at frequent intervals (underactive bladder)
- Toilet training
- Antibiotic therapy/prophylaxis (if urinary tract infections)
- Anticholinergics (if overactive bladder)
- Laxatives (if constipation/encopresis)
- Physiotherapy of the pelvic floor
- Psychological management if necessary

Treatment: enuresis

- Detailed information to the child and the parents
- Reduce fluid intake in the evening
- Bladder training and/or anticholinergics (if low bladder capacity)
- Enuresis alarms
- Desmopressin (specially for short stays away from home, i.e., school camp)
- Psychological treatment (eventually family therapy)

34.15 Bladder Rupture

General considerations

- Forceful trauma on a full urinary bladder (in small children the bladder is unprotected)
- Bladder rupture is usually associated with pelvic fracture

Classification

- Bladder rupture is classified as intra- or extraperitoneal (Table 34.7)

Table 34.7 Bladder rupture classification

Rupture	Description
Intraperitoneal	Rupture of the bladder in the abdominal cavity after severe, direct, blunt lower abdominal trauma
Extraperitoneal	Rupture of the bladder due to injury of the pelvic ring

Signs

- Lower abdominal pain, tenderness
- Gross hematuria on initial urination
- Patients are unable to void – urinary retention
- Peritonitis in cases of intraperitoneal rupture

Preoperative work-up

- Pelvic X-ray to exclude pelvic fracture
- Ultrasonography
- Retrograde cystography (difference between extra- and intraperitoneal bladder injury)
- Intravenous pyelography
- CT scanning

Therapy

- Broad-spectrum antibiotics
- Extraperitoneal ruptures: usually catheter drainage alone is enough
- Intraperitoneal ruptures: emergency surgical repair

Postoperative care

- Catheterization of the bladder for 2 weeks
- Before removing the bladder catheter, cystography is needed to document complete healing

34.16 Urethral Injuries

General considerations

- All posterior urethral injuries are associated with concomitant pelvic fractures
- Anterior urethral injury is common after "straddle" injury to the perineum
- These injuries are often associated with bladder rupture, bladder neck or vaginal injury

Signs

- Blood at the urethral meatus
- Urinary retention
- Scrotal and/or perineal hematoma (anterior urethral trauma)

Investigations

- Retrograde urethrography prior to catheterization
- Catheterization if there is no evidence of extravasations
- Urethrocystoscopy in girls

Therapy

- Primary urethra reconstruction in cases of injury due to pelvic fracture
  - Removal of hematoma with drainage
  - Realignment of the ruptured urethra and stenting
  - Suprapubic catheter
- Delayed urethra reconstruction
  - Transurethral catheter if possible, or suprapubic catheter
  - Delayed reconstruction should not be attempted until at least 6 months after trauma

Postoperative care

- Leave the urethral catheter in place until healing is achieved
- Antibiotics

Prognosis

- In severe cases, poor
- Urethral strictures
- Erectile dysfunction
- Urinary continence can be impaired
- Impotence is frequent
- These complications are mainly due to the initial trauma rather than the reconstructive surgery

35 Kidney Insufficiency

General considerations

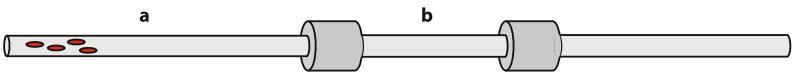
- In this condition the patients need either hemodialysis or peritoneal dialysis to treat terminal renal insufficiency
- In our experience catheter longevity is superior after laparoscopic implantation compared to open surgical implantation
- We describe the use of laparoscopy in the following situations
  - Primary laparoscopic implantation of a Tenckhoff catheter for peritoneal dialysis
  - Laparoscopy to reestablish continuous abdominal peritoneal dialysis (CAPD) in cases of a malfunctioning catheter
  - Secondary laparoscopic implantation of a peritoneal catheter because the original catheter has malfunctioned
- The cumulative probability of catheter malfunction with subsequent catheter replacement is, according to the National CAPD Registry of the National Institutes of Health (1988):
  - 18% after 12 months on CAPD
  - 32% after 24 months
- The cumulative probability of exit-site or tunnel infection is
  - 33% in the first year of CAPD
  - 49% in the second year

Indications

- Need for CAPD
- Any catheter malfunction

Preoperative work-up

- Routine laboratory investigations
- Preparation of an individual pig-tail catheter
  - Measure the distance of the coil to the deep cuff = intra-abdominal catheter part (*a*)
  - Measure the distance between the cuffs = subcutaneous tunnel length (*b*)



- Mark the site of the deep and the superficial cuff on the abdominal wall
- Evacuate the bladder (in case of residual kidney function)
- Prophylactic antibiotics

Operation: primary implantation

- The operative steps are illustrated in Fig. 35.1
- The catheter is immersed in sterile saline and both cuffs are squeezed and rotated several times to eliminate any air trapped within the cuff, since air inhibits the ingrowth of fibrous tissue
- Creating a pneumoperitoneum with the special Verres needle
- Inspect the abdominal cavity and select the deep cuff position
- Dissect over the implantation site of the deep cuff (1.5 cm) on the medial border of the rectus muscle on the left side above the level of the anterior superior iliac crest
- Open the rectus fascia (1 cm)
- Make a blunt dissection of the muscle fibers and expose the peritoneum
- Puncture the peritoneum with the split cannula at an angle of about 45° from the vertical towards the pelvis under laparoscopic control
- Retract the blunt trocar after positioning behind the bladder
- Dilate the peritoneal puncture
- Implant a catheter under laparoscopic control
- Remove the split cannula
- Orient the catheter with the curled portion open to the midline

- Position the deep cuff under the fascia and suture
- Pass the catheter through the subcutaneous channel with the tunnel instrument
- If needed, one additional 1.7-mm forceps may be used

Postoperative management

- CAPD 6 h after implantation

Complications

- Leakage of the implantation site

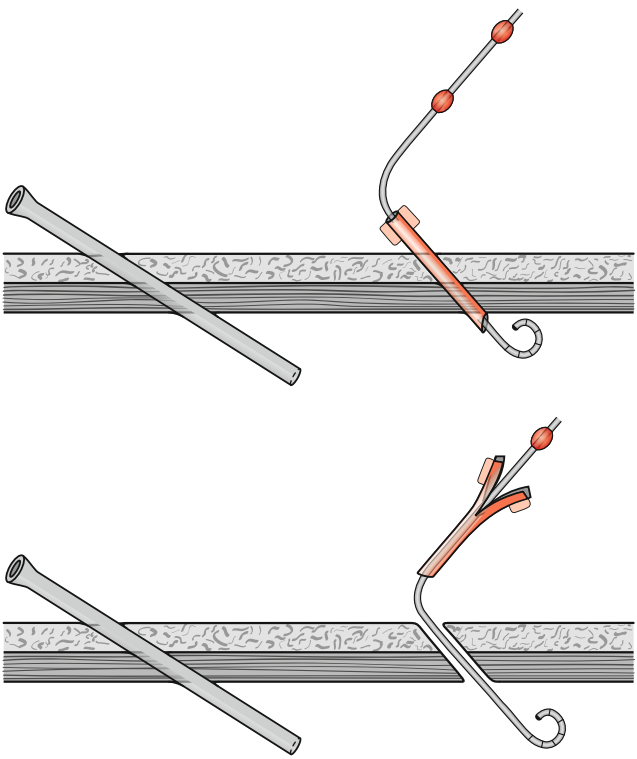


Fig. 35.1 Operative steps: laparoscopic Tenckoff implantation

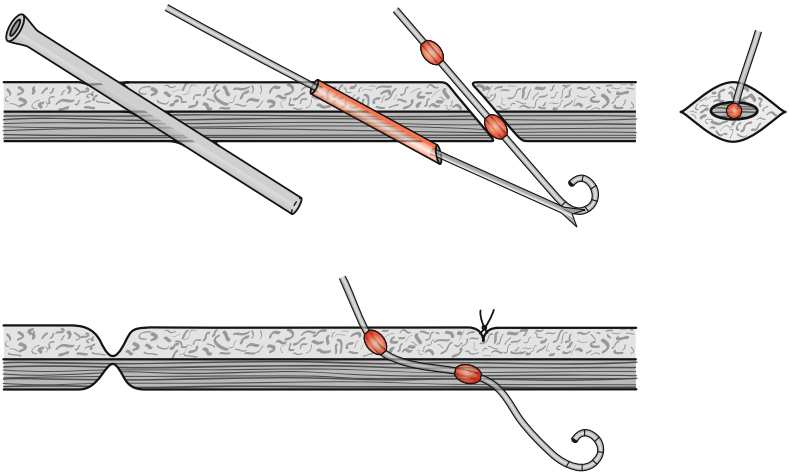


Fig. 35.1 *(continued)* Operative steps: laparoscopic Tenckoff implantation

36 Skin and Soft Tissues

36.1 Pigmented Nevi

General considerations

- The classification of nevi as junctional, compound or intradermal is determined by the exact location of the nevus cells in the skin
  - Junctional nevi may present at birth, in early childhood or during adolescence. They may occur anywhere on the body, are relatively small, flat, and irregular in color or shape
  - Compound and intradermal nevi are most frequently on the head and neck, and may become verrucous, raised, dome-shaped or pedunculated
- Clinically and pathologically nevi may be classified as non-melanocytic or melanocytic
  - Non-melanocytic nevi are occasionally associated with congenital syndromes, and the risk for malignant transformation is low
  - Melanocytic nevi have a risk for malignant transformation
- Excessive sun exposure is the main risk factor for development of malignant melanoma in melanocytic nevi patients
- Patients with melanocytic nevi should be observed with care (photographic documentation)

Operation

- Pigmented nevi are benign lesions and need to be removed only for cosmetic reasons or to avoid chronic irritation and infection caused by local traumatic damage

- A rapid increase in size, change in color and shape, itching, the development of satellite lesions, and pain are indications for adequate excision and histological evaluation
- The current recommendation for the management of large (giant) congenital melanocytic nevi is early excision because of the malignancy risk (often multiple surgeries and skin grafting are necessary)

36.2 Dermoid Cysts

General considerations

- Subcutaneous congenital cysts with dermal elements (hair follicles, sebaceous glands, lipid, and keratin)
- Location – head (under the lateral part of the eyebrow), nasal bridge, perineal raphe, and scrotum
- May penetrate the orbital bone
- Tendency to grow and to become infected

Signs

- Soft tumor that is most often fixed to bone or less commonly to other deep-lying tissues
- Painless mass 1–2 cm in diameter
- In cases of local infection, there are signs of inflammation

Preoperative work-up

- Physical examination (palpation with photographic documentation)
- Ultrasound diagnosis (for cervical tumors)
- X-ray
- CT (computed tomography) in cases where there is a paraorbital process

Operation

- Radical surgical excision

36.3 Lymphadenitis

General considerations

- Acute bacterial cervical adenitis (secondary to acute pharyngitis) and inguinal lymphadenopathy (resulting from infections of the lower extremity) are common (penicillin-resistant *Staphylococcus aureus* and *Streptococcus hemolyticus*)
- Cat scratch disease – the most common cause of non-bacterial lymphadenopathy
- Chronic supraclavicular – cervical lymphadenitis is frequently caused by tuberculous and atypical mycobacterial (bovine tuberculosis) infections

Signs

- Painful and swollen tumor
- Initially – local infiltration without skin inflammation
- Later – fluctuation with hot and red overlying skin
- Fever
- Leukocytosis
- In cat scratch disease – local superficial infection or pustule (3–5 days) is followed by regional lymphadenitis (in 10–12 days)
- Patients with atypical mycobacterial process are asymptomatic
- Tuberculous lymphadenitis patients may be symptomatic (pulmonary tuberculosis)

Preoperative work-up

- Take a history (oropharynx bacterial infection, local skin and soft tissue inflammatory damage, contact with domestic animals, BCG vaccination, family history)
- Physical and ultrasound examinations
- Diagnostic needle aspiration
- Examination for cat scratch disease, tuberculosis, bovine tuberculosis, and actinomycosis infection
- Start antibiotic therapy

Operation

- Perform incision and drainage with a Penrose drain (by suppuration)
- Lymph node extirpation (indicated after 6–8 weeks of ineffectual antibacterial treatment, for those with cat scratch disease or atypical mycobacterial infection)
- Fistula excision and lymph node extirpation (in tuberculous lymphadenitis)

Postoperative care

- Daily irrigation through the Penrose drain
- Postoperative antibacterial treatment for 7–10 days

Prognosis

- Good for acute lymphadenitis patients

36.4 Burns

General considerations

- Burn injuries are caused by thermal energy, electricity or chemicals
- Thermal injuries result from scald (85%) or flame (13%) burns
- Scalds are more common below the age of 5 years
- Flame burns are the predominant cause of thermal injury in older children
- Electrical and chemical burns are less common (approximately 2%)
- Burns are the second most common cause of accidental death in children under 5 years of age
- Hemodynamic, cardiopulmonary, renal, and metabolic disturbances develop rapidly following severe thermal injury
- Burns result in a varying degree of cell death and dysfunction (depends on the heat intensity, skin thickness and time of exposure to the thermal agent)

Classification

- Burns are classified according to the depth of tissue injured (Table 36.1)

Table 36.1 Classification of burns

Degree	Extent of damage		Features	Healing
First	Basal layer of the epidermis (superficial thickness)		▪ Red burn site ▪ Painful ▪ Dry ▪ No blisters	▪ Spontaneous healing in 3–8 days ▪ No scarring
Second	Epithelium and greater portion of dermal layer	Superficial partial thickness	▪ Red (blanches with pressure) ▪ Painful ▪ Moist ▪ Blistered	▪ Spontaneous healing in 7–20 days ▪ Scarring unusual ▪ Pigmentary changes possible
		Deep partial thickness	▪ Variable color, white to red (does not blanch with pressure) ▪ Painful on pressure ▪ Wet or waxy ▪ Unroofed blisters	▪ Slow healing in 3–6 weeks ▪ Significant scarring ▪ Risk of contracture
Third	Entire epidermis and dermal layers (Full thickness)		▪ White or charred (does not blanch with pressure) ▪ No sensation ▪ Dry	▪ Spontaneous healing impossible ▪ High risk of contracture

Diagnosis

- The body surface area in children varies with their age (the size of a burn injury should be estimated and documented by standardized charts)
- The initial estimation of burn depth is frequently not simple, because a burn is not a static injury
- Lund–Browder charts should be used to assess the percentage of body surface area involved (Fig. 36.1)

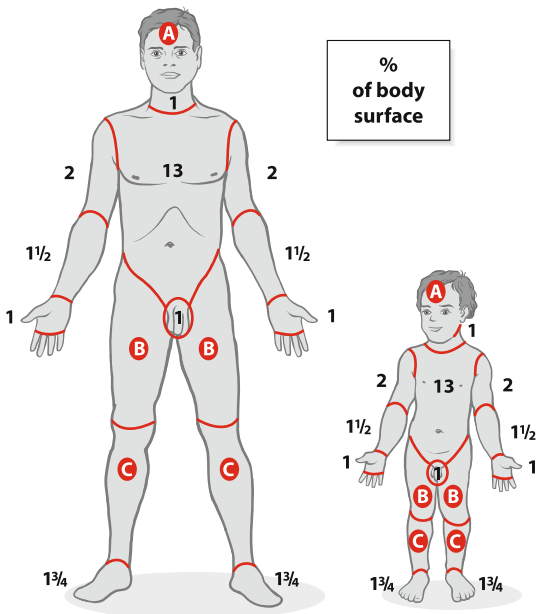


Fig. 36.1 Burn injury chart

Bodypart	Age (years)					
	0	1	5	10	15	Adult
A = ½ head	9½	8½	6½	5½	4½	3½
B = ½ of the thigh	2¾	1¼	4	4¼	4½	4¾
C = ½ of one lower leg	2½	2½	2¾	3	3¼	3½

Criteria for hospitalization

- Partial-thickness and full-thickness burns of >10% in patients less than 10 years old
- Partial-thickness and full-thickness burns of >20% in older patients
- Partial-thickness and full-thickness burns involving the eyes, face, ears, hands, feet, perineum, genitalia, anus, or overlying major joints
- Full-thickness burns of >5% at any age
- Electrical burns
- Inhalation burns
- Significant chemical burns
- All patients under 18 months old
- Patients with preexisting illnesses that could complicate recovery or increase mortality and morbidity
- Social issues (unsafe home environment, suspected abuse)

Treatment

- Initial management of severe burns
  - Separate the patient from the cause of the burn
  - Cover the burn wound with a clean cloth soaked in cool water or irrigate with cool tap water (15–20 min)
  - Saturate the dressings covering the wounds with an antimicrobial agent
  - Intubate early in cases of inhalational injury or carbon monoxide poisoning
  - Set up an intravenous infusion with Ringer's lactate (20 $ml \cdot kg^{-1} \cdot h^{-1}$ until more accurate estimates of fluid requirements are made)
  - Place a nasogastric tube (to prevent vomiting)
  - Site a urinary catheter with urinary output monitoring
  - Start intravenous pain management
- Fluid resuscitation during the first 24 h after the burn
  - Ringer lactate: 2000 $ml \cdot m^{-2}$ of body surface area every 24 h plus 5000 $ml \cdot m^{-2}$ of body surface area burned per 24 h. One-half of the calculated fluid is given over the first 8 h post-burn; the second half over the subsequent 16 h

- Minimum urine output: 1 $ml \cdot kg^{-1} \cdot h^{-1}$ (in children weighing less than 30 kg) and 30–40 $ml \cdot h^{-1}$ (for those weighing more than 30 kg)

- Fluid resuscitation after the first 24 h (three-quarters of the first day's allowance)
  - 1500 $ml \cdot m^{-2}$ of body surface area every 24 h plus 3750 $ml \cdot m^{-2}$ of body surface area burned every 24 h (plus electrolyte-colloid therapy)
- Inhalation injury may increase quantity of fluids required by 50%
- Caloric requirements: 7560 $kJ \cdot m^{-2}$ of body surface area plus the same amount for the area of body surface burned (20% protein, 60% carbohydrates, 20% fats)
- Antibiotics are usually not given prophylactically, but are administered when infections occur (hyperthermia, wound infection, leukocytosis, thrombocytopenia, tachypnea)
- Blood and other appropriate cultures (in the first day of infection)
- Nasogastric tube feeding for 12–24 h after burn injury
- Pain control (analgesics should be administered, especially for painful procedures)
- Intubation and respiratory support (upper airway obstruction, edema, hypoxemia, alveolar-capillary block syndrome)
- Tetanus prophylaxis

Operation

- Burn wound care
  - Gently cleanse the wounds and debride ruptured blisters
  - Apply topical agents to control microbial wound colonization and to reduce local sepsis (silver sulfadiazine – Silvadene®)
  - Apply the first dressing after 24 h
  - Debride the burn change the dressing once a day
  - Once the wound has epithelialized it is appropriate to leave the dressing on for 4–72 h
- Surgical treatment of burn wounds
  - Escharotomies (for circumferential burns of the extremities or chest wall burns)

- Perform an early complete excision as soon as the patient is hemodynamically stable, using a temporary skin substitute to cover the wounds or auto-grafting
- Make staged excisions (for patients with deep burns and for wounds of indeterminate depth)
- Apply cultured epithelial cells derived from a small skin biopsy specimen from the patient (in patients with massive burns). After several weeks the culture can produce epithelial sheets capable of covering wounds
- Carry out fasciotomy, frequent debridement of necrotic tissue, and amputation (for patients with electrical injury)

Rehabilitation

- The aim is to return the patient to as normal a home life as possible
- Cover the burn wound
- Give the patient active and passive range-of-motion exercises
- Splinting
- Compression garments (in hypertrophic scarring)
- Physical therapy

Prognosis

- Severe inhalation injury and sepsis are a lethal combination
- Survival depends on the burn size and depth, the presence of inhalation injury, and the patient's age
- Fluid retention during the first 48 h following resuscitation is critical for the survival of burns patients
- Abused patients have a higher mortality than those who are not abused

37 Osteoarticular System

37.1 Trigger Thumb

General considerations

- Trigger thumb is a congenital malformation in the thumb joint
- The thumb is flexed 20°–30° at the level of the interphalangeal joint
- The tendon may pass, forcefully, through the tendon pulley, on passive extension, but otherwise stays blocked and the child is unable to spontaneously extend the interphalangeal joint

Signs

- The thumb eventually "cracks" on passive dorsiflexion or remains partly flexed

Preoperative work-up

- Routine anesthetic evaluation

Operation

- Make a transverse skin incision at the transition between the thumb and the palm of the hand, at the level where a hard nodule is felt
- Incise the circular fibrous band, distal to the nodule, longitudinally, until the tendon's "proximal nodule" is visualized and the tendon is free to move

Postoperative care

- Immobilization of the thumb in extension
- Hyperextension, followed by active mobilization 10 days later

Prognosis

- Very good

37.2 Polydactyly

General considerations

- Incidence: 8 in 10,000 newborns
- Most often it is a sort of appendage on the ulnar side of the hand
- In polydactyly the appendage rarely constitutes a whole digit

Signs

- Cosmetic problems
- One finger can be functionally bound to the extra finger

Preoperative work-up

- X-ray of the hand
- Routine anesthetic evaluation

Operation

If there is only one appendage

- Make an elliptical incision at the base of the digit
- Excise the digit and the atrophic tendon structures

If a whole finger structure exists

- The metacarpal of the extra finger may join the metacarpal of the fifth digit
- Make a longitudinal elliptical incision, slightly overriding the extra finger
- Release the finger as far as the level of the junction of the two metacarpal bones
- Separate all tendons of the extra digit, near the bone
- Perform a bone section as a straight line

Postoperative care

- Normal dressings

Prognosis

- Very good in the simplest forms
- In the more severe forms, particularly those involving the thumb, special expertise may be required to improve prognosis

37.3 Syndactyly

General considerations

- During embryogenesis some cells are resorbed in order to create the interdigital spaces. If that separation process is disrupted in any way, the fingers remain bound together
- Incidence 7 in 10,000 newborns
- The simplest form (and rarest) consists only of skin connection
- When bones are joined (typically the terminal phalanx) special expertise is required

Preoperative work-up

- X-ray of the hand

Operation

- Make unsymmetrical zigzag incisions on the dorsal and volar aspects of the digits, raising a small skin flap (at the dorsal or volar aspect of the base of the fingers) to reconstitute the interdigital web
- Preparation in the midline, without damaging vessels and nerves or the base of the triangles
- Make a midline separation of the membrane, and eventual distal osteotomy (fused phalanges)
- Always divide the skin equally between the two fingers. Complementary full-thickness skin grafts are nearly always required on both sides

- When there is partial recurrence of the syndactyly with interdigital contracture at the base of the fingers at least 1 year later, a complementary correction can be performed; namely, using a V-shaped, inferiorly based flap, associated with multiple Z-plasties ("jumping man")

Postoperative care

- Dressings should be changed on postoperative day 7
- Only mobilize the fingers once the sutures are fully healed
- Physiotherapy

Prognosis

- Frequent recurrences in the form of contractures

37.4 Hand Trauma Treatment

Incisions on the hand

- A deformed hand is incapacitating
- The digital folds must not be incised at 90°, even when performing Z-plasties
- Damage to the dorsal veins of the hand should be avoided
- Permitted incisions are shown in Fig. 37.1

Tendon Sutures

- Tendon sutures are demonstrated in Figs. 37.2–37.4

Normal resting position of the hand

- Wrist: 30°–40° dorsiflexion
- Metacarpophalangeal joint: 90° flexion
- Proximal interphalangeal joint: 0° neutral
- Distal interphalangeal joint: 0° neutral

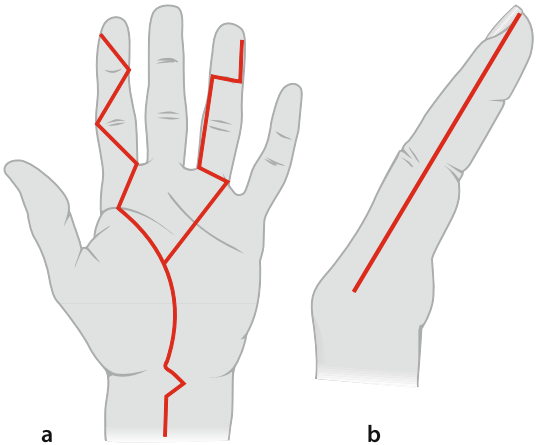


Fig. 37.1a, b Incisions permitted on the hand

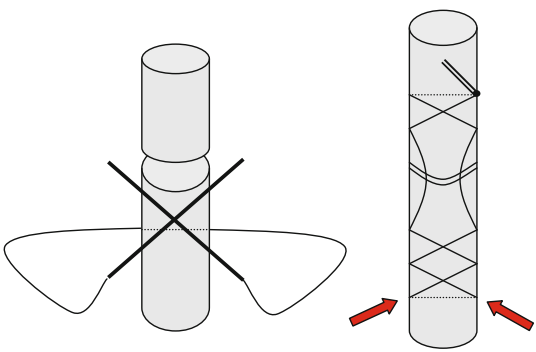


Fig. 37.2 Suture according to Bunnell

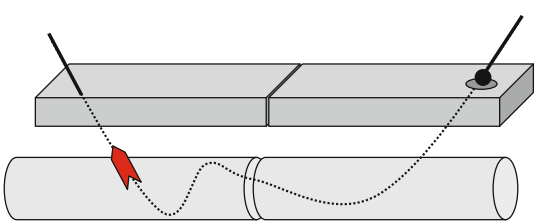


Fig. 37.3 Suture according to Lengemann

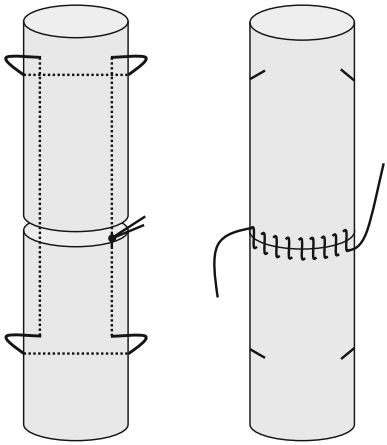


Fig. 37.4 Suture according to Kleinert

Dynamic immobilization

- The affected finger should be held with elastic traction that allows for more active stretching of the finger, but passive stretching of less than 20° (Fig. 37.5)
- The finger must be pulled in the direction of the scaphoid bone
- The thumb must be pulled in the direction of the metacarpal heads

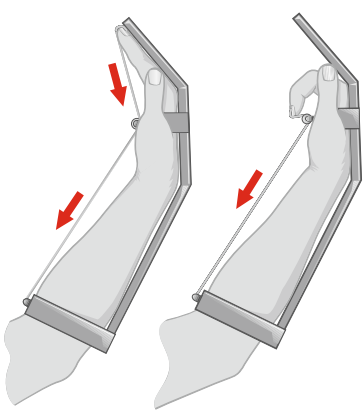


Fig. 37.5 Kleinert immobilization cast

37.5 Ganglions/Baker's Cyst

General considerations

- **Ganglions**
  - Cystic synovial structures at the level of joint capsules or tendon sheaths, with a gelatinous content
  - In children they appear more frequently near the hand joints and the volar aspect of the wrists
- **Baker's cysts**
  - Ganglions are located in the popliteal region. They present mainly in the first decade of life, particularly around the age of 6 years
  - Located medial to the gastrocnemius muscle and lateral to the semitendinosus and semimembranosus muscles

Signs

- Subcutaneous nodule, with mild pain on compression, or due to increased tension within the ganglion

Preoperative work-up

- Simple clinical evaluation
- Ultrasound

Operation

- Make an incision over the cyst
- Open the deep fascia (in the case of a Baker's cyst)
- Expose the cyst
- Dissect the cyst, avoiding rupture
- Any communication with the adjoining joint must be closed

Postoperative care

- A compressive bandage is advisable

Prognosis

- Recurrence rate is high (around 40%), only if there is incomplete removal of the capsule
- Occasionally, Baker's cysts undergo spontaneous remission, so that the indication for surgery should be limited to symptomatic lesions

37.6 Congenital Hip Dislocation

General considerations

- Malformation of the proximal femur, acetabulum, and capsule of the hip joint
- Familial tendency, incidence of 3%
- Girls are affected five times more than boys by the malformation: 60% on the left side, 20% on the right side, and 20% bilateral
- Examination of the hips should be made at birth and at 4 and 12 weeks of age

Classification

- Congenital hip dislocation classification is shown in Table 37.1 and Fig. 37.6

Table 37.1 Congenital hip dislocation classification

Grade	Description
I	Slight capsular instability. Acetabular eversion
II	Marked capsular instability, with subluxation. Marked acetabular eversion, and anteversion of the neck of the femur
III	Dislocation – the labrum and limbus interposed

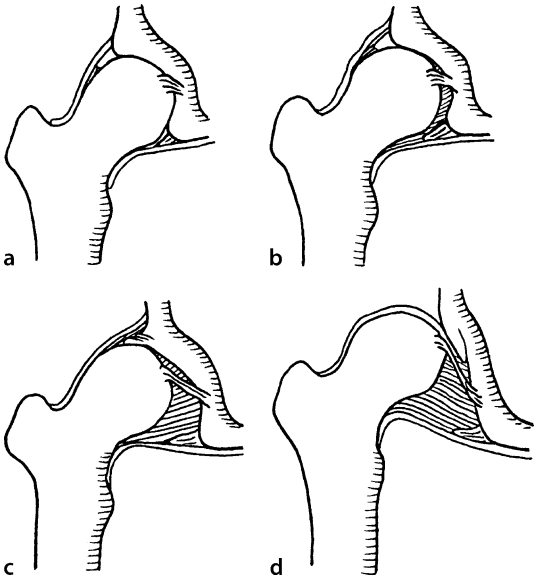


Fig. 37.6a–d Classification of hip dislocation. **a** Normal, **b** grade I, **c** grade II, **d** grade III

Signs

- Enquire about family history, breach presentation, oligohydramnios
- Barlow's sign, leading to dislocation of the hip
- Thomas's sign: with the knee extended the hip cannot be flexed
- Ortolani's sign: rebound when reintroducing the hip into the acetabulum
- Palmen's sign: free hip
- Asymmetry of the thigh folds
- Asymmetry of the buttock folds
- Apparent shortening of the femur on the affected side
- Unlevel pelvis

Investigations

- Ultrasound (see Fig. 8.8)
- X-ray the pelvis in special cases (after 6 months of age)

Conservative treatment

- In grade I, simple abduction until the age of 4 months
- In grade II, special abduction frame
- Overhead extension in true luxation
- Control at 6th week of therapy
- In luxation control at 3rd week of therapy

Operation

- Indicated when a stable situation, i.e., with a well-centered femoral head, is not possible
- Open reduction
  - Determine the approach to the hip joint (Smith Peterson or Watson)
  - Carry out reconstruction according to Salter (not in very young children)
  - Choose from the various different femoral osteotomies to centralize the femoral head in the joint

Prognosis

- The sooner the diagnosis (at birth) and treatment, the better the prognosis
- With early care, conservative treatment may be enough

37.7 Osteomyelitis

General considerations

- Hematogenous bone infection appearing in children following a bacteremia
- Direct infection of bone in children is rare
- Microbial infection with
  - *Staphylococcus aureus*
  - *Staphylococcus epidermidis*
  - α- and β-hemolytic streptococci
  - *Pneumococcus*
  - *Haemophilus influenzae*

- Mixed infection
- It is more frequent in newborns (27%) than in children
- Normally osteomyelitis starts at the metaphyses of the long bones
- Until the second year the metaphyses and epiphyses have the same blood vessels and can both be involved and the infection easily expands to involve the joint (osteoarthritis, particularly in the newborn)
- After the second year until puberty, metaphyses and epiphyses have different blood supply (Fig. 37.7)
- In the adult the bone is fed by only one vessel
- Differential diagnoses: bone tumors, acute leukemia, septic arthritis
- The pathophysiology of osteomyelitis is illustrated in Fig. 37.8

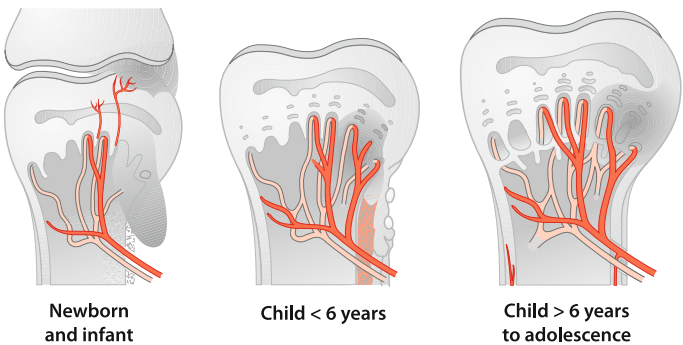


Fig. 37.7 Blood supply of the epiphysis

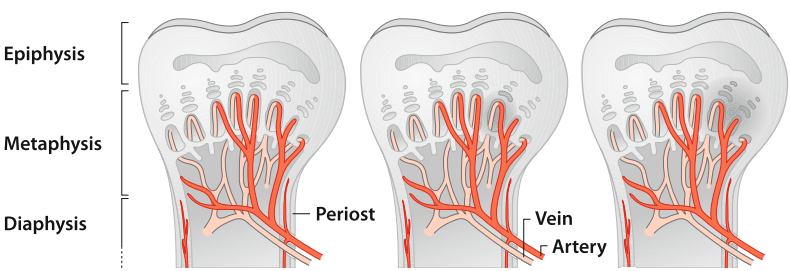


Fig. 37.8 Pathophysiology of osteomyelitis

Signs

- Functional impairment
- Fever
- Sepsis
- Symptoms of infection (swelling, warmness, redness, pain, functional impairment)

Preoperative work-up

- Routine anesthetic evaluation
- Blood count with differential
- C-reactive protein/erythrocyte sedimentation rate (ESR)
- Diagnostic confirmation (needle aspiration)
- X-ray only after 10–14 days in the newborn
- Ultrasound, to evaluate soft tissue abscesses and periosteal compromise
- Isotopic scintigraphy
- MRI

Antibiotic therapy

- Clindamycin (40 $mg \cdot kg^{-1} \cdot day^{-1}$, t.i.d.) or vancomycin (30 $mg \cdot kg^{-1} \cdot day^{-1}$)
- In neonates treat with cefotaxime (150 $mg \cdot kg^{-1} \cdot day^{-1}$, t.i.d.)
- Continue treatment until ESR normalizes
- Abscesses, necrotic tissues, bone sequestration, fistula or failure of conservative treatment are indications for surgery
- A flowchart for antibiotic therapy is outlined in Fig. 37.9

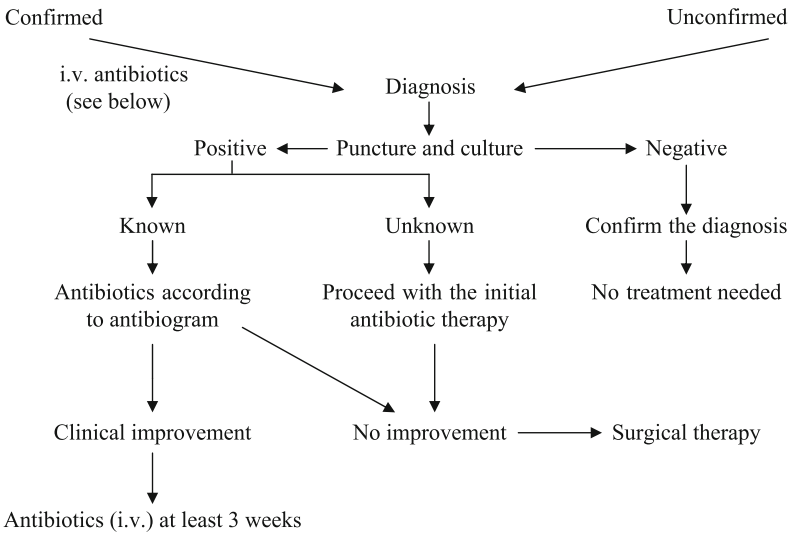


Fig. 37.9 Flowchart for antibiotic therapy

Operation

- Open the abscess, remove dead bone
- Perform aspiration and drainage
- Apply antibiotic collar (polymethylmethacrylate, PMMA) in some cases

Postoperative care

- Daily dressings
- Antibiotic therapy as indicated

Prognosis

- Original state restored in approximately 80%
- Important orthopedic problems arise when therapy is delayed or insufficient

38 Skeletal System

38.1 General Considerations

Due to the impulsive nature of children's movements, they very frequently injure their skeleton. One in seven children visiting the emergency room has a fracture. Pediatric surgeons must therefore have sound knowledge of the immature skeleton; importantly, the differences in the physical qualities of a child's compared to an adult's skeleton and the phenomenon of "growth" itself.

The aim of child-oriented treatment must be to achieve the greatest possible clinical efficiency, i.e., excellent results with minimal intervention. Hence, the only treatment strategy is:

The first treatment should be the correct and the definitive treatment.

Development and growth

- Compared to mature bone, immature bone is more capable of reaction and adaptation, but it is also more vulnerable. A fracture in an immature bone signals either the acceleration or deceleration of growth, which has the potential to induce deformity, and adds to the complications of the fracture itself
- Having said that, children's fractures heal rapidly. Depending on the child's age and the direction of the deformity, the bone can remodel to correct even the most angular of malunions. The most important area of injury in the immature skeleton is the growth plate or epiphysis

Regulation of epiphyseal growth

- The growth plate (epiphyseal line) is the primary center for growth in most bones and may be divided into two zones according to function: (1) the zone of growth, involved in both longitudinal and circumferential bone growth, and (2) the zone of matrix formation
- The growth plate is capable of responding to different stimuli (i.e., fracture or injury) with either compression (i.e., cessation of growth) or tension (i.e., bone resorption)

Growth and remodeling of metaphyseal and diaphyseal bone

- The metaphysis is the site of most rapid change in bone structure, as the deeper zones of the growth plate mature and the growth plate produces primarily trabeculae
- Circumferential growth of the diaphysis is a function of appositional bone formation by the periosteum, together with osteoclast resorption by the endosteum so as to enlarge the medullary cavity. As growth continues, the bone is capable of reducing, or even correcting, angular deformity by selective resorption and apposition, possibly driven by compression and tension forces (Table 38.1)

Table 38.1 Self-correction potential of the growing skeleton

Side-to-side displacement	Axial deformity (frontal/sagittal plane)	Shortening/lengthening	Rotation failures
Periosteal, endosteal	Epiphyseal		
Direct			Indirect

38.2 Classification

There are numerous systems for classifying bone fractures: according to localization, displacement and stability (Table 38.2); according to localization and involvement of the growth plate (Table 38.3); the Salter–Harris/Aitken system (Table 38.4); and the AO Pediatric Comprehensive Classification of Long-Bone Fractures (PCCF) (Table 38.6).

Table 38.2 Bone fracture classification according to location, displacement, and stability

Stability	Localization		Therapy
	Diaphyseal/ metaphyseal	Articular	
Sufficiently stable for initial retention	Transverse fractures, with tolerance limits depending on age or Oblique/spiral fractures of one bone of the lower leg or forearm	Non-displaced or minimally (<2 mm) displaced articular fractures	Immobilization with plaster in combination with cast wedging if necessary
Unstable fractures	All fully displaced fractures	Articular fractures with a gap >2 mm!	Reduction under anesthesia with either conservative (plaster) or operative stabilization

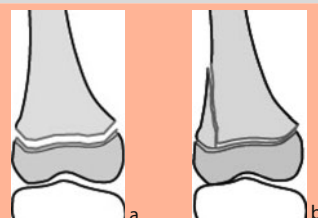
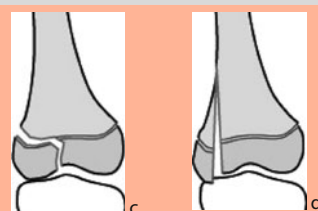
Table 38.3 Classification according to location and growth plate involvement

Shaft fracture	Diaphyseal	Stable	Non displaced fractures without shortening
		Unstable	Displaced fractures with shortening or having the tendency for shortening
		Greenstick	Bowing fractures with complete fracture of one cortex and incomplete facture of the cortex of the contra lateral side
	Metaphyseal	Buckle	Compression of the metaphyseal cortex of one side
		Bowing/ supracondylar	Greenstick fracture in the metaphysis
		Lig. avulsion	Ligament avulsion

Table 38.3 *(continued)* Classification according to location and growth plate involvement

Articular fracture	Epiphyseal	Aitken I Salter–Harris I + II	See Table 38.4
		Aitken II + III Salter–Harris III + IV	See Table 38.4
		Tillaux or bi-plane fracture	In puberty by partially closed growth plate
		Flake fracture	"Normally" in combination with joint dislocation
		Lig. avulsion	Bony or cartilage avulsion

Table 38.4 Salter–Harris/Aitken classification

Salter–Harris			Aitken
I	Epiphysiolysis (*a*)		
II	Epiphysiolysis with metaphyseal wedge (*b*)		I
III	Epiphyseal fracture (*c*)		II
IV	Epiphyseal fracture with metaphyseal wedge (*d*)		III

AO Pediatric Comprehensive Classification of Long-Bone Fractures (PCCF)

The AO Pediatric Comprehensive Classification of Long-Bone Fractures (PCCF) is available to all surgeons as a software package at www.aofoundation.org/aocoiac.

- Fracture location is related to the four long bones and their three segments, as well as the specific pediatric subsegments. The bones and segments within the bones follow a coding scheme similar to that in adults, but the identification of the segments differs. For pediatric long-bone fractures, the end segment has two subsegments:
  - Segment 1: Proximal – including epiphysis (E) and metaphysic (M) subsegments
  - Segment 2: Diaphysis (D)
  - Segment 3: Distal – including metaphysic (M) and epiphysis (E) subsegments
- For the radius/ulna and tibia/fibula bone pairs, both bones must be included in the defined square, see Fig. 38.1
- As malleolar fractures are uncommon in children, they are simply coded as distal tibia fractures (e.g., the fracture of the medial malleolus is a typical Salter–Harris III or IV fracture of the distal tibia, coded as 43)
- The original severity coding of A–B–C used in adults is replaced by a classification system that is known and accepted all over the world, determined according to diaphysis (D), metaphysis (M) and epiphysis (E)
- Epiphyseal fractures (E) involve the epiphysis and its growth plates
- Metaphyseal fractures (M) are identified according to the position of the defined square (where the center of the fracture lines must be located in the defined square) with one side over the growth plate
- For easier and more accurate application of the squares, and thus more reliable classification, a series of defined squares is copied to a transparency that is overlaid on the a.p. radiographic view
- The defined square does not apply to the proximal femur, where metaphyseal fractures are located between the growth plate of the head and the intertrochanteric line

- The morphology of the fracture is documented by a type-specific child code and a severity code, as well as an additional code for displacement of specific fractures

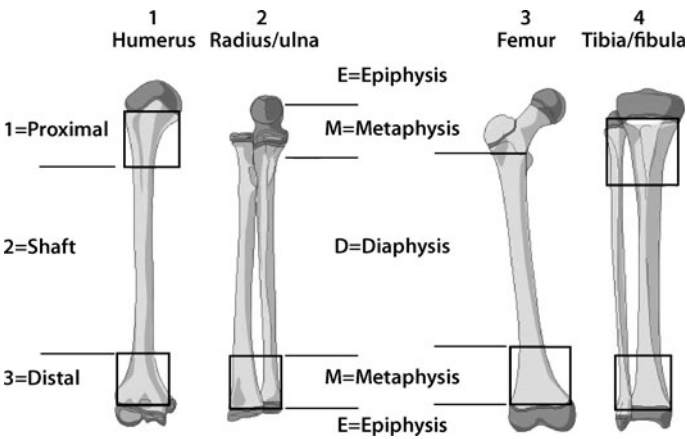


Fig. 38.1 Fracture location related to bone segments and subsegments. For children the defined square must be placed over the larger parts of the growth plate

Overall structure of pediatric fracture classification (Table 38.5)

Table 38.5 Overall classification system of pediatric fracture classification

Diagnosis							
Localization					Morphology		
Bone 1 2 3 4	Segment 1 2 3	-	Subsegment E M D	/	Child 1–9	Severity .1 .2	Displacement I–IV
Example							
Radius	Proximal	-	Diaphysis	/	Transverse	Simple	No angulation
2	1	-	D	/	4	.1	I

- Relevant pediatric fracture patterns, transformed into a "child code," are specific and grouped according to each of the fracture location categories of E, M or D (Tables 38.4 and 38.6)
  - Patterns of epiphyseal fractures include the known epiphyseal injuries I–IV according to Salter–Harris using the child codes E/1 to E/4. Other child codes (E/5 to E/9) are used to identify Tillaux (two-plane) fractures (E/5), tri-plane fractures (E/6), ligament avulsions (E/7), and flake fractures (E/8)
  - Three child patterns are identified for metaphyseal fractures, i.e., the buckle/torus or greenstick fractures (M/2), complete fractures (M/3) and osteoligamentous, musculo-ligamentous avulsion or just avulsion injuries (M/7)
  - Child patterns within segment 2 (diaphyseal fractures) include bowing fractures (D/1), greenstick fractures (D/2), toddler fractures (D/3), complete transverse fractures (angle ≤30°; D/4), complete oblique/spiral fractures (angle >30°; D/5), Monteggia (D/6), and Galeazzi lesions (D/7)
- The grade of fracture severity distinguishes between simple (0.1) and wedge (partially unstable fractures with three fragments including a fully separated fragment) or complex fractures (totally unstable fractures with more than three fragments) (0.2) (Fig. 38.2)
- Fracture displacement for specific fractures (Table 38.7)
  - Supracondylar humeral fractures (code 13-M/3) are given an additional code regarding the grade of displacement at four levels (I–IV)
  - Radial head fractures (code 21-M/2 or /3, or 21-E/1 or /2) are given an additional code (I–III) regarding the axial deviation and level of displacement (I = no angulation and no displacement, II = angulation with displacement that is less than half the bone diameter, III = angulation with displacement that is more than half the bone diameter)

- Paired bones
  - Except for the known Monteggia and Galeazzi lesions, when paired bones (i.e., radius/ulna or tibia/fibula) are both fractured with the same child pattern, a single classification code should be used with the severity code used to describe the worst of the two fractures
  - When a single bone is fractured, a small letter describing that bone (i.e., "r," "u," "t" or "f") should be added after the segment code (e.g., the code "22u" identifies an isolated diaphyseal fracture of the ulna)
  - When paired bones are fractured, each with a different child pattern (e.g., complete fracture of the radius and a bowing fracture of the ulna), each bone must be coded separately including the appropriate small letter (22r-D/5.1 and 22u-D/1.1). This allows for the detailed documentation of combined fractures of the radius and ulna, or those of the tibia and fibula in clinical studies, so their relative influence on treatment outcomes can be properly evaluated

Table 38.6 Classification of pediatric fractures

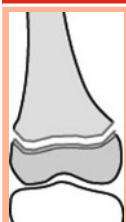
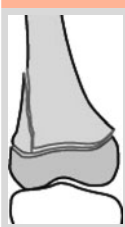
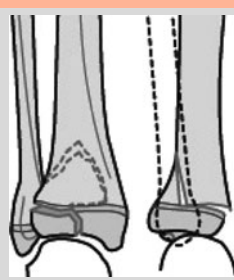
E = Epiphysis			
	E/1 S–H I		E/5 Tillaux (two plane) fractures
	E/2 S–H II		E/6 Tri-plane fractures

Table 38.6 *(continued)* Classification of pediatric fractures

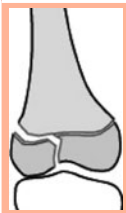
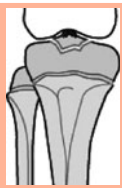
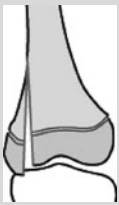
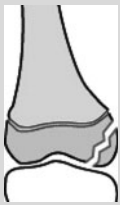
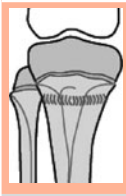
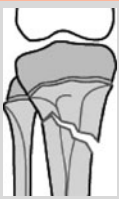
E = Epiphysis			
	E/3 S–H III		E/7 Ligament avulsions
	E/4 S–H IV		E/8 Flake fractures
			E/9 Other fractures
M = Metaphysis			
	M/2 Incomplete fracture (Torus/Buckle or greenstick)		
	M/3 Complete fracture		

Table 38.6. *(continued)* Classification of pediatric fractures

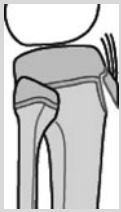
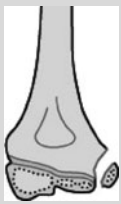
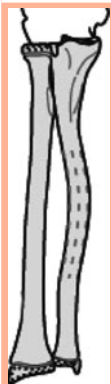
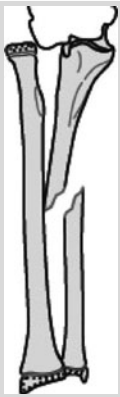
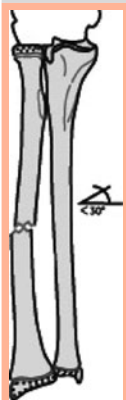
M = Metaphysis	
 	M/7 Ligament avulsion
	M/9 Other fractures
D = Diaphysis	
	D/1 Bowing fractures
	D/5 Complete oblique/ spiral fracture >30°

Table 38.6. *(continued)* Classification of pediatric fractures

D = Diaphysis	
 D/2 Greenstick fractures	 D/6 Monteggia lesions
 D/4 Complete transverse fractures	 D/7 Galeazzi lesions
	D/9 Other fractures

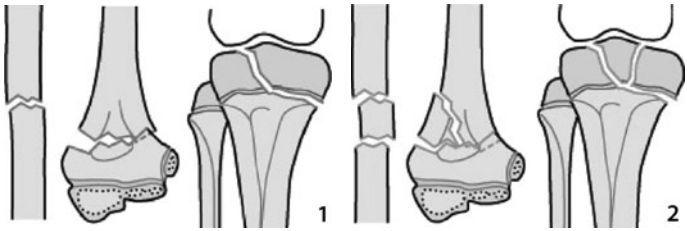
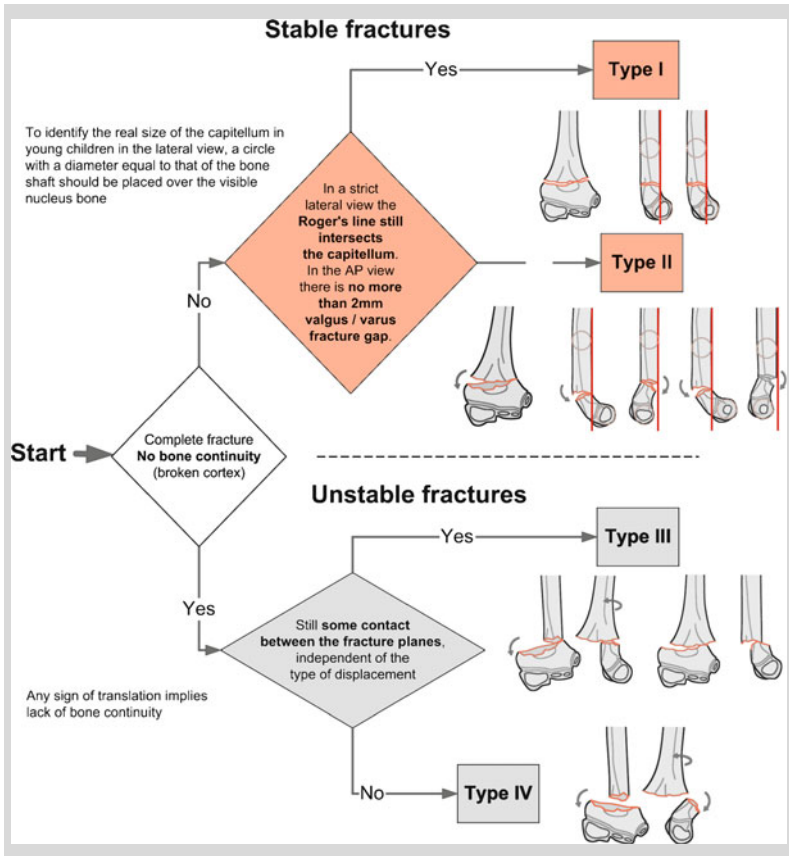


Fig. 38.2 Severity implies anticipated difficulties and method of treatment, not the prognosis

Table 38.7. Fracture displacement for specific fractures



Further rules for correct classification

- Fractures of the apophysis are recognized as metaphyseal injuries
- Transitional fractures with or without a metaphyseal wedge are classified as epiphyseal fractures
- Ligament avulsions
  - Intra-articular and extra-articular ligament avulsions are epiphyseal and metaphyseal injuries, respectively
  - The side of ligament-avulsion fractures of the distal humerus and of the distal femur is indicated by the small letter "u" (ulnar/medial) or "r" (radial/lateral) for the humerus and by "t" (tibial/medial) or "f" (fibula/lateral) for the femur
- Femoral neck fractures
  - Epiphysiolysis and epiphysiolysis with a metaphyseal wedge are coded as normal type-E epiphyseal Salter–Harris I and II fractures, E/1 and E/2
  - Fractures of the femoral neck are coded as normal type-M metaphyseal fractures coded from I to III
  - The intertrochanteric line limits the metaphysis

38.3 Skeleton Standard and Special X-rays

General remarks

- Not every fracture is visible on the X-ray in childhood
- An X-ray is only indicated if it has a therapeutic consequence
- If the decision to X-ray has been made, then it should be taken appropriately to provide the information desired
- It is obligatory to take X-rays in two planes, a.p. and lateral, even in emergency situations, including the proximal and distal joint of the broken segment
- X-ray of the opposite extremity is unnecessary, since no new knowledge or additional information can be gained
- A guide to radiological investigation of the skeleton is given in Table 38.8

Table 38.8 A guide to radiological investigation of the skeleton

Skull	
Skull a.p. and lateral	To see fractures
Skull Town's view	For occipital fractures and fractures of the foramen magnum
Spine	
Cervical, thoracic, lumbar a.p. and lateral	For a general overview
Cervical spine, oblique X-ray	Intervertebral foramina, small intervertebral joints
Cervical spine segmental functional X-ray	Intersegmental blocking, hypermobility by ligament avulsion
Ribs	
Thorax a.p. and lateral	Posterior rib segments
Thorax oblique	Anterior rib segments
Thorax tangential, special X-ray	For detail (after consultation with the radiologist)
Sternum	
Sternum lateral view	Special indications (funnel chest)
Shoulder	
Shoulder a.p. (under traction on both arms, 15 kg comparison of both sides)	Dislocation of the acromioclavicular joint
Shoulder in glenoid tangential projection	Overlapping free view of the humeral head and the glenoid joint ("true" a.p. view) Side difference indicates an injury of the acromion
Y-view (acromion, coracoid and the tangential view of the scapula together form the leg of the "Y")	Dislocation of the shoulder (needs a second view)
Shoulder transthoracal	Subcapital fractures of the humerus (second view needed)
Target X-ray	For special indications (suspicion of a Bankart lesion or Hill–Sachs lesion)

Table 38.8 *(continued)* A guide to radiological investigation of the skeleton

Elbow joint	
Elbow a.p. and lateral	Fractures
Elbow under imaging intensifier	For special indications (radial head, processus coronoideus)
Hand	
Hand dorso-palmar and lateral	Fractures and dislocations, swellings
Special view of the scaphoid (scaphoid quartet)	Fractures
Thorax	
Thorax a.p. and lateral	Aspiration, lung contusion, pneumothorax, heart, aortic arch silhouette
Pelvis	
Pelvis normal a.p.	Fractures
Hip joint axial	Fractures
Lauenstein X-ray of the ala of the ileum	Fractures of the ileum, the anterior wall
Obturator X-ray	Fractures of the anterior acetabular column, pubis and the posterior acetabular wall
Ankle joint	
A stress X-ray should not be taken. MRI shows ligament avulsions much better, however the diagnosis is of academic value since ligament avulsion is no longer treated surgically	
a.p. and lateral view	Fractures, joint incongruence
Foot	
Foot a.p.	Fractures
Foot in dorso-plantar oblique projection	Fractures
Foot in plantar-dorsal oblique projection (prone position of the child)	Medial tarsal bones (cuneiform medial and intermedium)
Foot lateral and axial	Calcaneus fractures

38.4 Development of Ossification Centers

- Figure 38.3 shows the normal development of ossification centers

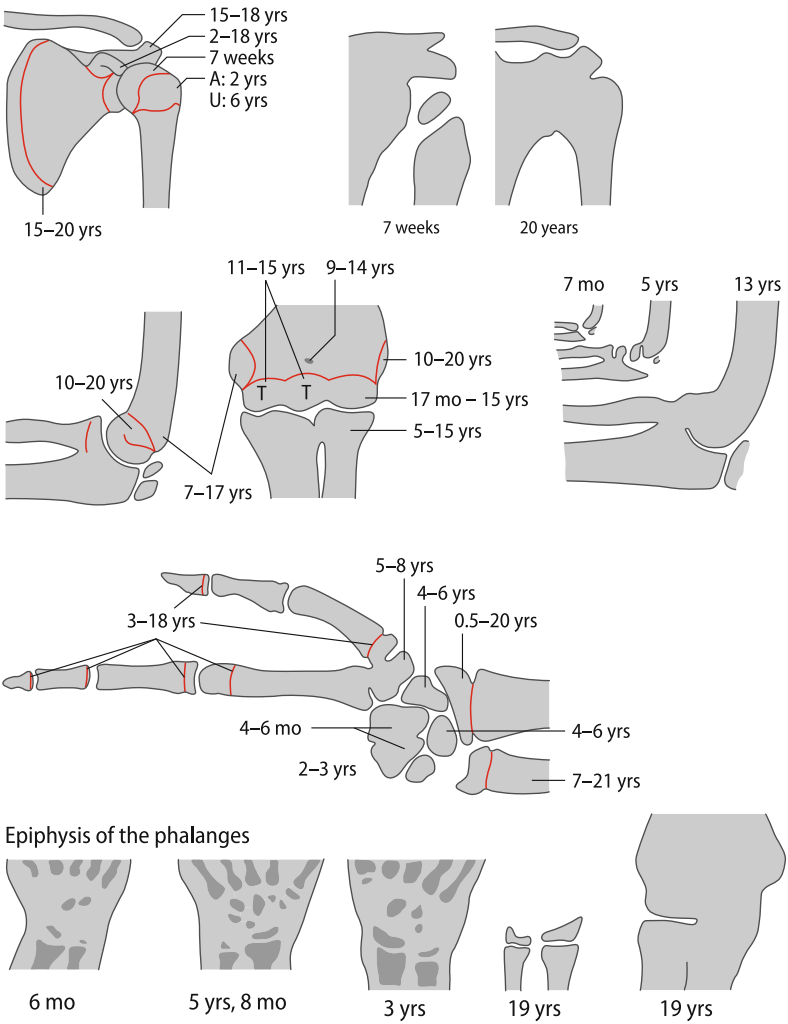


Fig. 38.3 Normal development of ossification centers. (*yrs* Years)

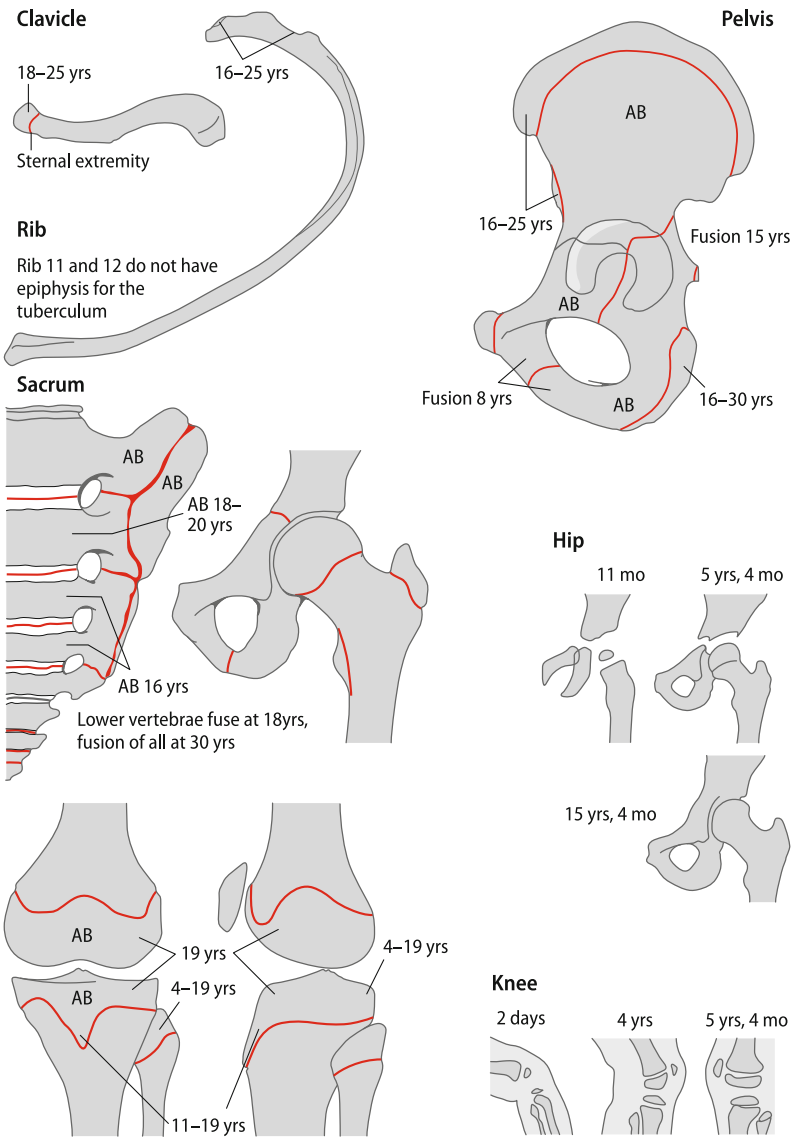


Fig. 38.3 *(continued)* Normal development of ossification centers. (*yrs* Years)

Vertebrae

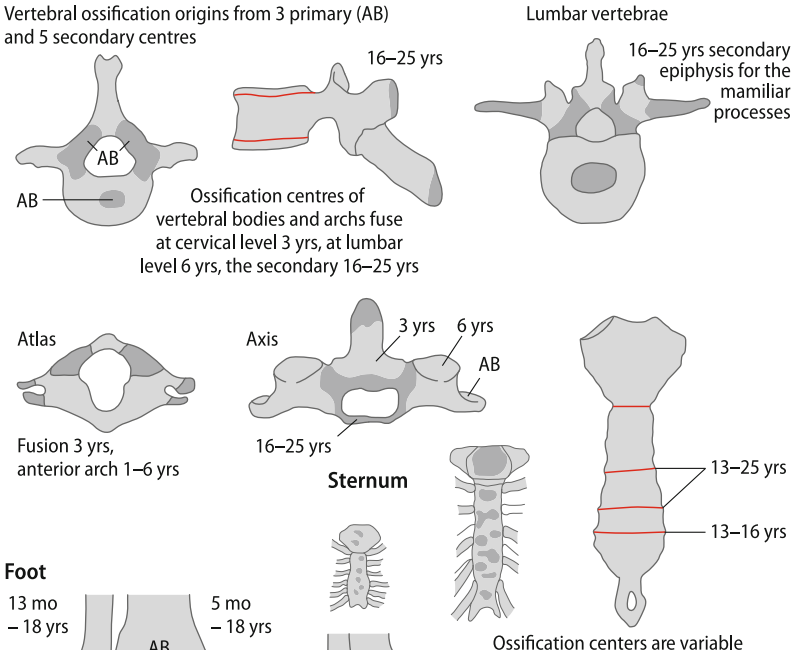


Fig. 38.3 *(continued)* Normal development of ossification centers. (*yrs* Years)

38.5 Plaster Immobilization

- Every physician who deals with fractures must know how to apply a cast

Plaster splint

- Indications
  - Fractures
  - Distortions
  - Pain after bone bruises
- The extremity must be covered with a cotton tube
- Wrapped with a thin half elastic cushion cotton
- Wrapped with a paper bandage
- Plaster gauzes are placed longitudinally along both sides of the extremity with a 3-cm plaster-free gap between them
- After edema regression the cast can be closed circularly

Circular plaster cast

- Indications
  - Functional treatment
  - Secondary post-plaster splint (after edema regression)
- Same steps as above
- The extremity is enclosed in a circular plaster
- Drying time for plaster ~1.5 h; for Scotch cast ~30 min
- A primarily applied circular plaster should be opened longitudinately, especially when the tissue is swollen after fresh injuries
- When swelling subsides, the plaster can be closed again

Plaster cast windows

- Indications
  - Open wounds
  - Pins
  - K-wires
- The earliest that windows can be made in the plaster is after drying

- The window in the plaster must always be covered with the piece that was taken out in order to prevent edema in this region
- The window in the plaster must not impair the stability

Plaster cast wedging

- Indications
  - Remaining angulation of an undisplaced fracture, after fracture stabilization without reduction
- The earliest the wedging of the plaster can be made is after 1 week. Swellings and pain must have gone completely
- The cut for the wedge must be at the deepest point of the concavity of the deformity
- The more peripheral the fracture, the more proximal should be the point for the wedge
- The child should not have any pain during this procedure

Radiological anatomy

- On X-ray, children's joints seem much greater than they really are (Fig. 38.4)
- The reason for this is the thick cartilage around the epiphysis

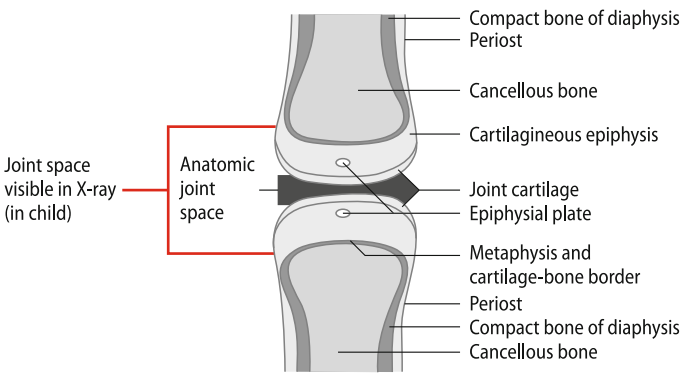
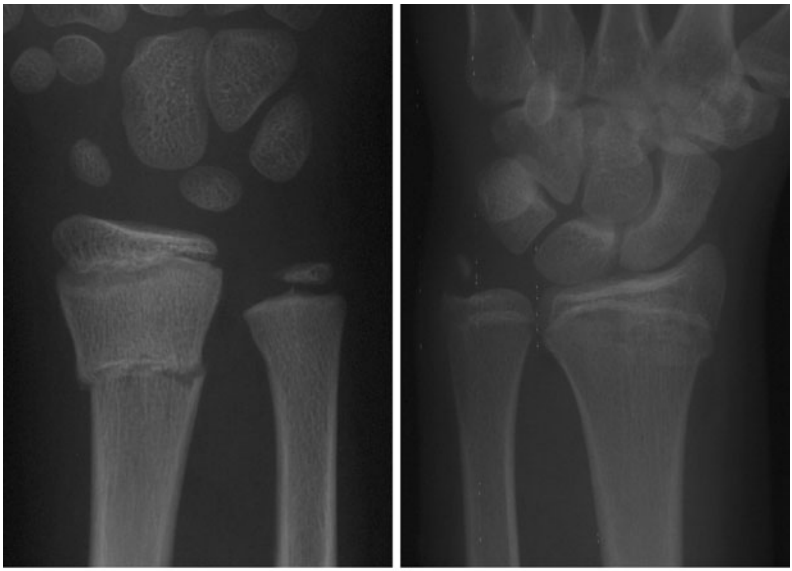


Fig. 38.4 Radiological anatomy of the joint in childhood.



a b

Fig. 38.4 *(continued)* Radiological anatomy of the joint in childhood. **a** 4 years old child, **b** 14 years old child

38.6 Therapy Principles

Closed reduction (mainly used for the upper extremity)

- Every fracture reduction should be carried out under anesthesia (plexus block, general anesthesia)
- Extend the fingers, hang a weight at the humerus and maintain the upper extremity with 90° flexion at the elbow for 20–30 min
- Reduce the fracture using a reduction maneuver that mimics the movement that led to the fracture in the first place
- Apply the plaster in the hanging position
- Immobilization of the fracture with a dorso-volar plaster splint

So-called semi open reduction

- Indication
  - Partial or total unstable displaced fractures that can be definitively reduced
- Closed reduction under sterile conditions as described above
- Followed by percutaneous K-wire fixation
- In addition, immobilization with a plaster splint is required

Extension

- Indications
  - Limited nowadays
  - The main indication is in fractures of the femur in children between birth and 3 (4) years of age, depending on the child's weight of the child; known as overhead extension
- Sufficient analgesia and sedation of the child
- First the plaster is placed on the healthy leg over its full length
- Then the injured leg is covered with plaster, however only distal to the fracture
- Fixation of the extension on the overhead arch so that the child's buttocks are raised (there should be space for a flat hand to move freely under the buttocks without touching them)

Osteosynthesis with K-wires

- Indications
  - Closed or open reduced metaphyseal fractures
  - Fractures of the hand and foot
- Fixation with K-wires is not suitable for stabilization of diaphyseal fractures
- Whenever possible, the K-wires should be placed percutaneously so that they can be removed without anesthesia
- The crossing points of the K-wires should be proximal to the fracture line
- If the epiphysis must be crossed, repeated attempts to fix it should be avoided
- In cases such as this, thin K-wires should be used instead

- Ensure that the K-wires penetrate the opposite cortex
- Daily care of the pins reduces the risk of infection
- Normally K-wires can be removed after 3–4 weeks

Osteosynthesis with external fixator (Ex-Fix)

- Indications
  - Comminuted fractures mainly in older children (femur, tibia, forearm)
  - Polytrauma
  - Long spiral fractures, e.g., spiral wedge of the femur in older children
- Normally closed reduction with or without extension table
- Type: Monotube®, tubular system, circular frame
- Place the Schanz screws under imaging intensifier
- In Monotube® systems, the distances between the entry points are predefined
- In "frame systems" one entry point should be near the fracture, the other further away from it
- All clamps must be open for reduction
- After sufficient reduction all clamps have to be closed and secured
- Daily care of the pins reduces the infection risk

Elastic stable intramedullary nailing (ESIN)

- Operation technique: see Sect. 38.15
- Indications
  - Transverse, oblique and short spiral, diaphyseal fractures in childhood (3–15 years)
- ESIN is minimally invasive, minimally traumatic, and sufficiently stable for movement and partial weight bearing; it is biologically and child friendly way, using special elastic nails, of enabling osteosynthesis

Lag screw osteosynthesis

- Indication
  - Articular and peri-articular fractures, Salter–Harris II fractures, mainly in the distal tibia and femur, femoral neck fractures

- Self-drilling and taping cannulated screws (dimensions 4/4.5/6.5 mm) are mainly used
- Directly position the extremity on the intensifier
- Rotate the extremity so that the fracture line is visible in a proper a.p. view
- Put the guide wire on the fragment parallel to the table of the intensifier
- Drill the guide wire into the bone to the contralateral cortex
- Measure the length
- Put the correct-sized cannulated screw over the guide wire
- Tighten the screws until the fracture is closed

Plate osteosynthesis

- Indications
  - Comminuted fractures, mainly in older children (femur, tibia, forearm), long spiral fractures with/without spiral wedge of the femur in older children
- Usage of plates is reserved to exceptions and in special cases
- When an osteosynthesis with a plate is indicated, we recommend the application of new types of plates such as LC-DCP and LCP plates. If possible, these types of plates can be applied using a minimally invasive technique (MIPO)

Possibilities for osteosynthesis

- Possiblities for osteosynthesis are illustrated in Fig. 38.5

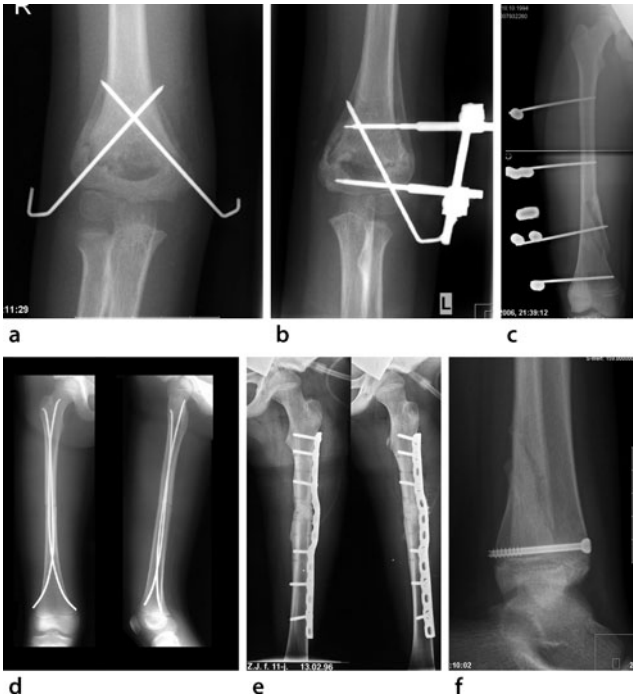


Fig. 38.5a–f Possibilities for osteosynthesis. **a** Kirschner wire, **b** combination Kirschner wire/external fixation, **c** external fixator, **d** elastic stable intramedullary nailing, **e** plate osteosynthesis, **f** lag screw osteosynthesis

- **Knee joint puncture (Fig. 38.6)**
  - Indications
    - Posttraumatic hemarthros (not before 24 h!)
    - Signs or suspicion of infection
  - Procedure
    - Apply local anesthesia or anesthetic cream well before the puncture
    - Puncture in the lateral proximal recess

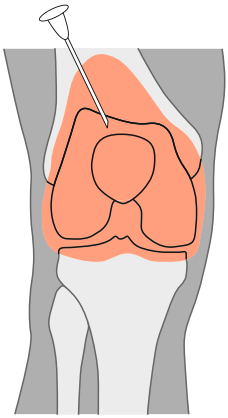


Fig. 38.6 Knee joint puncture

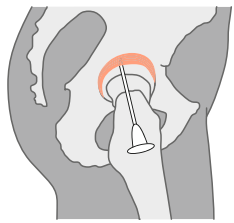


Fig. 38.7 Hip joint puncture

- **Hip joint puncture (Fig. 38.7)**
  - Indications
    - Posttraumatic hemarthros (not before 24 h!)
    - Sings or suspicion of infection
  - Procedure (three different approaches)
    - From lateral approach
    - From anterior approach (take care to avoid the artery and nerve!)
    - Ludloff approach (preferred by author)

Period of healing

- Guidelines for fracture immobilization in childhood are given in Table 38.9
- Metaphyseal fractures heal in half the time of the diaphyseal fractures
- Diaphyseal transverse fractures heal more slowly than diaphyseal oblique fractures

Table 38.9 Guidelines for fracture immobilization in childhood

Fracture	Period of immobilization (weeks) at age		
	<5 years	5–10 years	> 10 years
Clavicle	1	2	2–3
Humerus			
Proximal stable	1	2–4	3-4
Proximal unstable	1	2–4	3-4
Humerus shaft	2–3	3–4	4-6
Supracondylar	2–3	3–4	4-5
Radial condyle	3	3–4	4
Ulnar condyle/Y–fracture	2–3	3–4	3-4
Ulnar epicondyle (+dislocation of the elbow)	2–3	2–3	4
Radius proximal	1–2	2–3	3–4
Olecranon	2	2–3	3–4
Forearm shaft including greenstick fracture	3	4	4–6
Radius (+ radius + ulna) distal	2–3	3–4	4–5
Salter–Harris I radius distal	2	2–3	3–4
Carpal		4–6	5–8
Metacarpal			
Proximal and distal		2–3	3-4
Metacarpal shaft		3–4	4-6
Fingers			
Proximal and distal	1–2	2–3	3-4
Finger shaft	2–3	3–4	4-6
Femur			
Neck of the femur		4–6	6-8
Subtrochanteric fractures	2–4	4–5	5-6
Shaft	2–3	4–5	4-6
Distal	2–3	3–4	4-5
Tibia and lower leg			
Tibial spine fracture		3–4	4-6
Proximal metaphysis	2–3	3–4	4-5
Shaft	2–3	3–5	4-6
Distal and malleolar	2–3	3–4	4-5

Table 38.9 *(continued)* Guidelines for fracture immobilization in childhood

Fracture	Period of immobilization (weeks) at age		
	<5 years	5–10 years	> 10 years
Hind foot and calcaneus		4–6	5–10
Mid-foot and toes distal	2–3	3–4	4
Toes	1–2	2–3	3–4
Fibulotalar ligaments/osseous avulsion		3–4	4–6

Source: von Laer L (1991) Fractures and dislocation during growth. *Injury* **36**(2):356.

38.7 Spinal Injuries

General considerations

- Spinal injuries in childhood are not as rare as one assumes
- The most common injuries are simple fractures after anterior crush injuries or compression fractures of the vertebral body
- In injuries involving the atlas and axis, rotatory subluxation or dislocation is one of the most common lesions in children, rather than fractures of the atlanto-axial articulation
- Injuries of the thoracic and lumbar spine are more common in childhood than in adulthood. The majority of these fractures result from traffic accidents. Child abuse should also be considered
- Obstetric fractures involve mostly the cervical spine (with high mortality)
- End-plate fractures in younger children are followed by scoliosis
- Fractures of vertebral bodies have good remodeling capacity, depending of the child's age
- Diagnosis can be difficult due to congenital malformations (congenital non-union, hemivertebra, congenital vertebral fusions) or following diseases (post-traumatic malformations or Scheuermann's disease)
- The normal spine in children differs considerably from that in adults, especially in the cervical region

Classification

- Stable spine fractures with compression of the vertebral body or "end-plate" injuries
- Unstable fractures with involvement of the vertebral arch and the pedicles
- Ligament avulsion

Or

- Fractures involving end-plates with growth disturbance
- Fractures not involving end-plates

Investigation

- Diagnosis includes accurate evaluation of the level and extent of injuries to both chondro-osseous and nervous system tissues
- Radiographic evaluation must be carried out paying due attention to potentially severe unstable injuries and must include prior adequate immobilization of the spine
- Osseous injuries can be seen on an adequate a.p. and lateral view
- Oblique views may be necessary
- CT scan or MRI may be indicated

Therapy

- A number of unique problems can be encountered in the treatment of infants, children, and adolescents with spine injuries
- In any closed, nonoperative treatment regime, the spinal deformity must be reduced and adequately stabilized, and protected from re-displacement during the healing process
- Treatment guidelines are given in Table 38.10

Table 38.10 Treatment guidelines for spinal injuries in infants, children, and adolescents

Fracture	Therapy
Stable/non-displaced	Simple bed rest is indicated, since most children with stable (compression) fractures are asymptomatic within a few days or weeks. External support may be necessary
Stable displaced	Surgical stabilization with dorsal fusion
Unstable	Surgical stabilization with dorsal fusion

Prognosis

- The prognosis of undisplaced stable fractures is good, depending on the type of injury (growth plate/end-plates, wedge compression)
- The prognosis of displaced fractures depends on the accompanying neurological problems

38.8 Sling-Shot Injury of the Cervical Spine

General considerations

- Sling-shot injury of the cervical spine happens more frequently than fractures in childhood

Investigations

- Normal X-ray indicating a straightened position of the cervical spine

Therapy

- Pain management
- Immobilization with a soft ruff
- Physiotherapy

Prognosis

- Good

38.9 Pectoral Girdle and Clavicle

General considerations

- Fractures of the clavicle (Table 38.11) occur most frequently in children during the first 10 years
- Fractures of the scapula, which is highly mobile and well protected by muscles, are rare
- Fractures of the scapula result from direct violence, such as crushing injury, and traffic accidents

Table 38.11 Fractures of the clavicle

Morphology	Fractures of the mid-shaft are most common and range from greenstick to complete fractures
Signs	Pain, swelling, painful movement of the arm
Diagnosis	Clinically and radiologically
Correction potential	Good
Complications	Nerve problems, non-union, cosmetics
Nonoperative therapy	In principle non surgical → sternal brace or "figure-of-eight" orthesis pulling the shoulder backwards
Operative treatment	Only fully displaced, comminuted fractures in adults (author's preferred method is ESIN)
Immobilization	2–3 weeks
X-ray control	After 3 weeks
Follow-up	Only clinically

Gleno-humeral joint dislocation

- Subluxation and dislocation of the shoulder (Table 38.12) are rare in infants or young children
- The capsule of the shoulder joint has some intrinsic laxity that allows some displacement during stress
- "Dislocation" of the shoulder has also been described as a birth injury. However, great care should be taken before making such a diagnosis, as the proximal humerus is most likely to be fractured through the epiphysis

Table 38.12 Gleno-humeral joint dislocation

Morphology	Injury of the adolescent, appearing, in contrast to epiphysiolysis, in the form of a head dislocation after closure of the proximal epiphysis
Signs	Visible deformity, empty glenoid
Diagnosis	Clinical and radiography
Correction potential	None
Complications	Lesions of the axillary nerve
Nonoperative therapy	Immediate reduction and Gilchrist dressing
Operative treatment	There are no good operative methods in childhood
Immobilization	3 weeks
X-ray control	None
Follow-up	No control necessary

Prognosis

- For clavicle fractures, very good
- Prognosis of dislocation of the gleno-humeral joint depends on the time lapsed since the incident and the type of injury

38.10 Humeral Fractures

General information

- Rare in childhood, following direct trauma or obstetric injury
- Mostly transverse or oblique fractures

Fractures of the proximal humerus

- Proximal humeral fractures and their management are detailed in Table 38.13

Table 38.13 Proximal humeral fractures and their management

Morphology	About 60% are subcapital fractures; 38% are Salter–Harris II fractures; pure epiphysiolysis are rare
Signs	Deformation, pain
Diagnosis	X-ray; interpretation is often difficult in undisplaced fractures, displacement of the epiphyseal line is interpreted as a fracture Note the three ossification centers
Correction potential	Great potential, angulation in the sagittal and frontal plane is tolerated up to 60° in children <12 years old and up to 30° >12 years
Complications	Practically unknown; in neonates premature close of the growth plate is possible
Nonoperative therapy	Stable, undisplaced fracture, any age Stable fracture with angulation < 60°: < 10 years < 30°: > 10 years Or Stable fracture with tolerable displacement, any age Immobilization for 3–4 weeks in a Desault or Gilchrist dressing Or If anesthesia is needed for reduction, definitive treatment with stable fixation is recommended
Operative treatment	▪ Children >10–12 years of age ▪ Unstable fracture if a reduction under anesthesia is necessary ▪ Major displacement after nonoperative treatment ▪ Author's preferred method: ESIN ▪ No additional immobilization is needed ▪ Nail removal after 3–4 months
Immobilization	3 weeks (see above)
X-ray control	Nonoperative therapy: days 3–4 and weeks 3–4 Operative treatment: postoperatively and week 4
Follow-up	Week 3 or 4 radiological and clinical

Operative treatment

- Author's preferred method
  - ESIN from a monolateral, radial approach in an ascending technique (see Table 38.14 for corresponding operative technique)
  - Alternative: percutaneous (2.5-mm threaded) K-wire fixation
  - Open reduction is a rare exception

Table 38.14 Supracondylar humerus fracture

Morphology	See Table 38.5
Signs	Swelling, pain, visible deformation
Diagnosis	X-ray in two planes Classification: ▪ No displacement ▪ Displacement in one plane ▪ Displacement in two planes ▪ No bone contact
Correction potential	Practically nonexistent
Complications	▪ Radial (medial) nerve injury (deep branch) ▪ Premature closure of the growth plate after repeated drilling ▪ Varus deformity as a consequence of a rotational failure
Nonoperative therapy	Classification: type I and II ▪ Blount loop ▪ Dorso-volar plaster splint in 90° position
Operative treatment	Classification: type III and IV ▪ Closed reduction (in 90%–95% is possible) ▪ Percutaneous K-wire fixation (ascending crossed bilateral, parallel radial, ascending or descending monolateral radial) ▪ Small external radial fixator (method preferred by author) ▪ ESIN
Immobilization	Operative and nonoperative treatment; 3–4 weeks of plaster fixation Removal of the percutaneous K-wires at this time
X-ray control	Nonoperative treatment: after a few days and at week 3–4, depending of the child's age Operative treatment: after 3–4 weeks
Follow-up	2–3 months after injury functional, clinical examination No physiotherapy

Fracture of the humerus shaft (diaphyseal fractures)

- Fractures of the humeral shaft and their management are detailed in Table 38.15

Prognosis

- Very good

Table 38.15 Fracture of the humerus shaft (diaphyseal fractures) and their management

Morphology	Rare fractures
Signs	Deformity, pain
Diagnosis	X-ray (two images taken at 90° to one another)
Correction potential	There is a great potential in all planes
Complications	Damage to the radial nerve (long spiral fractures of the distal third)
Non operative therapy	Stable undisplaced fracture, any age Stable fracture with angulation <30° Or Stable fracture with tolerable displacement, any age Immobilization for 3–4 weeks in a Desault or Gilchrist dressing Or If anesthesia is needed for reduction, definitive treatment with stable fixation is recommended
Operative treatment	▪ Children >10–12 years of age ▪ Radial nerve irritation is not an indication for surgical intervention ▪ Unstable fracture if a reduction under anesthesia is necessary ▪ Major displacement after nonoperative treatment ▪ Author's preferred method: ESIN ▪ No additional immobilization is needed ▪ Nail removal after 3–4 months
Immobilization	3–4 weeks
X-ray control	Nonoperative therapy: days 3–4 and weeks 3–4 Operative treatment: postoperatively and week 4
Follow-up	Week 3 or 4 radiologically and clinically

38.11 Elbow Joint Region Fractures

General considerations

- Mostly children between 3 and 10 years old sustain these fractures
- Good knowledge of the child's anatomy of the distal humerus and the proximal forearm is imperative
- The X-ray is often difficult to interpret, nevertheless an X-ray of contralateral, uninjured site is unnecessary and no longer required
- Correct diagnosis should always be achieved before starting treatment

- Special fractures
  - Non-displaced fractures of the lateral condyle
  - Isolated fractures of the radial neck
  - Monteggia fractures
  - Rotation failures in supracondylar humerus fractures due to varus or valgus deformity resulting from the fracture

Classification

- Fractures in the region of the elbow joint are discussed in Tables 38.14, 38.16–38.26

Table 38.16 Regarding the joint

Articular	Fractures of the lateral condyle Transcondylar fractures of the humerus
Extra-articular	Supracondylar fracture of the humerus Epicondylar fractures

Table 38.17 Regarding the direction of displacement of the distal fragment

Fractures in extension	In 95% of all cases, the distal fragment is displaced dorsally
Fractures in flexion	The distal fragment is displaced ventrally only in 5%

Table 38.18 Localization of elbow fractures according to frequency

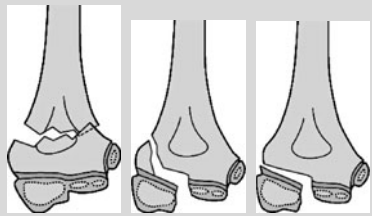
Supracondylar fractures (*a*)

Transcondylar fractures

Lateral condyle (*b1,2*)
Medial condyle (very seldom in childhood)

Fractures of the proximal radius (*e*)

Dislocation of the elbow joint



Ligament avulsions

Lateral epicondyle (*c*)
Medial epicondyle (*d*)

Olecranon fractures (*f*)

Monteggia fractures (isolated fracture of the ulna and dislocation of the radial head)

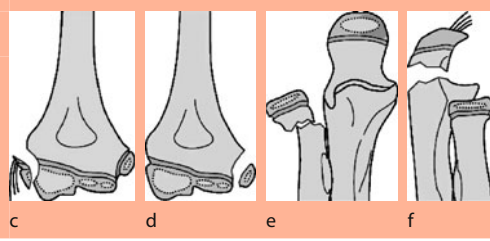


Table 38.19 Epiphysiolysis of the distal humerus

Morphology	Very rare fracture
Signs	Pain, deformity
Diagnosis	X-ray
Correction potential	None
Complications	Same as for supracondylar fractures (see Table 38.18)
Nonoperative therapy	Dorso-volar plaster splints
Operative treatment	If a reduction is indicated: fixation with crossed K-wires
Immobilization	3 weeks
X-ray control	Day 7 (nonoperative) 3–4 weeks end control
Follow-up	2 months

Table 38.20 Transcondylar, intercondylar fractures

Morphology	The entire distal epiphysis of the humerus is displaced posteriorly, laterally or forwards, depending on the injury mechanism. The most frequent fracture is that of the lateral condyle
Signs	Pain, swelling
Diagnosis	X-ray in two planes. Sometimes only the oblique view will disclose either displacement or evidence of the undisplaced fracture line
Correction potential	None
Complications	Delayed healing and blocked union with varus deformity, late ulnar nerve irritation, avascular necrosis of the capitulum
Nonoperative therapy	▪ Initial undisplaced fractures (long arm cast) followed by a cast-free X-ray control on day 4–5 ▪ Secondary displacement over 2 mm needs surgical intervention
Operative treatment	Initial displacement over 2 mm (open reduction and K-wire or screw fixation) Implant removal after 8-12 weeks
Immobilization	4 weeks
X-ray control	Undisplaced fractures day 4–5, cast free!! Consolidation is visible after 4–5 weeks
Follow-up	6 months and 1 year

Table 38.21 Proximal radial fractures

Morphology	65% subcapital = metaphyseal fractures of the radial neck 35% Salter–Harris II fractures
Signs	Pain, blockage of pronation and supination
Diagnosis	X-ray in two planes, sonography
Correction potential	None in lateral displacement Good in the sagittal and frontal plane up to 60°
Complications	Avascular necrosis, malunion or non-union, premature fusion of the growth plate, ectopic calcification, limited pronation and supination
Nonoperative therapy	<10 years of life up to 60° (long arm cast)

Table 38.21 *(continued)* Proximal radial fractures

Operative treatment	If anesthesia is needed for reduction Closed reduction by indirect manipulation Fixation with ESIN Trick: the fully displaced radial head can be manipulated by fixing the fragment with a percutaneous K-wire, and thereby moved to the right location (Joy-stick technique)
Immobilization	Nonoperative therapy: 2–3 weeks, then functional therapy Operative treatment: no immobilization is required
X-ray control	Nonoperative therapy: day 4 and 8 and after 3 weeks Operative treatment: only after 4 weeks
Follow-up	Clinical controls for 2 years after accident

Table 38.22 Dislocation of the elbow

Morphology	Mostly in children over 8 year of age Displacement direction correlates with deforming force direction
Signs	Deformity, pain, swelling, nerve irritation
Diagnosis	Clinically and X-ray The differential diagnosis of an elbow dislocation basically consists of distinguishing a dislocation from a supracondylar fracture, a lateral condylar fracture, or a transcondylar fracture
Correction potential	None
Complications	Medial ligament avulsion, fracture of the medial epicondyle Vascular and nerve complications
Nonoperative therapy	Immediate reduction of an acute posterior dislocation may often be accomplished without general anesthesia Dislocation of the radial head during this maneuver Long arm cast for 3 weeks, then functional therapy
Operative treatment	Re-fixation of the medial epicondyle with K-wire or screw Reconstruction of the medial and lateral ligaments if the elbow is unstable
Immobilization	3 weeks
X-ray control	Only after surgery in week 4
Follow-up	6 weeks and 6 months

Table 38.23 Fracture of the medial or lateral epicondyle

Morphology	Fracture of the medial or lateral epicondyle (nearly always as a result of an elbow dislocation)
Signs	Swelling, local pain (lateral or medial)
Diagnosis	X-ray in two planes
Correction potential	None
Complications	Non-union
Nonoperative therapy	Only undisplaced fractures Look for secondary displacement on day 3–4
Operative treatment	Displaced (> 2 mm) fractures: open reduction and K-wires or screw fixation Secondary displaced fractures after nonoperative therapy in the control
Immobilization	3–4 weeks, long arm cast
X-ray control	Undisplaced, cast free on day 3–4 Operated fractures in week 4
Follow-up	2–3 months after injury

Table 38.24 Olecranon fractures

Morphology	Olecranon fractures are usually undisplaced and incomplete, particularly in younger children Often seen in combination with other injuries
Signs	Swelling and pain, elbow in flexion
Diagnosis	X-ray in two planes, sometimes very difficult, especially in young children (absence of ossification centers)
Correction potential	None
Complications	Restricted movement
Nonoperative therapy	Long arm cast for undisplaced fractures
Operative treatment	Longitudinal pinning and cerclage wire fixation in dislocated fractures
Immobilization	Long arm cast for 4 weeks
X-ray control	Nonoperative therapy: days 5–6 and at week 4 Operative treatment: at week 4
Follow-up	Clinical control at week 8

Table 38.25 Subluxation of the radial head (Chassaignac)

Morphology	Mostly due to traction on the forearm in 1- to 3-year-old children Subluxation of the radial head
Signs	Painful pronation, elbow in extension
Diagnosis	Clinically, history
Correction potential	Good
Complications	Neglected fracture of the radial neck, persistent dislocation
Nonoperative therapy	Elbow in flexion → fast supination and extension → the click is noticeable
Operative treatment	Indicated only in neglected cases
Immobilization	None
X-ray control	None
Follow-up	None

Table 38.26 Monteggia lesions

Morphology	Fracture of the proximal third of the ulna in association with dislocation of the radial head This injury has to be suspected whenever a fracture anywhere along the ulna (bowing, greenstick injury) is visible without an obvious associated fracture of the radius
Signs	Swelling and pain, elbow in flexion
Diagnosis	X-ray in two planes including both wrist and elbow joints
Correction potential	None
Complications	Restricted movement, persistence of radial head dislocation
Nonoperative therapy	Long arm cast in undisplaced fractures with correctly centered radial head, towards the center of the humerus in lateral and a.p. X-ray view
Operative treatment	Displaced fractures with dislocated radial head ▪ Closed reduction of the fracture and radial head dislocation ▪ Fixation of the ulna with ESIN (method preferred by author) Secondary dislocation after nonoperative therapy
Immobilization	4 weeks
X-ray control	Nonoperative therapy : days 5–6 and at week 4 Operative treatment: at week 4
Follow-up	Clinical control at week 8

38.12 Forearm Fractures

General considerations

- Diaphyseal injuries are common in children
- The severity may vary from pure bowing (Table 38.27) to greenstick (Table 38.28) or even to a complete fracture (Table 38.29) with or without displacement
- The level of the fracture varies
- In children, the tendency for the radius and ulna fractures to be aligned is greater than in adults
- Great fracture variability
  - Same fracture type in both bones, but not aligned
  - Isolated fracture of the radius
  - Bowing of radius and ulna (Table 38.27)
  - Fracture of the ulna and bowing of the radius or reversed
- A Galeazzi fracture is the name given to fracture of the distal radius with dislocation of the distal radio-ulnar joint (Table 38.30)

Table 38.27 Bowing fracture of the forearm

Morphology	Plastic deformity of the shaft without fracture of the cortex → microfractures
Signs	Pain, deformity, restricted movement
Diagnosis	X-ray, two planes
Correction potential	None
Complications	Restricted movement (pronation and supination), re-fracture
Nonoperative therapy	<20° bending → long arm cast, cast wedging (only if necessary), no anesthesia
Operative treatment	>20° bending → closed indirect reduction and stabilization with ESIN technique
Immobilization	Nonoperative therapy → 4 weeks Operative treatment → immobilization is not required
X-ray control	At weeks 4
Follow-up	Over 1 year

Table 38.28 Greenstick fracture of the forearm shaft

Morphology	Plastic deformity of the shaft with one-sided cortex fracture
Signs	Pain, deformity, restricted movement
Diagnosis	X-ray, two planes
Correction potential	None
Complications	Restricted movement (pronation and supination), re-fracture
Nonoperative therapy	<20° bending → long arm cast, eventually cast wedging, no anesthesia
Operative treatment	>20° bending → closed indirect reduction and completion of the fracture → stabilization with ESIN technique
Immobilization	Nonoperative therapy → 4 weeks Immobilization is not required after operative treatment
X-ray control	At week 4
Follow-up	Over 1 year

Table 38.29 Complete diaphyseal forearm fracture

Morphology	Both cortexes are fractured, with or without displacement
Signs	Pain, swelling, deformity
Diagnosis	X-ray in two planes
Correction potential	Partial, 10°–15°
Complications	Mal-union with restricted movement, re-fracture
Nonoperative therapy	Only nondisplaced, stable fractures, using a well-molded cast with three-point fixation Angulation <15° can be treated by cast wedging
Operative treatment	All displaced unstable fractures at any age Failure of retention in nonoperative therapy (ESIN with the possibility of reducing the fracture by making a small incision at the level of the fracture in about 10% of cases)
Immobilization	5–6 weeks for nonoperative therapy No immobilization when ESIN is used
X-ray control	Nonoperative therapy: days 6–7 and weeks 5–6 ESIN: weeks 5–6 and before nail removal
Follow-up	6 months

Table 38.30 Forearm fractures – distal third (metaphyseal fractures)

Morphology	▪ Metaphyseal torus or buckle fractures ▪ Metaphyseal bowing and greenstick fractures ▪ Complete metaphyseal fractures with or without displacement ▪ Salter–Harris I and II fractures
Signs	Pain, swelling, deformity (medial nerve irritation)
Diagnosis	X-ray in two planes
Correction potential	Extremely good, children in <10 years, up to 50°
Complications	Correctly treated, practically none Sometimes an overgrowth of the radius is possible as well as premature closure of the growth plate
Nonoperative therapy	Long arm cast immobilization for torus, greenstick and bowing fractures without reduction Complete fractures should be reduced under general anesthesia since muscle relaxation is an essential part of the reduction In the hands of an experienced surgeon 95% of all fractures can be reduced and stabilized nonoperatively
Operative treatment	Only complete unstable fractures of the distal radius in older or adolescent children need surgical stabilization (K-wires or external fixator) Plate fixation is an exception
Immobilization	3–4 weeks for the majority No immobilization for external fixator or plating
X-ray control	Days 6–7 and weeks 3–4
Follow-up	If there is a malunion at consolidation

Treatment of forearm fractures without anesthesia

- Place child in supine position (see Fig. 38.8)
- Place upper extremity in 90° abduction on the edge of the table with their elbow in 90° flexion
- Cover the whole arm with a stocking
- Elevate the forearm using finger traps
- A counterweight is attached across the upper arm with the elbow at 90° flexion, to an extent that the child can still tolerate
- This position is maintained for 15–20 min
- Reduce the fracture by pressing both hands together to stretch the interosseous membrane

- X-ray control
- Apply a well padded dressing
- Apply a well molded long arm cast (must be opened after drying) or dorso-volar long arm splint (author's preferred method)

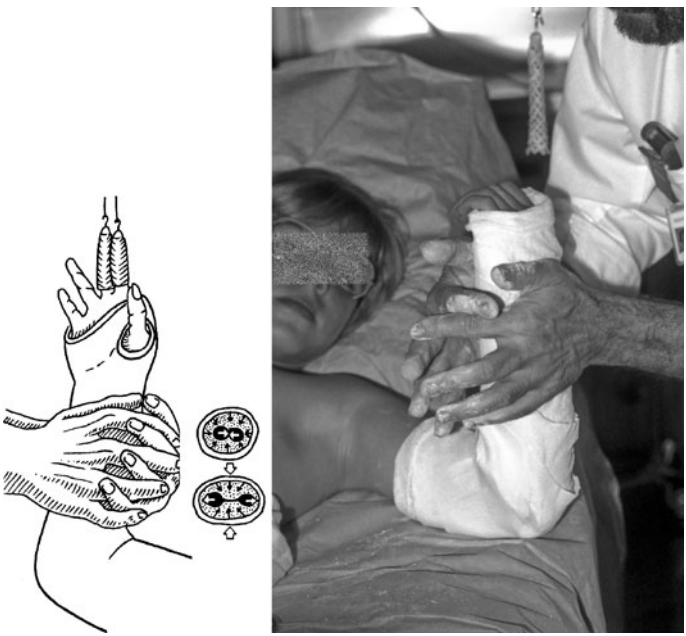


Fig. 38.8 Treatment of forearm fractures without anesthesia

38.13 Wrist and Hand Fractures

General considerations

- Whereas carpal injuries and multiple unstable fractures of the metacarpals are rare in children, other hand and finger fractures are frequent in children, especially phalangeal fractures and interphalangeal dislocations
- Fractures of the scaphoid are rare in children under 12 years of age. The treatment is nonoperative with a scaphoid cast for 6 weeks

- Displaced fractures are treated operatively, similar to fractures in adults
- Diagnosis can be difficult and often requires special X-ray techniques
- Fractures of the metacarpals are the most frequent hand fractures
- Each metacarpal has only one epiphysis; for MC I it is proximal (Table 38.31), for MC II–V, distal (Table 38.32)
- Hence, more proximal fractures can be found on the MC I and more distal fractures on the MC II–V

Table 38.31 Fracture of the first metacarpal

Morphology	Mostly metaphyseal torus fractures or Salter–Harris I and II fractures; shaft fractures are rare
Signs	Pain, deformity
Diagnosis	X-ray in two planes
Correction potential	Possible in all planes, exception the frontal plane
Complications	Premature closure of the growth plate
Nonoperative therapy	Undisplaced metaphyseal and diaphyseal fractures treated with a forearm cast without reduction
Operative treatment	Displaced metaphyseal fractures, closed reduction and ± K-wire fixation Displaced diaphyseal fractures, closed reduction ± osteosynthesis (mini-ESIN, author's preferred method)
Immobilization	Proximal fractures: 2–3 weeks Shaft fractures: 3–4 weeks, independent of the fixation
X-ray control	Non operative therapy: day 4 and week 3 Operative treatment: week 4
Follow-up	End of treatment

Table 38.32 Fracture of metacarpals II–V

Morphology	Proximal fractures are rare and mostly undisplaced Subcapital fractures are more frequent, especially metacarpal V
Signs	Pain, deformity When the metacarpal or phalanx bones are involved, the uniform plane of the fingernails is disrupted, and the finger affected overlaps the others
Diagnosis	X-ray in two planes

Table 38.32 *(continued)* Fracture of metacarpals II–V

Correction potential	Very good Remodeling is never capable of correcting a rotational deformity of the fingers
Complications	Axial deviations
Nonoperative therapy	Undisplaced basal fractures and fractures of the shaft Well-fitting plaster cast or splint or "Iselin splint" without reduction
Operative treatment	Displaced proximal fractures: closed reduction ± K-wire fixation and plaster cast Displaced shaft fractures: closed reduction ± plaster cast or mini-ESIN
Immobilization	Proximal fractures: 2–3 weeks Shaft fractures 3–4 weeks, independent of the fixation
X-ray control	Nonoperative: day 3 or 4 and weeks 3–4 Operative: weeks 3–4
Follow-up	End of treatment

38.14 Pelvic Ring Fractures

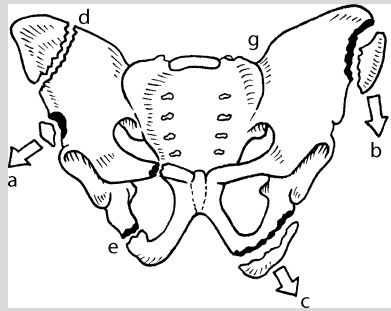
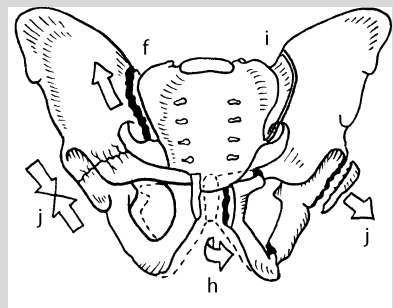
General information

- Pelvic fractures are rarely isolated; most appear as accompanying injuries after heavy direct or multiple trauma
- The degree of severity ranges from a simple fracture of the pubic bone to a complete pelvic ring disruption

Classification

- The classification of pelvic ring fractures is given in Table 38.33

Table 38.33 Classification of pelvic ring fractures

Fractures of the pelvic ring without delayed deformities (stable fractures)	
Avulsion of the apophyses of: ▪ the inferior iliac spine (*a*) ▪ the superior iliac spine (*b*) ▪ the ischial tuberosity (*c*)	
Ilium ala fractures (*d*)	
Pubic arch fractures (pubis and ischium) (*e*)	
Fractures of the ilium (*f*)	
Iliosacral joint loosening (*g*)	
Fractures with severe delayed deformities (unstable fractures)	
Symphysis separation (*h*)	
Sacroiliac joint disruption (*i*)	
Acetabular fractures (*j*)	

Therapy

- Treatment of pelvic ring fractures is given in Table 38.34

Hip dislocation

- Hip dislocation and its management is discussed in Table 38.35

Table 38.34 Treatment of pelvic ring fractures

	Undisplaced/stable	Displaced/stable
Avulsion of apophyses	Crutches/analgesia <10 days	Same
Ilium ala fracture	Crutches until pain free	Open reduction with screw or K-wire fixation
Pubic arch fracture (pubis and ischium)	Crutches until pain free	Same
Fracture of the ilium	Crutches until pain free	Same
Symphysis loosening	Crutches until pain free	Same
Complete unstable symphysis separation	Crutches for 3–4 weeks	External fixator or Recco-plate fixation in older children
Sacro-iliac joint disruption	External fixator	Reduction, external fixator ± transarticular screw or 4-hole plate
Acetabular fractures	Spica cast 5–6 weeks	Open reduction and screw/plate fixation

Table 38.35 Hip dislocation and its management

Morphology	Different types: superior-iliac, posterior-iliac, anterior-pubic, fracture dislocation Very rare in childhood
Signs	Pain, the involved limb is shorter and is held in flexion, adduction, and internal rotation
Diagnosis	Clinical, X-ray, CT
Correction potential	None
Complications	Femoral head necrosis, re-dislocation, secondary hip dysplasia
Nonoperative therapy	Within the first 8 h, aspiration of the joint; if there is any sign of incongruence, open reduction is indicated
Operative treatment	Incongruence after reduction Combined with fracture of the acetabular rim
Immobilization	Depending on the injury and treatment: 1–6 weeks
X-ray control	In weeks 4–6 Scintigraphy or MRI if indicated additionally
Follow-up	When necrosis is suspected: every 6 months

38.15 Lower Limb Fractures

Femoral neck fractures

- Femoral neck fractures and their management are discussed in Table 38.36
- The capital femoral and trochanteric epiphyses have cartilaginous continuity along the posterior superior femoral neck due to embryonal development
- Damage to this cartilaginous continuity, as in a femoral neck fracture, may seriously impair normal development of the neck
- Femoral head necrosis results from vessel damage in this region!

Table 38.36 Femoral neck fractures and their management

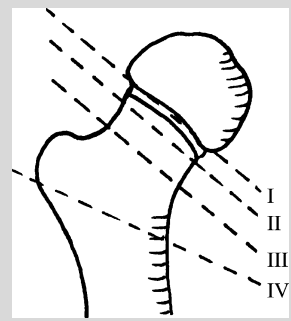
Morphology	Very rare injury in childhood: I Transphyseal injury II Transcervical injury III Cervico-trochanteric injury IV Pertrochanteric injury 
Signs	Pain, the involved limb is shorter and is held in flexion, adduction, and internal rotation
Diagnosis	Clinical, X-ray in two planes, CT
Correction potential	Limited
Complications	Femoral head necrosis, malunion, non-union varus deformity
Nonoperative therapy	Only undisplaced fractures Joint aspiration
Operative treatment	Displaced fractures Open reduction (recommended) Threaded K-wire fixation in small children Screw or angular plate fixation in older children

Table 38.36 (*continued*) Femoral neck fractures and their management

Immobilization	Depending on the injury and treatment: 4–8 weeks
X-ray control	In weeks 4–6 Scintigraphy recommended MRI (using titanium implants if possible) when indicated
Follow-up	If necrosis is suspected every 6 months

Femoral shaft fractures

- Femoral shaft fractures and their management are discussed in Table 38.37

Table 38.37 Femoral shaft fractures and their management

Morphology	Most frequent injury of the lower leg ▪ Subtrochanteric fractures ▪ Fractures (transverse or oblique) of the proximal and middle third
Signs	Pain, deformity, restricted movement, blood loss, shock
Diagnosis	X-ray in two planes, including both hip and knee joint
Correction potential	Very good, depending of the child's age
Complications	Leg length discrepancy, rotation failure, deviation of the axis
Nonoperative therapy	Children <3–4 years → outpatient overhead extension or for stable fractures initial spica cast (not for children with multiple injuries)
Operative treatment	Children 4–13/14 years, depending on their weight → closed reduction and ESIN as the first method of choice Unstable, complex fractures → external fixation or minimal invasive plating (MIPO)
Immobilization	3–4 weeks No immobilization after ESIN, MIPO, Ex-Fix treatment
X-ray control	Overhead extension or spica cast: weeks 3–4 Operative treatment: weeks 5–6, before implant removal
Follow-up	Children >10 years until growth stops

Operative technique with elastic stable intramedullary nailing (ESIN)

- See Fig. 38.9 for operative technique
- Place child in supine position
- For children <8 years: free position, fixed on the standard table
- For children >8 years: fracture table is recommended (especially for transverse fractures)
- Decide on the direction of the nailing: retrograde (normally) or antegrade (special situations)
- Make a preliminary reduction with the aid of an image intensifier on the fracture table
- The nail entry point is normally one finger's breadth about the proximal tip of the patella, which corresponds to 2–3 cm proximal the epiphyseal line
- Make a skin incision about 3–4 cm in the distal direction from the planned entry point in the bone
- Create the nail entry point by penetrating the near cortex with the awl or drill bit
- Insert the awl vertically down to the bone, then lower the awl about 10° to the cortical surface whilst rotating it so that the bone cortex is perforated in an upwards showing angle. With a rotating motion, continue to penetrate the cortical bone at an upward angle
- The nail diameter should be one-third of the narrowest diameter of the medullary canal and as long as the bone when bent. Both nails must be bent in the same way
- The greatest curvature of the nails must be at the level of the fracture
- Drive the first nail to the level of the fracture
- In a similar manner to that previously described, open the femur on the opposite side
- Drive the second nail up to the level of the fracture
- Visualize the fracture with fluoroscopy and decide which nail will be easier to pass across the fracture and will most effectively pull the proximal fragment into alignment
- Drive this nail across the fracture, monitoring its position with fluoroscopy

- Advance this nail into the proximal fragment only so far as to ensure that the reduction is maintained
- Position the second nail in the same manner
- Both nails can now be advanced to the proximal epiphysis
- Cut the nails to the right length outside of the skin
- Control the rotation of the leg
- The final position of the nails is achieved when the end points are placed in the proximal fragment
- The distal ends of the nails should poke out at least 8 mm from the cortex in order to facilitate easy removal, whilst the low profile minimizes soft-tissue irritation
- If the fracture is distracted, release traction and impact the patient's heel
- Close the skin
- Do not remove the nail before complete consolidation, wait at least 4–5 months

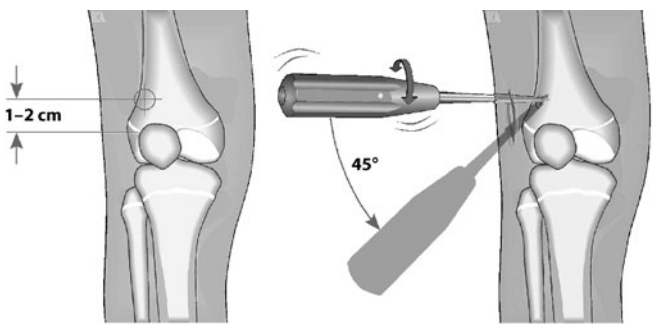


Fig. 38.9 Operative technique with Elastic-Stable-Intramedullary-Nailing (ESIN)

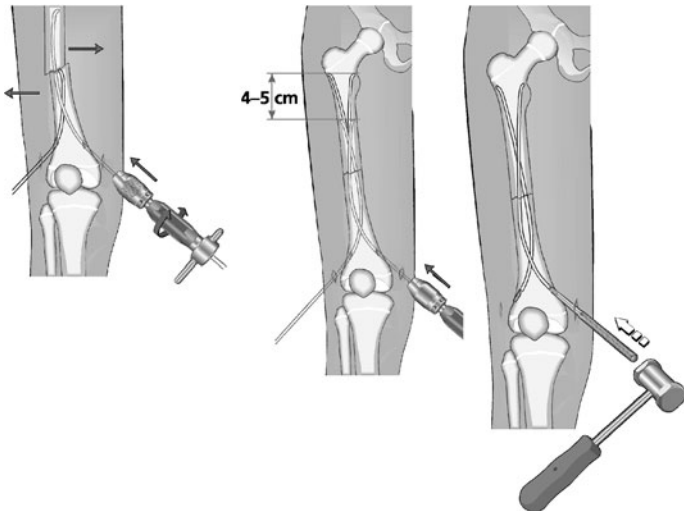


Fig. 38.9 *(continued)* Operative technique with Elastic-Stable-Intramedullary-Nailing (ESIN)

38.16 Distal Femur and Proximal Tibia Fractures

General considerations

- Very rare fractures in childhood, following high-energy trauma or traffic accidents
- The radiological diagnosis is not easy, however it is not as difficult as for the elbow
- Hemarthros indicates a severe trauma
- Osteochondral fragments, "flake fractures," must be looked for

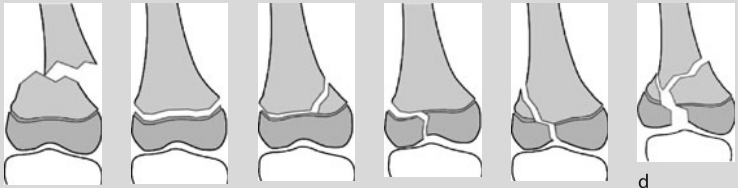
Classification

- Distal femur and proximal tibia fractures are described in Table 38.38
- Supracondylar and condylar fractures of the femur and their management are described in Table 38.39
- Fractures of the proximal tibial epiphysis and their management are detailed in Table 38.40

Table 38.38 Distal and proximal tibia fractures

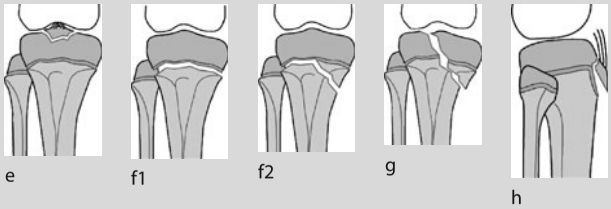
Supracondylar and condylar region

- Supracondylar fractures (*a*)
- Salter–Harris I + II fractures (*b1,2*)
- Unicondylar Salter–Harris III + IV fractures (*c1,2*)
- Bicondylar Salter–Harris III + IV fractures (*d*)



Fractures of the proximal tibia "Tibiaplateau"

- Tibia spine injuries (*e*)
- Salter–Harris I + II fractures (*f1,2*)
- Salter–Harris III + IV fractures (*g*)
- Tibial tuberosity fracture (*h*)



Fractures of the proximal tibia metaphysis

- Simple metaphyseal fracture (*i*)
- Complex metaphyseal fracture (*j*)

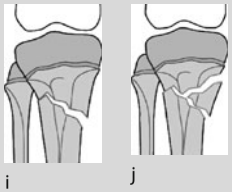


Table 38.39 Supracondylar and condylar fractures of the femur

Morphology	▪ Supracondylar buckle fractures ▪ Complete transverse or oblique fractures, metaphyseal or physeal (Salter–Harris I + II) ▪ Uni- or bicondylar (Salter–Harris III + IV) fractures (very rare in childhood)
Signs	Pain, knee stiffness, swelling, pulseless lower leg
Diagnosis	X-ray in two planes
Correction potential	Good, depending on the child's age
Complications	None for supracondylar fractures Varus or valgus angulation following premature partial closure of the growth plate, limitation of knee motion, leg length discrepancy
Nonoperative therapy	All undisplaced fractures independent of the age Long leg cast immobilization in 10° flexed
Operative treatment	All displaced fractures, which need reduction ▪ Closed (extra-articular fractures) reduction and K-wires or Ex-Fix stabilization ▪ Open (articular fractures) reduction and K-wires or screw fixation ▪ Any fracture that needs a reduction under anesthesia should be treated with a stable, definitive fixation
Immobilization	4–6 weeks
X-ray control	Nonoperative: days 4, 10 and week 4 Operative: postoperative and weeks 4–6
Follow-up	Up to 2 years, depending on the fracture type

Table 38.40 Fractures of the proximal tibial epiphysis and their management

Morphology	Fracture types: ▪ Complete ▪ Type "greenstick" → "Judge fracture" ▪ Type bowing and torus or buckle
Signs	Pain, swelling
Diagnosis	X-ray in two planes, → medial open wedge fracture
Correction potential	No correction of valgus deformity
Complications	Consolidation problems with progressive valgus angulation

Table 38.40 *(continued)* Fractures of the proximal tibial epiphysis and their management

Nonoperative therapy	Stable, undisplaced fractures with valgus angulation <10° → long leg cast, cast wedging after 8 days to compress the medial cortex
Operative treatment	Displaced fractures and fractures with valgus angulation >10° → closed or open reduction K-wires or medial Ex-Fix stabilization (compression on the medial cortex)
Immobilization	~ 5 weeks
X-ray control	Nonoperative therapy: days 4, 8 and week 5 Operative treatment: postoperative and week 5
Follow-up	Every 6 months for 2 years

38.17 Patella Fractures and Dislocations

General considerations

- Patellar dislocations (Table 38.41) are common when considering the entire spectrum of acute and chronic subluxation and dislocation injuries
- More frequently, chronic subluxation mimics actual dislocation
- Dislocation of the patella is frequent in young girls
- Prerequisites are "genua valga," being overweight, and "patella alta"
- Fractures of the patella (Table 38.42) result from direct trauma and high-energy extension trauma (such as high jumping)

Table 38.41 Patellar dislocation and its management

Morphology	Nearly always dislocations in the lateral direction The dislocation may be complete or incomplete
Signs	Pain, swelling, blocking of the knee, hemorrhage in the joint
Diagnosis	Clinic signs, X-ray in two planes
Correction potential	None
Complications	Overlooked "flake fracture" Repeated dislocations
Nonoperative therapy	Reduction and extension of the knee while the hip is flexed ± aspiration of the hemarthros if the knee is painful
Operative treatment	If there are any signs of osteochondral fractures arthroscopy is indicated Re-fixation or removal of the fragment
Immobilization	The limb should be immobilized in a cylindrical cast
X-ray control	After reduction
Follow-up	Physiotherapy if habitual dislocation is suspected

Table 38.42 Patellar fractures and their management

Morphology	▪ Incomplete and complete fractures ▪ Inferior and superior fractures ▪ Longitudinal and transverse fractures ▪ "Sleeve" fracture ▪ Variations of the norm → bipartite patella
Signs	Pain, swelling, blocking of the knee, hemorrhage in the joint
Diagnosis	Clinic, X-ray in two planes
Correction potential	Partial, a cartilaginous gap is always filled out with fibrous cartilage
Complications	Nonunion, pre-arthrosis
Nonoperative therapy	Fissures, undisplaced, stable fractures → cylindrical cast
Operative treatment	All displaced fractures → circumferential wiring of the patella Implant removal after 4–5 months
Immobilization	4–5 weeks
X-ray control	Nonoperative therapy: day 6 and week 5 Operative treatment: postoperative and week 5
Follow-up	6 months after implant removal

38.18 Tibial Shaft Fractures

General considerations

- Fracture of the tibial shaft (Table 38.43) is one of the most frequent fractures of the lower leg in childhood
- Injuries of this region vary considerably with both age and mechanism of injury
- In infants and children, the typical injury is a spiral tibial fracture with an intact fibula
- This circumstance influences the kind of treatment and the outcome
- Complete fractures of the tibia and fibula (Table 38.44) are unstable, and often shortened with rotational failures
- Special types: toddler fractures or bowing of the fibula or (rarely) of the tibia
- Fractures of the tibial shaft (isolated tibial fractures)

Table 38.43 Tibial shaft fractures

Morphology	Fractures of the middle and distal third Spiral fractures are more frequent than transverse fractures
Signs	Pain, swelling, angulation
Diagnosis	X-ray in two planes including the knee and ankle joint
Correction potential	Good, depending on the age No correction for rotation deformities
Complications	Different rotation of the feet, remaining angulation
Nonoperative therapy	Undisplaced and stable fractures with angulation <10° (open long leg cast and if necessary cast wedging on days 4–5)
Operative treatment	The indication for operative therapy is rare Shortening of the tibia with bowing of the fibula (Ex-Fix or MIPO) ESIN can produce nonunion because of the blocking fibula
Immobilization	4–5 weeks No immobilization is required after operative treatment
X-ray control	Nonoperative therapy: days 4 and 10 and weeks 4–5 Operative treatment: postoperative, week 5
Follow-up	Every 6 months, up to 2 years after the procedure

Table 38.44 Complete fracture of the tibia and fibula

Morphology	Fractures of the middle and distal third Spiral fractures are more frequent than transverse fractures Very often fully displaced as the stabilizing effect of the fibula is missing
Signs	Pain, swelling, angulation
Diagnosis	X-ray in two planes including the knee and ankle joint
Correction potential	Good, depending on the child's age No correction for rotation deformity
Complications	Different rotation of the feet, remaining angulation, leg length discrepancy
Nonoperative therapy	Undisplaced and stable fractures with angulation <10° (open long leg cast and if necessary cast wedging on days 4–5)
Operative treatment	The indication for surgery is rare Displaced/unstable fractures (oblique and spiral) Method of choice → ESIN or Ex-Fix (MIPO in older children)
Immobilization	4–5 weeks No immobilization required after operative treatment
X-ray control	Nonoperative therapy: days 4 and 10 and weeks 4–5 Operative treatment: postoperative, week 5
Follow-up	Every 6 months, for up to 2 years after the procedure

38.19 Distal Tibia and Ankle Joint Fractures

General considerations

- Like the proximal region, the distal metaphysis may sustain injury in patterns of varying severity
- Due to the microstructural differences between the thick diaphysis and the thinner metaphysis, greenstick and torus fractures are common
- Fractures of the ankle joint are classified (Table 38.45) according to the maturation of the epiphysis and the child's age
- The epiphysis begins to close from the age of 12 in girls, 14 in boys
- Fracture types vary depending on epiphyseal maturation
- Extra-articular (Table 38.46) and intra-articular (Table 38.47) fractures and their management are discussed below

Table 38.45 Classification of distal tibia and ankle joint fractures

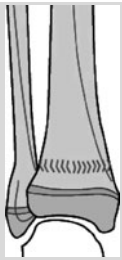
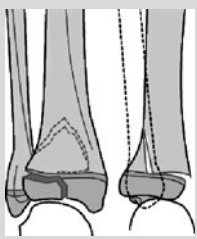
Extra-articular		
Metaphyseal torus or green-stick fracture (*a*)	 a	 b
Complete metaphyseal fracture (*b*)		
Epiphysiolysis (Salter–Harris I) (*c*)		
Epiphysiolysis with metaphyseal wedge (*d*)	 c	 d
Articular fractures		
Epiphyseal fracture (Salter–Harris III) (*e*) or Tillaux (two-plane) fracture	 g	 e
Epiphyseal fracture with metaphyseal wedge (Salter–Harris IV) (*f*)		
Tri-plane fracture (Salter–Harris IV) (*g*)		 f

Table 38.46 Extra-articular fractures and their management

Morphology	Metaphyseal torus fracture Metaphyseal bowing fracture Complete metaphyseal fracture Epiphysiolysis (Salter–Harris I) Epiphysiolysis with metaphyseal wedge
Signs	Pain, deformation, restricted movement
Diagnosis	X-rays in two planes Look for accessory ossification Documentation under image intensifier may be necessary
Correction potential	Very good
Complications	Alteration of leg length, valgus deformity, premature closure of the growth plate, fibulo-tibial synosthosis
Nonoperative therapy	Metaphyseal torus and bowing fracture (plaster cast immobilization and cast wedging if necessary) Metaphyseal complete fracture (stable reduction, plaster cast) Stable, Salter–Harris I + II fractures (reduction, plaster cast)
Operative treatment	Complete displaced, unstable metaphyseal fractures (closed or open reduction and Ex-Fix or K-wires or ESIN fixation when distal fragment >4 cm) Displaced, unstable Salter–Harris I + II fractures (closed or open reduction and minimal invasive screw fixation under image intensifier with cannulated self drilling, self-tapping screws or K-wire fixation)
Immobilization	4–6 weeks
X-ray control	Nonoperative therapy: day 6, weeks 4–5 Operative treatment: postoperative, week 5
Follow-up	Every 6 months, for up to 2 years after the procedure

Table 38.47 Articular fractures (Salter–Harris III and IV) and their management

Morphology	Epiphyseal fractures (Salter–Harris III) Epiphyseal fractures with metaphyseal wedge (Salter–Harris IV) "two-plane" or "tri-plane" fractures
Signs	Pain, deformation, restricted movement
Diagnosis	X-ray in two planes Look for accessory ossification Documentation under image intensifier may be necessary

Table 38.47 *(continued)* Articular fractures (Salter–Harris III and IV) and their management

Correction potential	Moderate
Complications	Alteration of leg length, valgus deformity, premature closure of the epiphysis, non-union
Nonoperative therapy	Cast or splint for undisplaced fractures with an articular gap <2 mm
Operative treatment	Displaced unstable Salter–Harris III + IV fractures (closed or open reduction and minimally invasive screw fixation under image intensifier with cannulated self drilling, self tapping screws or K-wire fixation)
Immobilization	4–6 weeks
X-ray control	Nonoperative therapy: day 6, weeks 4–5 Operative treatment: postoperative, week 5
Follow-up	Every 6 months for up to 2 years after the procedure

Bi- and tri-plane fractures: "fractures of Tillaux"

- This particular type of injury affects a part of the anterolateral tibial epiphysis
- The segment may extrude anteriorly and laterally
- Ankle congruity is of concern because juvenile two-plane fractures involve the weight-bearing articular surface
- The fracture may be accompanied by a posterior metaphyseal fragment and in this case the fracture is called a "tri-plane fracture"
- The exact diagnosis can often be difficult when only based on standard a.p. X-ray projection of the distal tibia and fibula
- Exact examination under the image intensifier can clarify the diagnosis
- However, CT may exhibit far greater accuracy than plain radiographs in delineating the degree of joint displacement and fragment separation
- This fracture type occurs especially in adolescents
- A schematic and model illustration of a bi-plane fracture at the ankle joint is shown in Fig. 38.10
- A schematic and model illustration of a tri-plane fracture at the ankle joint is shown in Fig. 38.11

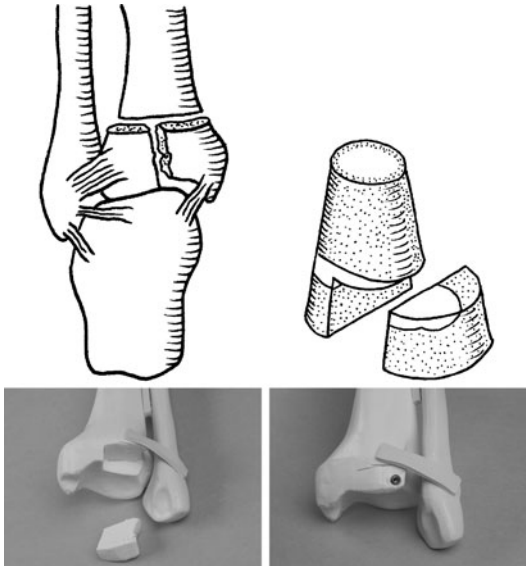


Fig. 38.10 Schematic and model illustration of a bi-plane fracture at the ankle joint

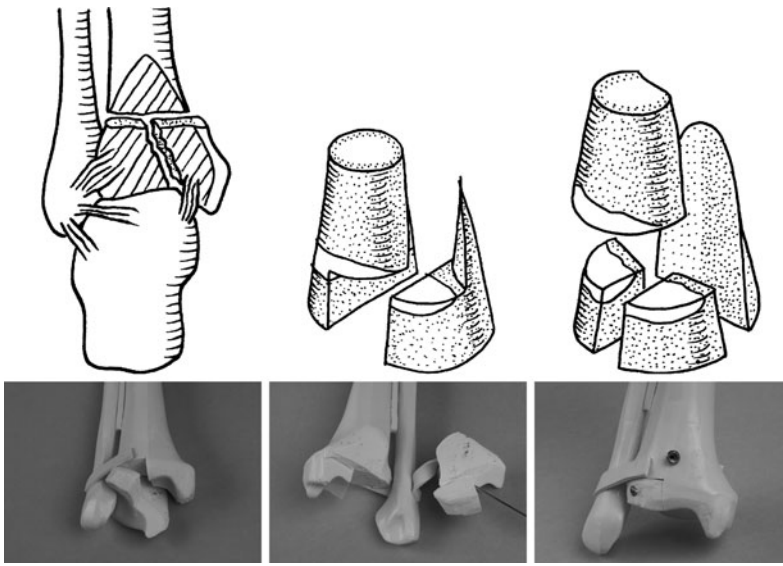


Fig. 38.11 Schematic and model illustration of a tri-plane fracture at the ankle joint

Therapy

- The aim of therapy is exact reconstruction of the joint surface
- Undisplaced (<2-mm gap) bi-plane and tri-plane fractures are treated nonoperatively by a well-padded compression dressing and posterior splint. After the swelling has disappeared the fixation is changed to a Sarmiento type cast
- Displaced bi- and tri-plane fractures are treated surgically
- Reduction of the fracture and retention are achieved using a minimally invasive technique such as that using cannulated, self-drilling, self-tapping screws
- The extremity is placed directly on the image intensifier
- The leg must be turned so that the fracture can be seen exactly in an a.p. view
- The screws must be placed exactly perpendicular to this plane. The adaptation of the fracture can easily be observed with the intensifier

38.20 Foot Fractures

General considerations

- Fractures of the talus and the calcaneus are rare in childhood
- Fractures of the metatarsals are frequent, the first and fifth rays in particular are involved
- The first metatarsal may be injured proximally, either in the metaphysis or the proximal growth plate
- Solitary fractures of the metatarsal diaphysis are usually undisplaced
- Symptomatic accessory bones make diagnosis difficult

Therapy

- The treatment of foot fractures is detailed in Table 38.48 and complications arising are listed in Table 38.49

Table 38.48 The treatment of foot fractures

Fracture type	Therapy	Immobilization
Calcaneus	Undisplaced → non-weight-bearing cast	6–8 weeks
	Displaced → open reduction and screw or plate stabilization, non-weight-bearing cast	
Talus	Undisplaced → non-weight-bearing cast	6–8 weeks
	Displaced → open reduction and screw or K-wire fixation, non-weight-bearing cast	
Metatarsal	Undisplaced → non-weight-bearing cast	3–4 weeks
	Displaced → open reduction and screw or K-wire fixation, non-weight-bearing cast	

Table 38.49 Complications of foot fractures

Fracture type	Complication
Calcaneus	Arthrosis, stiffness in the subtalar joint, pain
Talus	Necrosis, arthrosis of the ankle joint, stiffness in the ankle joints
Metatarsals	Non-union, pain

PART IV
APPENDICES

39 Embryology

39.1 Embryological Organ Development

- Embryological organ development is detailed in Fig. 39.1

39.2 Fetal Circulation

- The embryological origins of the fetal circulation are described in Table 39.1 and Fig. 39.2

39.3 Newborn Icterus

- Newborn icterus is illustrated in Fig. 39.3

Table 39.1 The embryological origins of the fetal circulation

Fetal structures	From/to	Remnant
Umbilical veins	Umbilicus/ductus venosus	Hepatic teres ligament
Ductus venosus	Umbilical veins/inferior vena cava	Ligamentum venosum
Foramen ovale	Right atrium/left atrium	Closed atrial septum
Arterial duct	Pulmonary arteries/descending aorta	Ligamentum arteriosum
Umbilical arteries	Common ileal artery/umbilicus	Superior vesicle artery Lateral vesicoumbilical ligament

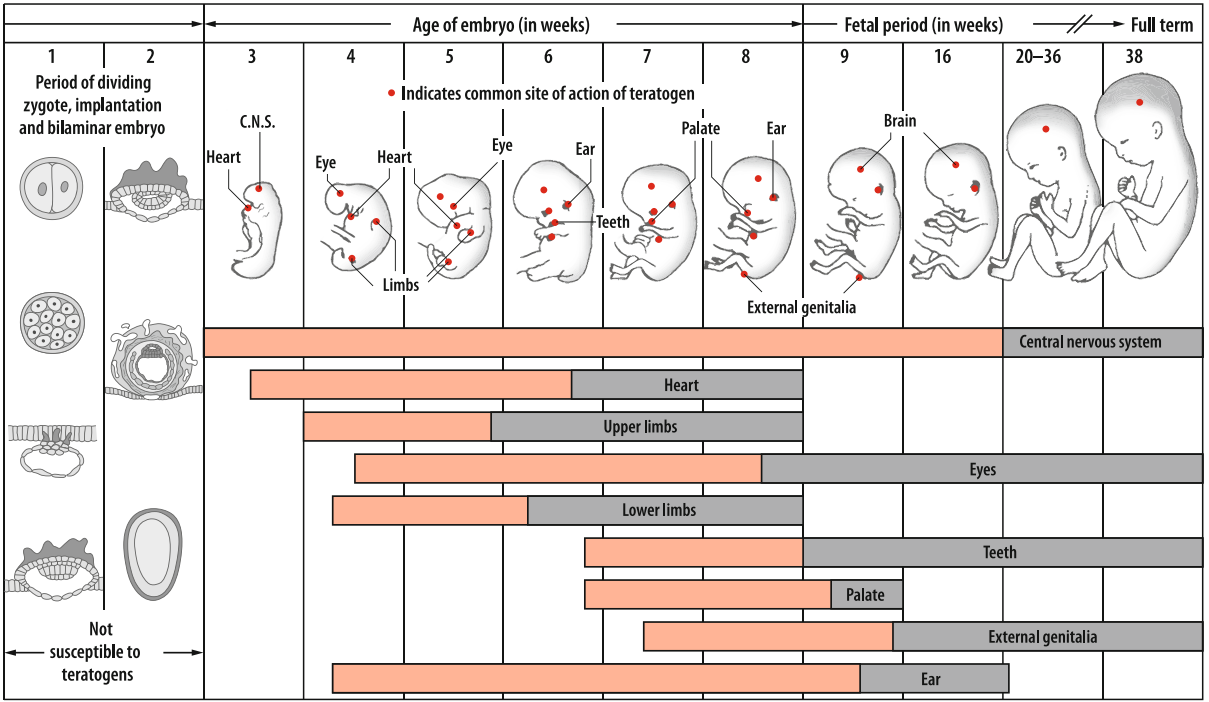


Fig. 39.1 Embryological organ development

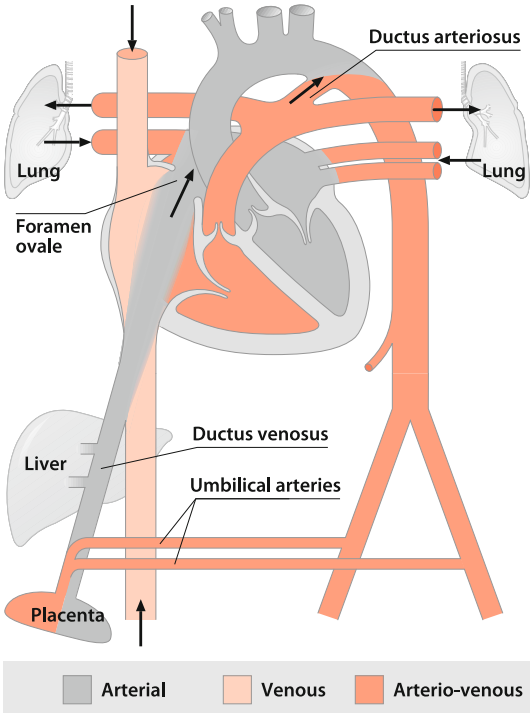


Fig. 39.2 Fetal circulation

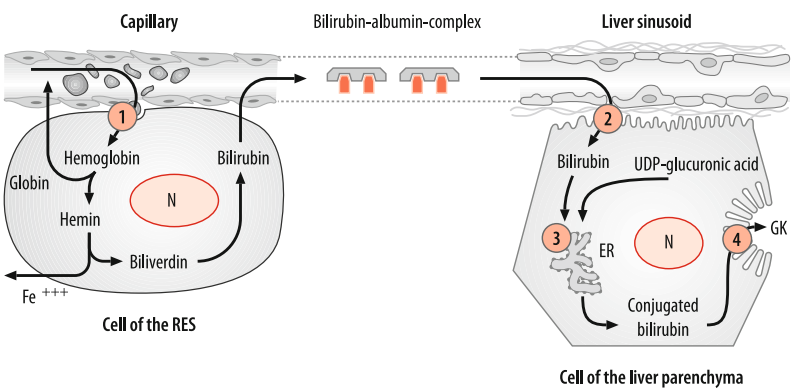


Fig. 39.3 Newborn icterus

39.4 Segmental Innervation

- Sensory dermatomes are shown in Fig. 39.4

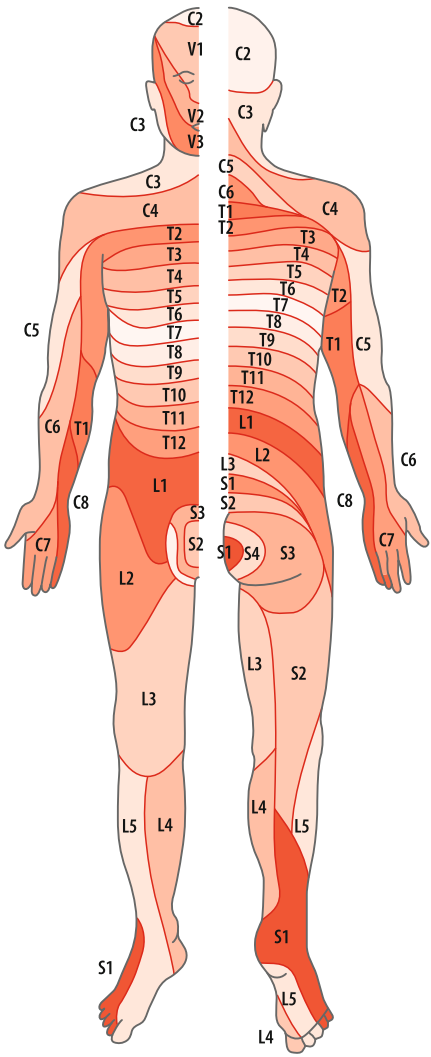


Fig. 39.4 Segmental innervation

40 Vaccination

40.1 Vaccines

- Vaccines are given to prevent, ameliorate or treat infectious diseases. They consist of attenuated or killed microorganisms, or of antigenic proteins derived from them (Table 40.1)

40.2 Rabies Vaccination

- The indications for rabies vaccination and recommended schedules are given in Table 40.2

Table 40.1 Vaccines, their characteristics and route of application

Vaccine (abbreviation)		Disease	Characteristics	Application	Combined vaccines
Inactivated vaccines	Bacteria	Tetanus (T)	Toxoid	i.m.	DT, Td, DPT
		Diphtheria (D,d)			
		Pertussis (P)	Inactive bacteria		
		Cholera			
		Haemophilus influenzae (Hib)	Surface polysaccharide		
		Meningococci A, C			
		Pneumococci			
	Viruses	Hepatitis B (HB)	HbsAg	i.m.	
		Early summer meningoencephalitis	With formaldehyde-inactivated virus		
		Poliomyelitis			
		Influenza	With β-propiolactone-inactivated virus		
		Rabies			
Live vaccines	Bacteria	Bacillus Calmette-Guérin (BCG)	Attenuated bovine mycobacterium	i.c.	
		Typhus	Enzyme-deficient *Salmonella typhi* mutant	p.o.	
	Viruses	Measles	Attenuated virus	s.c.	MMR
		Mumps			
		Rubella			
		Poliomyelitis	Avirulent poliovirus	p.o.	
		Yellow fever	Attenuated virus	s.c.	
		Varicella	Attenuated virus	s.c.	

Table 40.2 Rabies vaccination, its indications and schedule

Vaccine: HDC (human diploid cell culture)		
Vaccination plan	**Indication**	**Application**
Prophylactically	▪ Regular visits to areas where there is rabies ▪ Exposure to rabid animal/cadaver where there has definitely been no contact with saliva and no skin lesions before or after contact ▪ If vaccination has already been initiated, however the animal was not infected	One immunization on days 0, 28, 56 and a year later Or One immunization on days 0, 7, 21 and a year later
Post-exposure	▪ Exposure to animal with suspected rabies, contact with saliva and/or with skin lesion ▪ Exposure to rabid animal/cadaver and modes of contact are unknown, no skin lesion ▪ Re-exposure and the last vaccination was >5 years previously	One immunization on days 0, 3, 7, 14, 30, 90
Post-exposure simultaneous vaccination	▪ Animal with suspected rabies is not available for examination, direct contact with the animal skin or saliva (animal bite) occurred	20 IU·kg^{-1} human rabies immunoglobulin + schedule for post-exposure vaccination

For all vaccination schedules: vaccination needs to be repeated in 2–5 years		
Re-exposure	**Light cases**	**Severe cases**
Full vaccination within <1 year	One dose on day 0	Vaccination on days 0, 3
Full vaccination within previous 1–5 years	One dose on days 0, 3	Vaccination on days 0, 3, 7

41 Notifiable Infectious Diseases

- The occurrence of a notifiable disease (Table 41.1) must, by law, be reported to a health or local government authority

Table 41.1 Notifiable diseases, their etiology and their notification procedure rules; applicable to most European Union countries

Disease	Pathogen agent	Notification in				Special rules
		Suspicion	Disease	Secretor	Death	
Meningitis	All germs		×		×	
Encephalitis	All germs		×		×	
Cytomegaly	Cytomegalovirus (CMV)		×[a]		×[a]	[a]Only inborn
Early summer meningo-encephalitis	Flavi virus		×		×	
Yellow fever	Flavi virus		×		×	
Japanese B-encephalitis	Flavi virus		×		×	
Hemorrhagic fever	Hanta virus, Puumala virus	×	×		×	
Hepatitis A	Picornavirus		×		×	
Hepatitis B	Hepadnavirus		×		×	
Hepatitis C	Toga virus (?)		×		×	
Hepatitis D	RNA virus		×		×	
Hepatitis E	?		×		×	
Herpes simplex	Human herpes virus 1		×[a]			[a]Only in meningoencephalitis
Herpes genitalis	Human herpes virus 2		×[a]			
Influenza					×	
Measles	Paramyxovirus		×[a]		×	[a]Only in hospitals
Mumps	Paramyxovirus		×[a]		×[a]	[a]Only meningitis in schools, kindergarten, hospitals
Smallpox	Orthopoxvirus	×	×		×	

Table 41.1 *(continued)* Notifiable diseases, their etiology and their notification procedure rules; applicable to most European Union countries

Disease	Pathogen agent	Notification in				Special rules
		Suspicion	Disease	Secretor	Death	
Poliomyelitis	Enterovirus	×	×		×	
Roseola	Rubivirus		×[a]		×[a]	[a]Only embryopathy
Rabies	Rhabdovirus	×	×		×	
Varicella-zoster	Human Herpes virus 3		×[a]			[a]Only in encephalitis
Borreliosis	*Borrelia recurrentis*	×	×		×	
Botulism	*Clostridium botulinum*	×	×		×	
Brucellosis (Mediterranean fever)	*Brucella melitensis*		×		×	
Cholera	*Vibrio cholerae*	×	×		×	Quarantine
Diphtheria	*Corynebacterium diphtheriae*		×		×	
Spotted fever	*Rickettsia prowazekii*	×	×		×	
Gaseous gangrene	*Clostridium perfringens*		×		×	
Leprosy	*Mycobacterium leprae*	×	×		×	
Listeriosis	*Listeria monocytogenes*		×[a]		×[a]	[a]Only inborn
Syphilis (lues)	*Treponema pallidum*		×[a]		×[a]	[a]Connatal, others anonymous
Anthrax	*Bacillus anthracis*	×	×		×	
Ornithosis	*Chlamydia psittaci*	×	×		×	

Table 41.1 *(continued)* Notifiable diseases, their etiology and their notification procedure rules; applicable to most European Union countries

Disease	Pathogen agent	Notification in				Special rules
		Suspicion	Disease	Secretor	Death	
Paratyphus A,B,C + other Salmonelloses	*Salmonella paratyphi* and others	×	×	×	×	
Pertussis	*Bordetella pertussis*		×[a]		×	[a]Epidemic
Plague	*Yersinia pestis*	×	×			Quarantine
Q-Fever	*Coxiella burnetii*		×		×	
Malleus	*Pseudomonas mallei*		×		×	
Scarlatina	*Streptococci group* A		×[a]		×	[a]Only public institutions
Shigellosis	*Shigella*	×	×	×	×	
Tetanus	*Clostridium tetani*		×		×	
Trachoma	*Chlamydia trachomatis*		×		×	
Tuberculosis	*Mycobacterium bovis, M. avium, M. tuberculosis*		×		×	
Rabbit fever	*Francisella tularensis*	×	×		×	
Typhus	*Salmonella typhi*	×	×	×	×	
Malaria	*Plasmodia*		×[a]		×	[a]In recurrence
Toxoplasmosis	*Toxoplasma gondii*		×[a]		×[a]	[a]Only inborn
Trichinosis	*Trichinella spiralis*		×		×	

42 Fever Types

- Fever take many forms, which are described in Table 42.1 and Fig. 42.1

Table 42.1 Examples of fever courses

Fever type	
Continuum (amplitude <1°C)	Typhus, virus pneumonia
Remittent	Non-diagnostic
Intermittent	Sepsis, malignancy
Periodical	Virus infection, poliomyelitis (biphasic), malaria
Undulant	Hodgkin's disease, Bang's disease

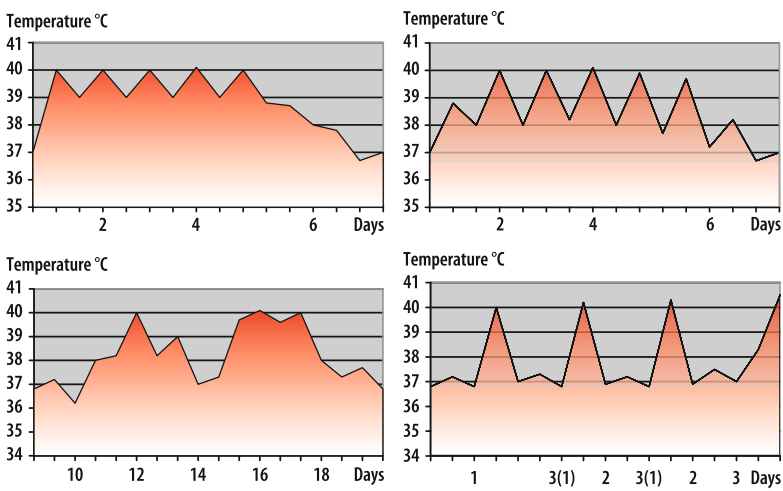


Fig. 42.1 Fever types

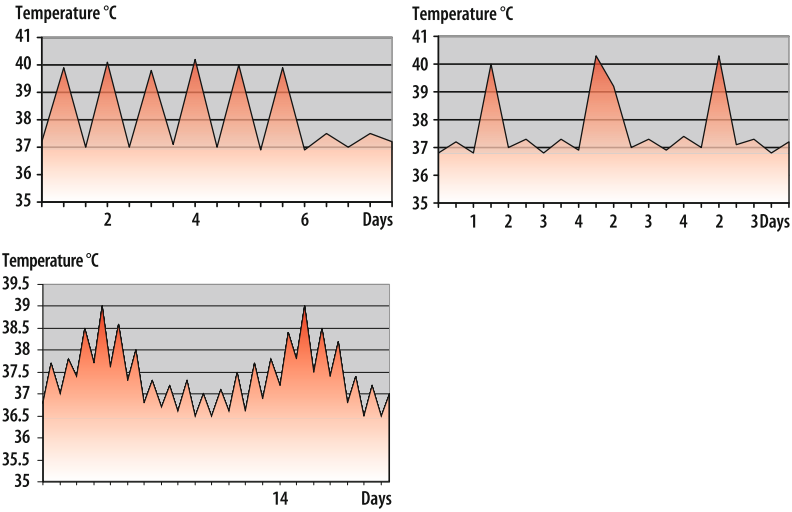


Fig. 42.1 *(continued)* Fever types

43 Catheter Size

- Catheter sizes are measured using the French scale (also known as the Charrière scale) (Fig. 43.1), where the conversion to metric measurement is: 1 French = 1 Charrière = 0.33 mm

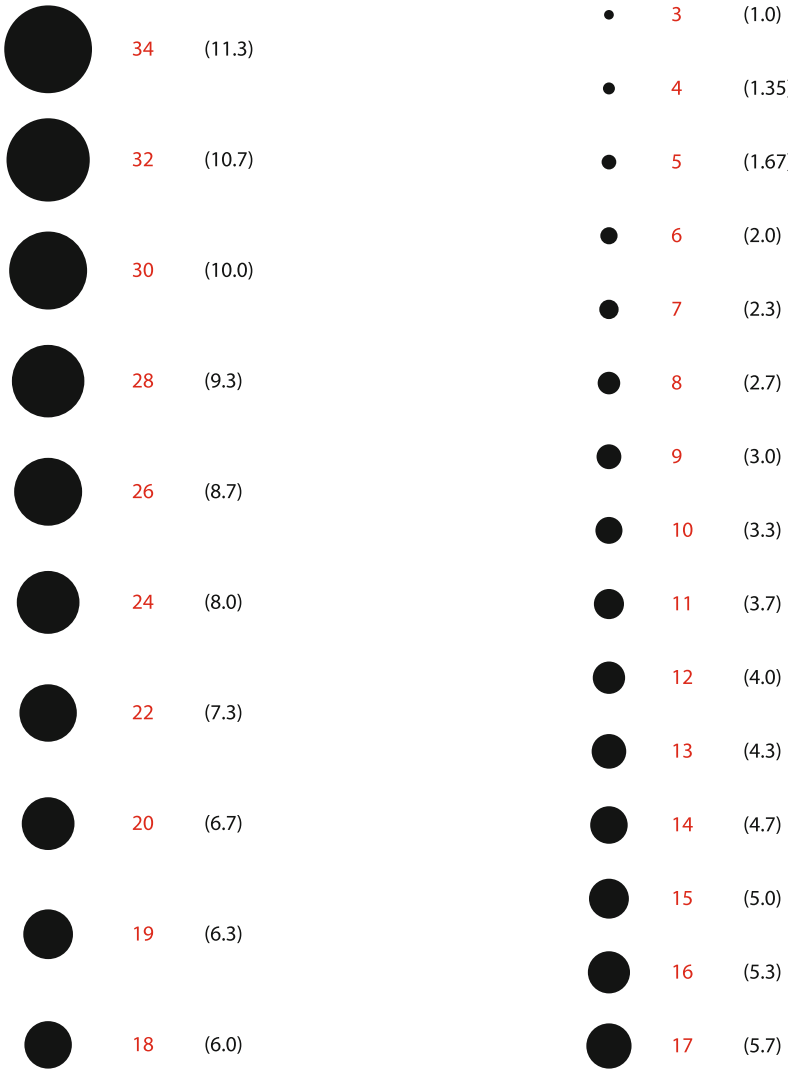
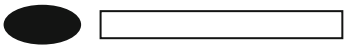


Fig. 43.1 Catheter sizes

In case of oval shaped catheters please put a stripe of paper around the catheter marking the circumference. Then put your stripe besides a ruler and see how many mm you measured. You get an approximately value in French by comparing the value to the above table.



3.2 mm = between 9 and 10 French

Table 44.1 Plasma albumin: its measurement and normal values from birth to 6 years of age. Albumin measured using electrophoresis (S) using the Ponceau-S stain. Values are the mean and 2 SD in *parentheses*. The value obtained with amido-black stain is given as a percentage (%)

Age group	Albumin ($g \cdot l^{-1}$, %)	α_1-Globulin ($g \cdot l^{-1}$, %)	α_2-Globulin ($g \cdot l^{-1}$, %)	β-Globulin ($g \cdot l^{-1}$, %)	γ-Globulin ($g \cdot l^{-1}$, %)
Newborn	37.1 (32.7–45.3) $g \cdot l^{-1}$	1.7 (1.1–2.5) $g \cdot l^{-1}$	4.2 (2.6–5.7) $g \cdot l^{-1}$	4.2 (2.5–5.6) $g \cdot l^{-1}$	7.4 (3.9–11) $g \cdot l^{-1}$
	68.1%	3.1%	7.7%	7.6%	13.5%
Infants	49.2 (35.7–51.3) $g \cdot l^{-1}$	1.7 (1.3–2.5) $g \cdot l^{-1}$	6.2 (3.8–10.8) $g \cdot l^{-1}$	5.3 (3.5–7.1) $g \cdot l^{-1}$	5.6 (2.9–11) $g \cdot l^{-1}$
	69.6%	2.7%	10.0%	8.7%	9.0%
1–2.5 years	45.1 (36.5–53.3) $g \cdot l^{-1}$	1.9 (l.1–2.9) $g \cdot l^{-1}$	6.9 (4.9–9.4) $g \cdot l^{-1}$	5.9 (4.3–7.9) $g \cdot l^{-1}$	7.5 (4.7–11.8) $g \cdot l^{-1}$
	67.0%	2.8%	10.3%	8.7%	11.2%
2.5–6 years	45.6 (33.1–52.2) $g \cdot l^{-1}$	1.9 (0.9–2.9) $g \cdot l^{-1}$	6.6 (4.3–9.5) $g \cdot l^{-1}$	5.9 (3.5–7.6) $g \cdot l^{-1}$	7.7 (4.5–12.1) $g \cdot l^{-1}$
	67.4%	2.7%	9.7%	8.8%	11.4%
>6 years	46.0 (40.0–52.5) $g \cdot l^{-1}$	1.8 (1.2–2.5) $g \cdot l^{-1}$	6.4 (4.3–8.6) $g \cdot l^{-1}$	5.7 (4.1–7.9) $g \cdot l^{-1}$	8.4 (5.9–13.7) $g \cdot l^{-1}$
	67.4%	2.6%	8.4%	8.3%	12.3%

Table 44.2 Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Acetylsalicylic acid (S) Salicylic acid (S)		1.1–2.2 mmol·l^{-1}	15–30 mg·dl^{-1}
Epinephrine (adrenaline) (24-h urine) {HPCL}	3–6 years	13.1 (4.9–27.3) nmol·day^{-1} 6.6 (2.9–13.1) µmol·mol^{-1}	2.4 (0.9–5.0) µg·day^{-1} 9.2 (4.1–18.3) µg·g^{-1}
	6–10 years	22.4 (10.9–53.5) nmol·l^{-1} 6.4 (2.0–11.9) µmol·mol^{-1}	4.1 (2.0–9.8) µg·day^{-1} 9.0 (4.1–16.6) µg·g^{-1}
	10–16 years	26.2 (8.7–51.3) nmol·l^{-1} 4.7 (1.3–8.7) µmol·mol^{-1}	4.8 (1.6–9.4) µg·day^{-1} 6.5 (1.8–12.2) µg·g^{-1}
Lipids in stool (Stool)			< 4.5 g·day^{-1}
Aldolase (S)	4–8 days		6.4 (0.6–12.2) U·l^{-1}
	2–3 months		3.8–7.7 U·l^{-1}
	4–12 months		3.1 (0.6–5.6) U·l^{-1}
	2–6 years		2.4 (0.6–4.2) U·l^{-1}
	7–15 years		2.4 (1.0–3.8) U·l^{-1}
Albumin in liquor (L) {Electroph.}	Children	Prealbumin Albumin α_1-Globulin α_2-Globulin ß-Globulin τ-Globulin ß + τ-Globulin γ-Globulin	8 (3–13)% 59 (40–70)% 4 (2–9.5)% 6.5 (3.5–12)% 10 (7–14.5)% 4 (2–7)% 14 (9–21.5)% 7.5 (3–13)%
Albumin (total) (S) {Biuret reaction} (24-h-urine)	Newborn		5.69 (4.52–6.86) mg·dl^{-1}
	Infants		5.95 (4.57–7.33) mg·dl^{-1}
	Children		6.93 (5.85–8.01) mg·dl^{-1}
	Any age		≤ 150 mg·l^{-1}·0.73 m^{-2}
	Any age		≤ 40 mg·dl^{-1}
Ammonia (EDTA/heparin)	1 day	97 (30–144) µmol·l^{-1}	165 (51–245) µg·dl^{-1}
	5–6 days	67 (53–144) µmol·l^{-1}	114 (53–144) µg·dl^{-1}
	> 5 years	36 (24–48) µmol·l^{-1}	61 (41–82) µg·dl^{-1}

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Amylase (S)	Infants		→ 40 U·l^{-1}
	Children		→ 50 U·l^{-1}
Antithrombin III (Citrat-P) {Immundiff.}			♀ 28.6 (21–57) mg·dl^{-1}
			♂ 43.2 (26–53) g·dl^{-1}
α-1-Antitrypsin (S) {Nephelometry}	0–7 days		200–400 mg·dl^{-1}
	8 days–12 months		130–240 mg·dl^{-1}
	> 1 year		130–300 mg·dl^{-1}
Apolipoprotein A I	1–7 days		76 (22–130) mg·dl^{-1}
	Children		118 (42–194) mg·dl^{-1}
Apolipoprotein A II	Children		44 (18–70) mg·dl^{-1}
	1–7 days		53 (9–90) mg·dl^{-1}
Apolipoprotein B	Children		86 (74–98) mg·dl^{-1}
Bilirubin (S)	Infants	4.89 (1.73–13.8) µmol·l^{-1}	0.3 (0.1–0.8) mg·dl^{-1}
	Children	6.05 (1.7–21.5) µmol·l^{-1}	0.4 (0.1–1.3) mg·dl^{-1}
Bile acids (total) (S)	1–7 days	9.5 (1.96–17.0) µmol·l^{-1}	3.9 (0.8–6.9) mg·l^{-1}
	7–30 days	11.8 (5.54–18.1) µmol·l^{-1}	4.8 (2.3–7.4) mg·l^{-1}
	1–9 months	6.14 (1.2–11.1) µmol·l^{-1}	2.5 (0.5–4.5) mg·l^{-1}
	2 years	2.93 (0–6.09) µmol·l^{-1}	1.2 (0–2.5) mg·l^{-1}
	4 years	3.80 (0–8.14) µmol·l^{-1}	1.6 (0–3.3) mg·l^{-1}
	> 4 years	4.25 (1.49–7.01) µmol·l^{-1}	1.7 (0.6–2.9) mg·l^{-1}
Blood volume	Newborn	Plasma volume 41 ml·kg^{-1}	Total erythrocyte volume 43 ml·kg^{-1}
	10–18 years	51 ml·kg^{-1}	26 ml·kg^{-1}
Calcium (S)	Newborn	2.27 (1.76–2.78) mmol·l^{-1}	9.1 (7.1–11.1) mg·dl^{-1}
	Infants	2.39 (2.04–2.73) mmol·l^{-1}	9.6 (8.2–10.9) mg·dl^{-1}
	Children	2.35 (2.09–2.61) mmol·l^{-1}	9.4 (8.4–10.5) mg·dl^{-1}

Table 44.2 *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Calcium ionized (S)	3 days	1.10 (1.03–1.17) mmol·l^{-1}	4.42 (4.14–4.70) mg·dl^{-1}
	3 months	1.15 (1.07–1.24) mmol·l^{-1}	4.62 (4.28–4.96) mg·dl^{-1}
	6 months	1.15 (1.06–1.24) mmol·l^{-1}	4.60 (4.24–4.96) mg·dl^{-1}
	6 years	1.14 (1.04–1.25) mmol·l^{-1}	4.58 (4.16–5.00) mg·dl^{-1}
	6–16 years	1.10 (1.00–1.20) mmol·l^{-1}	4.40 (4.0–4.80) mg·dl^{-1}
Calcium (U)	16–20 years	116 (14–492)µmol·mmol^{-1} creatinine	41 (5–174) µg·mg^{-1} creatinine
Chloride (S)	Newborn	105 (95.2–116) mmol·l^{-1}	
	Infants	103 (92.9–112) mmol·l^{-1}	
	Children	103 (94.5–111) mmol·l^{-1}	
(L)	→ 3 months	108–123 mmol·l^{-1}	
	→ 1 year	113–127 mmol·l^{-1}	
	→ 12 years	117–131 mmol·l^{-1}	
(U)	1 day	0.43 (0.07–0.79) mol·kg^{-1}	
	7 days	2.08 (0.08–0.8) mmol·kg^{-1}	
	Infants	1.61 (0.53–2.69) mmol·kg^{-1}	
	2 years	3.49 (1.59–5.39) mmol·kg^{-1}	
	4–5 years	3.26 (0.28–6.24) mmol·kg^{-1}	
	6–10 years	73.3 (61.7–84.9) mmol·kg^{-1} ♂	
		40.8 (32.6–49.0) mmol·kg^{-1} ♀	
	10–14 years	120 (104–136) mmol·day^{-1} ♂	
		105 (80.6–129 mmol·day^{-1} ♀	
(Sweat)	Infants	12.3 (2.5–22.1) mmol·l^{-1}	
	1–10 years	15.3 (0–31.5) mmol·l^{-1}	
	10–16 years	19.9 (1.5–38.3) mmol·l^{-1}	
Cholesterol total (S) {Enzymatic}	Newborn	1.94 (0.97–3.63) mmol·l^{-1}	75 (38–140) mg·dl^{-1}
	Infants	3.52 (2.34–4.98) mmol·l^{-1}	136 (90–193) mg·dl^{-1}
	2–5 years	4.31 (1.95–6.3) mmol·l^{-1}	167 (75–244) mg·dl^{-1}
	6–9 years	4.54 (3.1–6.63) mmol·l^{-1}	176 (120–246) mg·dl^{-1}
	10–12 years	4.47 (2.66–6.25) mmol·l^{-1}	173 (103–242) mg·dl^{-1}
	> 12 years	4.18 (2.57–5.79) mmol·l^{-1}	162 (99–224) mg·dl^{-1}

Table 44.2 *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
HDL-cholesterol (S) {Electrophor.}	Newborn	0.78 (0.34–1.37) mmol·l^{-1}	30 (13–53) mg·dl^{-1}
	5–18 years	1.22 (0.57–2.3) mmol·l^{-1}	47 (22–89) mg·dl^{-1}
LDL-cholesterol (S) {Electrophor.}	Newborn	2.07 (1.16–3.03) mmol·l^{-1}	80 (45–117) mg·dl^{-1}
	5–18 years	3.4 (1.53–5.61) mmol·l^{-1}	132 (59–217) mg·dl^{-1}
Cholinesterase (S)	1–15 years		5.8 (3.5–8.5) kU·l^{-1}
Chymotrypsin (Stool)	Children and adolescents		20.8 (4.5–43.5) U·g^{-1}
Ceruloplasmin (S)	Newborn		14 (6–20) mg·dl^{-1}
	Children		31 (23–43) mg·dl^{-1}
C-reactive protein (S) {Nephelometry}	1–3 days		≤ 1.2 mg·dl^{-1}
	≥ 4 days		< 0.6 mg·dl^{-1}
Complement (S)			36.9 (28–49) mg·dl^{-1}
C1-inactivated {Immundiff.}			60–220 mg·dl^{-1}
C3-factor {Nephelometry}	Newborn		60–150 mg·dl^{-1}
	1–12 months		80–170 mg·dl^{-1}
	1–2 years		80–120 mg·dl^{-1}
	Children		10– 40 mg·dl^{-1}
C4-factor	Newborn		5–30 mg·dl^{-1}
	1–12 months		10–40 mg·dl^{-1}
CH50 (total hemolytic activity)	Children		33 (20–50) U·l^{-1}

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Creatinine (S) {Jaffe reaction} Creatinine (24-h urine)	Newborn	53.5 (2.15–104) µmol·l^{-1}	0.6 (0.02–1.2) mg·dl^{-1}
	Infants	45.8 (9.71–81.8) µmol·l^{-1}	0.5 (0.1–0.9) mg·dl^{-1}
	Children	55.7 (21.0–90.4)) µmol·l^{-1}	0.6 (0.2–1.0) mg·dl^{-1}
	0–0.5 year	0.13–0.53 mmol·l^{-1}	15–60 mg·day^{-1}
	0.5–1 year	0.49–0.8 mmol·l^{-1}	50–90 mg·day^{-1}
	1–2 years	0.71–1.42 mmol·l^{-1}	80–160 mg·day^{-1}
	2–3 years	0.97–1.59 mmol·l^{-1}	110–180 mg·day^{-1}
	3–4 years	1. 15–2.30 mmol·l^{-1}	130–260 mg·day^{-1}
	4–5 years	1.86–3.45 mmol·l^{-1}	210–390 mg·day^{-1}
	5–7 years	2.30–4.60 mmol·l^{-1}	260–520 mg·day^{-1}
	7–10 years	3.19–6.37 mmol·l^{-1}	360–700 mg·day^{-1}
	11–13 years	8.53 (7.01–10.1) mmol·l^{-1}	792–1140 mg·day^{-1}
	> 13 years	71–265 µmol·kg^{-1}	8–30 mg·kg^{-1}
Creatinine clearance (S) (24-h urine)	5–7 days	50.6 (39.0–62.2) ml·min^{-1}·l^{-1}·0.73 m^{-2}	
	1–2 months	64.6 (53.0–76.2) ml·min^{-1}·l^{-1}·0.73 m^{-2}	
	3–4 months	85.8 (76.2–95.4) ml·min^{-1}·l^{-1}·0.73 m^{-2}	
	5–8 months	87.7 (63.9–112) ml·min^{-1}·l^{-1}·0.73 m^{-2}	
	9–12 months	86.9 (70.1–104) ml·min^{-1}·l^{-1}·0.73 m^{-2}	
	3–6 years	130 (120–140) ml·min^{-1}·l^{-1}·0.73 m^{-2}	
	7–13 years	136 (124–149) ml·min^{-1}·l^{-1}·0.73 m^{-2}	
Creatine kinase CK (S)	Infants		47.7 (16.7–136) U·l^{-1}
	Children		38.8 (15.6–93.8)U·l^{-1}
Creatine kinase MB (S)	> 4 months		0.5–5 U·l^{-1}
Copper (S)	0–0.5 years	3.1–11.0 µmol·l^{-1}	20–70 µg·dl^{-1}
	6 years	14.7–19.8 µmol·l^{-1}	90–190 µg·dl^{-1}
	Children	10.3–21.4 µmol·l^{-1}	66–136 µg·dl^{-1}
Copper (U)		5.7–119 µmol·l^{-1} creatinine	3.2–67 µg·dl^{-1} creatinine

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)			
		SI units		Conventional units	
Dopamine (24-h urine) {HPLC}	3–6 years	1.06 (0.25–1.31) μmol·day^{-1}		163 (38–309) μg·day^{-1}	
		516 (205–852) μmol·mol^{-1} creatinine		603 (239–995) μg·day^{-1} creatinine	
	6–10 years	1.31 (0.28–2.22) μmol·day^{-1}		200 (43–340) μg·day^{-1}	
		410 (73.7–690) μmol·mol^{-1} creatinine		479 (86–806) μg·day^{-1} creatinine	
	10–16 years	1.91 (1.41–2.62) μmol·day^{-1}		292 (216–401) μg·day^{-1}	
		339 (200–586) μmol·mol^{-1} creatinine		396 (234–684) μg·day^{-1} creatinine	
Erythrocyte fragility (Heparin blood) {Osmotic resistance}	Any age			0.46–0.42% NaCl	
				0.34–0.30% NaCl	
Erythrocyte parameter (EDTA blood)	Age	No (×106·μl^{-1})	Mean cell volume (μm^3)	Mean cell hemoglobin (pg)	Mean cell hemoglobin concentration (g·dl^{-1})
	Birth	3.9–6.5	98–118	31–37	30–36
	1–3 days	4–6.6	95–121	31–37	29–37
	7 days	3.9–6.2	88–126	28–40	28–38
	14 days	3.6–6.2	86–119	28–40	28–38
	1 month	3.0–5.4	85–123	28–40	29–37
	2 months	2.7– 4.9	77–118	26–34	29–37
	3–6 months	3.1– 4.5	74–108	25–35	30–36
	0.5–2 years	3.7–5.3	70–86	23–31	30–36
	2–6 years	3.9–5.3	75–87	24–30	31–37
	6–12 years	4.0–5.2	77–95	25–33	31–37
	♀ 12–18 years	4.1–5.1	78–102	25–35	31–37
	♂ 12–18 years	4.5–5.3	78–98	25–35	31–37

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Ferritin (S)	14 days		23.8 (9–62.8) µg·dl^{-1}
	1 month		24.0 (14.4–39.9) µg·dl^{-1}
	2 months		19.4 (8.7–43.0) µg·dl^{-1}
	4 months		9.1 (3.7–22.3) µg·dl^{-1}
	6 months		5.1 (1.9–14.2) µg·dl^{-1}
	9 months		3.9 (1.4–10.3) µg·dl^{-1}
	12 months		3.1 (1.1–9.1) µg·dl^{-1}
	1–10 years		4.3 (1.5–11.9) µg·dl^{-1}
α_1-Feto-protein (S) {RIA, EIA}	Preterm		13.4 (9.0–21.8) mg·dl^{-1}
	Newborn		4.8 (2.0–11.8) mg·dl^{-1}
	→ 14 days		3.3 (1.0–9.8) mg·dl^{-1}
	14–28 days		0 95 (0.05–3.5) mg·dl^{-1}
	6 months		1.3 (0.5–3.2) mg·dl^{-1}
Fatty acids (free) (P)	Newborn	435–1375 µmol·l^{-1}	
	4 months – 10 years	500–900 µmol·l^{-1}	
Folic acid (P), (S) {RIA}		4.3–23.6 nmol·l^{-1}	1.9–14.0 ng·ml^{-1}
Fructose (B) (U)		<10 mg·dl^{-1}	<0.6 mmol·l^{-1}
	Newborn	<3.9 mmol·l^{-1}	<70 mg·dl^{-1}
	Infants	<1.1 mmol·l^{-1}	<20 mg·dl^{-1}
	Children	<0.06 mmol·day^{-1}	<10 mg·day^{-1}
Coagulation factors			
(Citrat-P)	1 day		245 (190–300) mg·dl^{-1}
Fibrinogen	> 1 day		315 (255–375) mg·dl^{-1} = adult values (100%)
Factor II	Newborn	45 (30–60)%	6 months later
Factor V	Newborn	100 (60–140)%	After birth
Factor VII	Newborn	55 (40–70)%	6 months later

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Factor VIII	Newborn	105 (70–140)%	After birth
Factor IX	Newborn	30 (20–40)%	6 months later
Factor X	Newborn	55 (40–70)%	After 2 months
Factor XI	Newborn	30%	6 months later
Factor XII	Newborn	50%	After 2 months
Factor XIII	Newborn	100 (70–130)%	After birth
Antithrombin III	Newborn	45 (30–80)%	3–6 months later
Plasminogen	Newborn	50%	6 months later
Protein C	Newborn	35 (20–50)%	12 months later
Protein S	Newborn	35 (10–60)%	6 months later
Fibrin products	Any age	< 10 µg·ml^{-1}	
Thromboplastin time, TPZ	Newborn	80 (40–100)%	
Partial thromboplastin time (PTT)	< 6 months	38 (30–47) sec	
	Children	33 (25–41) sec	
Thrombin time (plasma thrombin time)	> 1 month	9–11 sec	
Glucose (B)	1 day	2.22–5.55 mmol·l^{-1}	40–100 mg·dl^{-1}
	2 days	2.22–5.0 mmol·l^{-1}	40–90 mg·dl^{-1}
	5 days	2.22–4.16 mmol·l^{-1}	40–75 mg·dl^{-1}
	Infants	3.33–5.0 mmol·l^{-1}	60–90 mg·dl^{-1}
	Children	3.33–5.55 mmol·l^{-1}	60–100 mg·dl^{-1}
Glucose-6-phosphate-dehydrogenase (B)			131 (105–157) mU·l0^{-9} erythrocytes
Glucose tolerance test (B)	Children		1-h value: ≤ 8.3 mmol·l^{-1} 150 mg·dl^{-1} 2-h value: ≤ 7.3 mmol·l^{-1} 131 mg·dl^{-1}

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Glutamate dehydrogenase GLDH (S)	Newborn		1.9 (0–6.6) U·l^{-1}
	1–6 months		1.5 (0–4.3) U·l^{-1}
	7–12 months		0.9 (0–3.5) U·l^{-1}
	2–3 years		0.9 (0–2.6) U·l^{-1}
	3–5 years		→ 3.2 U·l^{-1}
	Children		→ 3.5 U·l^{-1}
Glutamate oxaloacetate transaminase ASAT (S)	Newborn		21.9 (5.91–37.9) U·l^{-1}
	Infants		17.3 (7.38–27.3) U·l^{-1}
	Children		13.4 (4.7–22.2) U·l^{-1}
Glutamate pyruvate transaminase ALAT (S)	Newborn		12.4 (4.47–32.4) U·l^{-1}
	Infants		14.9 (6.23–35.7) U·l^{-1}
	Children		9.61 (4.5–20.5) U·l^{-1}
γ-Glutamyl transpeptidase GGT (S)	Newborn		47.8 (13.9–163) U·l^{-1}
	2–3 months		13.3 (1.95–90.8) U·l^{-1}
	> 4 m		7.32 (3.1–17.3) U·l^{-1}
Hematocrit (EDTA-B)	1 day		58 (45–72)%
	7 days		55 (43–67)%
	14 days		50 (42–66)%
	1 month		43 (31–55)%
	2 months		35 (28–42)%
	3–6 months		36 (33–39)%
	0.5–2 years		37 (34–40)%
	2–6 years		40 (35–45)%
	6–12 years		41 (36–46)%
	12–18 years		43 (37–49)%

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Hemoglobin (total) (EDTA-B)	1 day	12.1 (9.0–14.5) mmol·l^{-1}	19.5 (14.5–23.4) g·dl^{-1}
	7 days	10.9 (8.7–13.7) mmol·l^{-1}	17.5 (14.0–22.0) g·dl^{-1}
	14 days	10.2 (8.1–12.4) mmol·l^{-1}	16.5 (13.0–20.0) gl·dl^{-1}
	1 month	8.7 (6.2–11.1) mmol·l^{-1}	14.0 (10.0–18.0) g·dl^{-1}
	2 months	7.1 (5.6–8.7) mmol·l^{-1}	11.5 (9.0–14.0) g·dl^{-1}
	3–6 months	7.1 (5.9–8.4) mmol·l^{-1}	11.5 (9.5–13.5) g·dl^{-1}
	0.5–2 years	7.5 (6.5–8.4) mmol·l^{-1}	12.0 (10.5–13.5) g·dl^{-1}
	2–6 years	7.8 (7.1–8.4) mmol·l^{-1}	12.5 (11.5–13.5) g·dl^{-1}
	6–12 years	8.4 (7.1–9.6) mmol·l^{-1}	13.5 (11.5–15.5) g·dl^{-1}
	12–18 years	♀ 8.7 (7.5–9.9) mmol·l^{-1}	14.0 (12.0–16.0) g·dl^{-1}
		♂ 9.0 (8.1–9.9) mmol·l^{-1}	14.5 (13.0–16.0) g·dl^{-1}
Hemoglobin A_2 (EDTA-B) {Chromatography}	Newborn		0.19–0.6%
	1 month		0.71–1.38%
	2 months		1.08–2.01%
	3 months		1.44–2.08%
	5 months		1.50–2.38%
	6 months		1.60–2.40%
	Children		2.08–3.17%
Hb $A1_c$ (glycosylated Hb) (EDTA-B)	6–12 months		10.0 (5.4–14.6)%
	→ 2 years		9.2 (7.0–11.4)%
	→ 4 years		7.7 (5.7–9.7)%
	5–12 years		7.1 (5.3–8.9)%
	13–20 years		7.1 (5.5–8.7)%
CO-Hb (B) {Hemoglobin-electrophor.}		1.2% from total hemoglobin	

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Hb 1 (EDTA-B)		$Hb\ A_1$ / $Hb\ A_2$	Hb F
	Newborn	17.7 (13–22)% / 0.25 (0.05–0.45)%	81.7 (77–86)%
	Adult	97% / 2.5%	0.5%
	7 days	60.1 (51.4–68.3)%	
	1 month	45.5 (30.3–58.3)%	
	2 months	26.6 (18.8–39.2)%	
	3 months	14.5 (4.5–27.1)%	
	4 months	9.6 (0.2–15.9)%	
	6 months	1.0 (0–8.4)%	
	1 year	≤ 1.3%	
Fe_2+ Hb (B)	Newborn	0–0.37 $mmol \cdot l^{-1}$	0–0.58 $g \cdot dl^{-1}$
	Infants	0–0.19 $mmol \cdot l^{-1}$	0–0.29 $g \cdot dl^{-1}$
	Children	0–0.2 $mmol \cdot l^{-1}$	0–0.33 $g \cdot dl^{-1}$
Haptoglobin (S) {Nephelometry}	→ 7 days		40 $mg \cdot dl^{-1}$
	→ 1 year		110 $mg \cdot dl^{-1}$
	Children		10–140 $mg \cdot dl^{-1}$
Homovanillic acid (24-h urine) {HPLC}	3–6 years	14.3 (7.7–23.6) $\mu mol \cdot l^{-1}$	2.6 (1.4–4.3) $mg \cdot day^{-1}$
		6.1 (3.4–9.6) $mmol \cdot mol^{-1}$ creatinine	9.9 (5.4–15.5) $mg \cdot g^{-1}$ creatinine
	6–10 years	19.8 (11.5–25.8) $\mu mol \cdot l^{-1}$	3.6 (2.1–4.7) $mg \cdot day^{-1}$
		5.0 (2.7–7.1) $mmol \cdot mol^{-1}$ creatinine	8.0 (4.4–11.5) $mg \cdot g^{-1}$ creatinine
	10–16 years	23.6 (13.2–47.7) $\mu mol \cdot l^{-1}$	4.3 (2.4–8.7) $mg \cdot day^{-1}$
		3.3 (2.0–6.4) $mmol \cdot mol^{-1}$ creatinine	5.3 (3.3–10.3) $mg \cdot g^{-1}$ creatinine

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Iron (S)	14 days	11–36 µmol·l^{-1}	63–201 µg·dl^{-1}
	6 months	5–24 µmol·l^{-1}	28–135 µg·dl^{-1}
	1 year	6–28 µmol·l^{-1}	35–155 µg·dl^{-1}
	2–12 years	4–24 µmol·l^{-1}	22–135 µg·dl^{-1}
Iron-binding capacity (S)	14 days	34 (18–50) µmol·l^{-1}	191 (105–277) µg·dl^{-1}
	6 months	58 (40–76) µmol·l^{-1}	321 (219–423) µg·dl^{-1}
	1 year	64 (50–78) µmol·l^{-1}	358 (282–434) µg·dl^{-1}
	Children	59 (43–76) µmol·l^{-1}	331 (239–423) µg·dl^{-1}
Immunoglobulin (S) {Nephelometry}			
IgA	Newborn		<10 mg·dl^{-1}
	Infants		10–70 mg·dl^{-1}
	1–3 years		20–130 mg·dl^{-1}
	Children		40–240 mg·dl^{-1}
IgG	7 days		700–2000 mg·dl^{-1}
	1–12 weeks		150–900 mg·dl^{-1}
	3–12 weeks		200–800 mg·dl^{-1}
	1–3 years		400–1300 mg·dl^{-1}
	4–7 years		600–1600 mg·dl^{-1}
	> 8 years		700–1800 mg·dl^{-1}
IgM	Newborn		<20 mg·dl^{-1}
	Infants		20–100 mg·dl^{-1}
	1–3 years		50–200 mg·dl^{-1}
	Children		50–220 mg·dl^{-1}
IgE {Enzyme-immunoassay}	Newborn		1.5 IU·ml^{-1}
	Infants		15 IU·ml^{-1}
	1–5 years		60 IU·ml^{-1}
	6–9 years		90 IU·ml^{-1}
	10–15 years		200 IU·ml^{-1}

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Lactate (P)	1 h	0.9–2.7 mmol·l^{-1}	8.1–24.3 mg·dl–1
	5 h	0.9–2.0 mmol·l^{-1}	8.1–18.0 mg·dl–1
	1 day	0.8–1.2 mmol·l^{-1}	7.2–10.8 mg·dl–1
	7 days	0.5–1.4 mmol·l^{-1}	4.5–12.6 mg·dl–1
	Children	0.9–1.8 mmol·l^{-1}	8.1–16.2 mg·dl–1
(L)		0.9–2.8 mmol·l^{-1}	8–25 mg·dl^{-1}
Lactate dehydrogenase LDH (S)	Newborn		410 (200–838) U·l^{-1}
	Infants		285 (156–521) U·l^{-1}
	Children		213 (131–344) U·l^{-1}
Leucine aminopeptidase LAP (S)	Newborn		15–33 U·l^{-1}
	1–6 months		13–39 U·l^{-1}
	7–12 months		15–35 U·l^{-1}
	1–2 years		13–31 U·l^{-1}
	2–3 years		12–31 U·l^{-1}
	Children		14–36 U·l^{-1}
Lipase (S)	Newborn		→ 80 U·l^{-1}
	Children		→ 115 U·l^{-1}
Leukocytes (L)	Newborn	0–20·µl^{-1}	
	Infants	0–5·µl^{-1}	
	Children	0–4·µl^{-1}	

Leukocytes (EDTA-B)	Age	Total No.	Neutrophils	No./µl % Lymphocytes	Monocytes	Eosinophils
	1 h	18.1 (9.0–30.0)	11.0 (6.0–26.0) 61%	5.5 (2.0-11.0) 31 %	1.1; 6%	0.4; 2%
	12 h	22.8 (13.0–38.0)	15.5 (6.0–28.0) 68%	5.5 (2.0-11.0) 24 %	1.2; 5%	0.5; 2%
	1 day	18.9 (9.4–34.4)	11.5 (5.0–21.0) 61%	5.8 (2.0-17.5) 31 %	1.1; 6%	0.5; 2%

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)			
		SI units		Conventional units	
	7 days	12.2 (5.0–21.0)	5.5 (1.5–10.0) 45%	5.0 (2.0-17.0) 41 %	1.1; 9% 0.5; 4%
	14 days	11.4 (5.0–20.0)	4.5 (1.0–9.5) 40%	5.5 (2.0-17.0) 48 %	1.0; 9% 0.4; 3%
	1 month	10.8 (5.0–19.5)	3.8 (1.0–9.0) 35%	6.0 (2.5-16.5) 56 %	0.7; 7% 0.3; 3%
	6 months	11.9 (6.0–17.5)	3.8 (1.0–8.5) 32%	7.3 (4.0-13.5) 61 %	0.5; 5% 0.3; 3%
	1 year	11.4 (6.0–17.5)	3.5 (1.5–8.5) 31%	7.0 (4.0-10.5) 61 %	0.6; 5% 0.3; 3%
	2 years	10.6 (6.0–17.0)	3.5 (1.5–8.5) 33%	6.3 (3.0-9.5) 59 %	0.5; 5% 0.3; 3%
	4 years	9.1 (5.5–15.5)	3.8 (1.5–8.5) 42%	4.5 (2.0-8.0) 50 %	0.5; 5% 0.3; 3%
	6 years	8.5 (5.0–14.5)	4.3 (1.5–8.0) 51%	3.5 (1.5-7.0) 42 %	0.4; 5% 0.2; 3%
	8 years	8.3 (4.5–13.5)	4.4 (1.5–8.0) 53%	3.3 (1.5-6.8) 39 %	0.4; 4% 0.2; 2%
	10 years	8.1 (4.5–13.5)	4.4 (1.8–8.0) 54%	3.1 (1.5-6.5) 38 %	0.4; 4% 0.2; 2%
	16 years	7.8 (4.5–13.0)	4.4 (1.8–8.0) 57%	2.8 (1.5-5.2) 35 %	0.4; 5% 0.2; 3%
	21 years	7.4 (4.5 –11.0)	4.4 (1.8–7.7) 59%	2.5 (1.0-4.8) 34 %	0.3; 4% 0.2; 3%

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Potassium (S) {Photometry} (U)	Newborn	4.84 (3.56–6.11) $mmol \cdot l^{-1}$	
	Infants	4.74 (3.65–5.83) $mmol \cdot l^{-1}$	
	Children	4.14 (3.13–5.15) $mmol \cdot l^{-1}$	
	1 day	0.36 (0.08–0.5) $mmol \cdot kg^{-1} \cdot day^{-1}$	
	2 days	0.45 (0.17–0.93) $mmol \cdot kg^{-1} \cdot day^{-1}$	
	7 days	0.95 (0–2.25) $mmol \cdot kg^{-1} \cdot day^{-1}$ (breast-fed infants)	
		2.11 (0.51–3.71) $mmol \cdot kg^{-1} \cdot day^{-1}$ (formula-fed infants)	
	1–6 months	2.27 (0–4.89) $mmol \cdot kg^{-1} \cdot day^{-1}$	
	1–2 years	4.06 (2.94–5.18) $mmol \cdot kg^{-1} \cdot day^{-1}$	
	4–5 years	2.33 (0.83–3.83) $mmol \cdot kg^{-1} \cdot day^{-1}$	
	6–10 years	22.5 (17.4–27.5) $mmol \cdot day^{-1}$ ♀	
		31.9 (27.0–36.7) $mmol \cdot day^{-1}$ ♂	
	10–14 years	38.0 (30.9–45.0) $mmol \cdot day^{-1}$ ♀	
		39.3 (34.4–44.3) $mmol \cdot day^{-1}$ ♂	
Renin (EDTA-P)		2.15 (1.72–2.59) $\mu g \cdot l^{-1} \cdot h^{-1}$	
Reticulocytes (EDTA-B)	1 day		20–60‰
	7 days		3–10‰
	14 days		0–10‰
Thrombocytes (B) {Machine count}		(150–500)·103·μl^{-1}	
Transferrin (S)	14 days		158–268 $mg \cdot dl^{-1}$
	0.5–6 months		202–302 $mg \cdot dl^{-1}$
	0.5–1 year		261–353 $mg \cdot dl^{-1}$
	1–16 years		240–360 $mg \cdot dl^{-1}$
	>16 years		200–340 $mg \cdot dl^{-1}$

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Triglycerides (S)	Newborn	0.66 (0.12–2.60) mmol·l^{-1}	58 (11–230) mg·dl^{-1}
	Infants	1.29 (0.50–2.32) mmol·l^{-1}	114 (44–205) mg·dl^{-1}
	<6 years	1.04 (0.42–2.09) mmol·l^{-1}	92 (37–185) mg·dl^{-1}
	6–9 years	0.72 (0.32–1.39) mmol·l^{-1}	64 (28–123) mg·dl^{-1}
	10–12 years	0.74 (0.26–1.80) mmol·l^{-1}	65 (23–159) mg·dl^{-1}
	Children	0.77 (0.33–1.70) mmol·l^{-1}	68 (29–150) mg·dl^{-1}
Urine – Erythrocytes – Leukocytes		<5·µl^{-1} <10·µl^{-1}	
Volume (per 24 h)	≤6 months	35 ml·kg^{-1}·day^{-1}	
	≤2 years	200 ml·day^{-1}	
	≤7 years	400 ml·day^{-1}	
	≤15 years	800 ml·day^{-1}	
	>15 years	≤2000 ml·day^{-1}	

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Uric acid (S)	Newborn	182 (38.2–326) $\mu mol \cdot l^{-1}$	3.1 (0.6–5.5) $mg \cdot dl^{-1}$
	Infants	197 (68.1–325) $\mu mol \cdot l^{-1}$	3.3 (1.1–5.5) $mg \cdot dl^{-1}$
	Children	232 (111–353) $\mu mol \cdot l^{-1}$	3.9 (1.9–5.9) $mg \cdot dl^{-1}$
Uric acid (U)	7 days	1.55 (0.18–2.91) g uric acid·g^{-1} creatinine	1.04 (0.12–1.96) mol uric acid·mol^{-1} creatinine
	1 year	1.45 (0.62–2.27) g uric acid·g^{-1} creatinine	0.98 (0.42–1.53) mol uric acid·mol^{-1} creatinine
	2 years	2.37 (0.85–2.01) g uric acid·g^{-1} creatinine	1.60 (0.57–1.35) mol uric acid·mol^{-1} creatinine
	6 years	0.88 (0.59–1.26) g uric acid·g^{-1} creatinine	0.59 (0.30–0.85) mol uric acid·mol^{-1} creatinine
	10 years	0.61 (0.22–1.00) g uric acid·g^{-1} creatinine	0.41 (0.15–0.67) mol uric acid·mol^{-1} creatinine
	18 years	0.40 (0.16–0.69) g uric acid·g^{-1} creatinine	0.30 (0.11–0.52) mol uric acid·mol^{-1} creatinine
Urea-N (S)	Newborn	3.93 (1.06–6.79) $mmol \cdot l^{-1}$	11.0 (3.0–19.0) $mg \cdot dl^{-1}$
	Infants	4.60 (2.04–7.17) $mmol \cdot l^{-1}$	12.9 (5.7–20.1) $mg \cdot dl^{-1}$
	Children	5.06 (2.12–8.0) $mmol \cdot l^{-1}$	14.2 (6.0–22.5) $mg \cdot dl^{-1}$
Urea-N (U)	Newborn		0.15–1.0 $g \cdot day^{-1}$
	Infants		1.0–4.0 $g \cdot day^{-1}$
	Children		4.0–8.0 $g \cdot day^{-1}$
	Adolescents		8.0–20.0 $g \cdot day^{-1}$

Table 44.2. *(continued)* Laboratory values of commonly used examinations. (*B* Blood, *L* liquor, *P* plasma, *S* serum, *U* urine)

Substance (Material) {Method}	Age groups	Reference values (x +2SD)	
		SI units	Conventional units
Vanillin almond acid (24-h urine)	Newborn	1.8 (0.6–3.0) µmol·day^{-1}	0.35 (0.11–0.59) mg·day^{-1}
	1–6 months	3.2 (0.3–6.1) µmol·day^{-1}	0.64 (0.06–1.22) mg·day^{-1}
	0.5–2 years	5.4 (3.0–7.8) µmol·day^{-1}	1.08 (0.60–1.56) mg·day^{-1}
	2–4 years	8.1 (4.3–11.9) µmol·day^{-1}	1.61 (0.85–2.37) mg·day^{-1}
	6–8 years	9.7 (6.3–13.1) µmol·day^{-1}	1.93 (1.25–2.61) mg·day^{-1}
	8–10 years	11.7 (7.9–l5.5) µmol·day^{-1}	2.33 (1.57–3.09) mg·day^{-1}
	10–12 years	14.7 (9.1–20.3) µmol·day^{-1}	2.93 (1.81–4.05) mg·day^{-1}
	12–15 years	18.8 (8.6–28.9) µmol·day^{-1}	3.73 (1.71–5.75) mg·day^{-1}
Zinc (S)	Newborn	11.6 (7.9–15.3) µmol·l^{-1} ♀	75.7 (51.7–99.7) µg·dl^{-1}
		11.2 (7.6–13.3) µmol·l^{-1} ♂	73.5 (49.5–87) µg·dl^{-1}
Zinc (U)	Children	12.6 (9.8–16.8) µmol·l^{-1}	82.5 (63.8–110) µg·dl^{-1}
		595 (155–1470) µmol·mol^{-1} creatinine	

45 Commonly Used Pharmaceuticals

See next page for Table 45.1.

Table 45.1 Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Acetazolamide		Initially: 25 mg·kg^{-1}·day^{-1} Daily increase up to 100 mg·kg^{-1}·day^{-1}	i.v., p.o. 3×	Hydrocephalus therapy: Lasix 1–2 mg·kg^{-1}·day^{-1} Sodium hydrogen carbonate 1 mg·kg^{-1}·day^{-1} K^+, Na^+
Acetylcysteine	100 mg·ml^{-1} 20 mg·ml^{-1}	10 mg·kg^{-1}·day^{-1}	i.v., p.o., 3×	Intratrachial: 1:10 diluted Antidote for paracetamol intoxication 300 mg·kg^{-1}·20 h^{-1} in 3 doses (150 mg/50 mg/100 mg)
Aciclovir	25 mg·ml^{-1} 200 mg/tbl	30 mg·kg^{-1}·day^{-1}	i.v., p.o. 3×	Infusions over 30 min
Adrenaline	1 mg·ml^{-1} (1:10,000)	0.1^{-1}.0 µg·kg^{-1}·min^{-1}	0.1 ml·kg^{-1}·h^{-1} if diluted	Dilution: 3 ml + 47 ml G 5% = 60 µg·ml^{-1}
Allopurinol	100 mg·tbl^{-1} 300 mg·tbl^{-}	10 mg·kg^{-1}·day^{-1}	3×	

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Amphotericin B	5 mg·ml^{-1} 1 mg·0.2 ml^{-1}	0.1 mg·kg^{-1}·day^{-1} 0.3 mg·kg^{-1}·day^{-1} 0.5 mg·kg^{-1}·day^{-1}	First day: 0.02 ml·kg^{-1}·day^{-1} Second day: 0.06 ml·kg^{-1}·day^{-1} Days 3–7: 0.1 ml·kg^{-1}·day^{-1} Every 2 days: 0.1 ml·kg^{-1}·day^{-1}	Applied as infusion, diluted in G 5% every 6 h K^+ ↓, Bc ↓, Temp↑ Nephrotoxic, hepatotoxic
Atracurium	25 mg/2.5 ml = 10 mg·ml^{-1}	0.3–0.6 mg·kg^{-1}	i.v. 0.03–0.06 ml·kg^{-1}	
Atropine	0.5 mg·ml^{-1}	0.02 mg·kg^{-1}	i.v., s.c. 0.04 ml·kg^{-1} 0.1 ml·2.5 kg^{-1}	Temp↑, HR ↑
AT III	500 U/10 ml = 50 U/ml		1 U·kg^{-1}	Rise to 1%$^{-1}$–1.5%
β-Acetyldigoxin	0.2 mg·ml^{-1} 0.1 mg·tbl^{-1}	Newborns: 0.03 mg·kg^{-1} for 36 h then 0.005 mg·kg^{-1}·day^{-1} 4 weeks old: 0.04 mg·kg^{-1} for 36 h then 0.01 mg·kg^{-1}·day^{-1}	 0.15 ml·kg^{-1} for 36 h 2×0.12 ml·kg^{-1} 1:10 diluted 0.2 ml·kg^{-1} for 36 h 2×0.25 ml·kg^{-1} 1:10 diluted	By K^+ ↓: AV disturbances Bradycardia Ventricular tachycardia EHL 35–70 h Level: 1.0–2.5 mg·ml^{-1}

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Calcium gluconate	1 mmol Ca^{2+}/tbl 0.225 mmol·ml^{-1} = 0.9 mval·ml^{-1}	Theoretically required: 4 mmol Ca^{2+}·kg^{-1}·day^{-1}	2×½ tbl	1 mmol Ca^{2+} = 40 mg Ca
Calcium gluconate 10%			i.v. 0.5^{-1}.0 ml·kg^{-1}	Inject slowly
Captopril	25 mg·tbl^{-1} 50 mg·tbl^{-1}	0.3–5 mg·kg^{-1}·day^{-1}	0.2 mg·kg^{-1} then slowly increase	Angiotensin converting enzyme blocker
Ceruletide	40 µg/2 ml = 20 µg·ml^{-1}	2 ng·kg^{-1}·min^{-1} over 15–80 min	i.v. 1.5 ml·kg^{-1}·h^{-1} if diluted	Dilution: 2 ml in 500 ml NaCl 0.9% = 80 ng·ml^{-1} EHL 3–5 min
Chloral hydrate	0.6 g, 3 g = 200 mg·ml^{-1}	30–50 mg·kg^{-1}	p.o., rectal 0.2 ml·kg^{-1}	
Cisapride	1 mg·ml^{-1}	0.6 mg·kg^{-1}·day^{-1}	p.o. 3×	Stimulates bowel motility, antagonizes morphine effect
Clemastine	2 mg·5 ml^{-1} 0.5 mg·10 ml^{-1}		i.v., i.m. 0.6 ml·10 kg^{-1} 2×	Alertness ↓
Clenbuterol	1 µg·ml^{-1} syr 59 µg·ml^{-1} dr	0.8^{-1}–1.5 µg·kg^{-1}·day^{-1}	0.8^{-1}–1.5 ml·kg^{-1}·day^{-1}	Sympathicomimetic, anabolic
Clonazepam	1 mg/2 ml = 0.5 mg·ml^{-1}	0.05–0.1 mg·kg^{-1}	0.1–0.2 ml·kg^{-1}	May induce tonic convulsion

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Colistin	1,000,000 IU/ 3 ml = 333,000 IU·ml^{-1} 500,000 IU·tbl^{-1}	100,000 IU·kg^{-1}·day^{-1} to 200,000 IU·kg^{-1}·day^{-1}	0.3–0.6 ml·kg^{-1}·day^{-1}	Bowel sterilization without resorption
Dantrolene	20 mg/amp	1–2.5 mg·kg^{-1} then 7.5 mg·kg^{-1}·day^{-1}	Until 30 mg·kg^{-1} total doses	Myotonolytic Intracellular calcium antagonist for malignant hyperthermia
Dexamethasone	4 mg·ml^{-1}	Days 1–3: 2×0.25 mg·kg^{-1} Days 4–6: 1×0.30 mg·kg^{-1} Days 7–9: 1×0.27 mg·kg^{-1} Days 25–27: 0.1 mg·kg^{-1} Days 29/31/34: 0.1 mg·kg^{-1}		In BPD longer treatment more effective
Dexamethasone	4 mg·ml^{-1}	0.6 mg·kg^{-1}·day^{-1}	i.v. 3–4× 0.15 ml·kg^{-1}·day^{-1}	

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Dexpanthenol	500 mg·2 ml^{-1}		Infants: 1–2 ml·day^{-1} Children: 2–4 ml·day^{-1} 4–6 ml·day^{-1}	Bowel atony, paralytic ileus
Diazepam	10 mg·2 ml^{-1}	0.5 mg·kg^{-1}	0.1 ml·kg^{-1}	Long EHL, mucous obstruction, cerebral perfusion ↓
Dimetindene	1 mg·20 dr^{-1} 1 mg·cps^{-1} 1 mg·ml^{-1}		Infants: 3×4–8 dr 1–8 years: 3×10–15 dr 9^{-1}4 years: 3×20 dr	
Piritramide (Dipidolor®)	7.5 mg·ml^{-1}	0.1 mg·kg^{-1} (0.05–0.3 mg·kg^{-1})	0.1 ml·7.5 kg^{-1}	Longer and stronger effect than fentanyl, less effective than morphine
Dobutamine	250 mg/50 ml = 5 mg·ml^{-1}	5 µg·kg^{-1}·min^{-1} to 20 µg·kg^{-1}·min^{-1}	0.1 ml·kg^{-1}·h^{-1} if diluted	Dilution: 30 ml + 20 ml G 5% = 3 mg·ml^{-1}

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Dopamine	250 mg/50 ml = 5 mg·ml^{-1} 50 mg/5 ml = 10 mg·ml^{-1}	4 µg·kg^{-1}·min^{-1} = 1.152 ml·kg^{-1}·day^{-1} 4 µg·kg^{-1}·min^{-1} = 0.576 ml·kg^{-1}·day^{-1}	0.1 ml·kg^{-1}·h^{-1}. if diluted	Dilution 12 ml + 38 ml G 5% = 2.4 mg·ml^{-1} GFR ↑, Na-secretion ↑
Droperidol	2.5 mg·ml^{-1}	0.1–0.2 mg·kg^{-1} Prf: 0.1 mg·kg^{-1}·h^{-1}	0.04–0.08 ml·kg^{-1}	Strong neuroleptic, RR ↓, extrapyramidal symptoms
Droperidol + fentanyl	2.5 mg droperidol + 0.05 mg fentanyl per ml		0.035 ml·kg^{-1}·h^{-1}	RR ↓
Esmolol hydrochloride	100 mg/10 ml = 10 mg·ml^{-1}	0.5 mg·kg^{-1}	0.05 ml·kg^{-1}·min^{-1} = 0.1 ml·2 kg^{-1}	β-Blocker with short EHL (4 min)
Fe^{2+}	44 mg·ml^{-1} (= 22 dr) 2 mg·dr^{-1}	2 mg·kg^{-1}·day^{-1}	Initially 1 dr·day^{-1} for 7 days then 1 dr·kg^{-1}·day^{-1}	
Fentanyl	50 µg·ml^{-1}	5 µg·kg^{-1} 3 µg·kg^{-1}·h^{-1}	0.1 ml·kg^{-1} 0.1 ml·kg^{-1}·h^{-1} if diluted	Dilution: 30 ml + 20 ml G 5% = 30 µg·ml^{-1} RR ↓, constipation, bronchospasm
Flunitrazepam	2 mg·ml^{-1} 2 mg·tbl^{-1}	0.01–0.03 mg·kg^{-1} 0.01 mg·kg^{-1}·h^{-1}	0.1 ml·5 kg^{-1}	Dilution: 2 mg·2 ml^{-1}

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Folic acid	2 $mg \cdot ml^{-1}$	0.5 $mg \cdot kg^{-1} \cdot week^{-1}$	i.v. 0.25 $ml \cdot kg^{-1} \cdot week^{-1}$	
Furosemide	10 $mg \cdot ml^{-1}$ 40 $mg \cdot tbl^{-1}$	1 $mg \cdot kg^{-1}$	0.1 $ml \cdot kg^{-1}$	Total doses: up to 10 $mg \cdot kg^{-1} \cdot day^{-1}$
Ganciclovir	50 $mg \cdot ml^{-1}$	10 $mg \cdot kg^{-1}$	i.v. 0.5 $ml \cdot kg^{-1}$ if diluted	Dilution: 1 ml + 4 ml Ringer = 10 $mg \cdot ml^{-1}$
Glycerol trinitrate	10 $mg \cdot ml^{-1}$ = 1 $mg \cdot ml^{-1}$	1 $\mu g \cdot kg^{-1} \cdot min^{-1}$	0.1 $ml \cdot kg^{-1} \cdot h^{-1}$ if diluted	Dilution: 30 ml + 20 ml G 5% = 0.6 $mg \cdot ml^{-1}$
Heparin	100 $IU \cdot ml^{-1}$	CVA: 50 $IU \cdot kg^{-1} \cdot day^{-1}$ Low dose: 100 $IU \cdot kg^{-1} \cdot day^{-1}$ High dose: 400 $IU \cdot kg^{-1} \cdot day^{-1}$	0.5 $ml \cdot kg^{-1} \cdot day^{-1}$ 1.0 $ml \cdot kg^{-1} \cdot day^{-1}$ 4.0 $ml \cdot kg^{-1} \cdot day^{-1}$	
Hydrochlorothiazide	25 $mg \cdot tbl^{-1}$	1 $mg \cdot kg^{-1} \cdot day^{-1}$ BPD: 2–4 $mg \cdot kg^{-1} \cdot day^{-1}$ + aldactone	3×	In ascites combined with aldactone

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Insulin	40 $IU \cdot ml^{-1}$	0.1 $IU \cdot kg^{-1} \cdot h^{-1}$	0.1 $ml \cdot kg^{-1} \cdot h^{-1}$ if diluted	Dilution: 1 ml + 30 ml NaCl 0.9% + 9 ml Hum-Alb5% = 1 $IU \cdot ml^{-1}$ If Bg is very high then additional insulin application with a Prf. 0.05 $IE \cdot kg^{-1} \cdot h^{-1}$
Inzolen® infant	Mg, Fe, Zn, Cu Mn, Cr, Co, F, I		1 $ml \cdot kg^{-1} \cdot day^{-1}$	For newborns and premature infants
KCl	1000 mg tbl 13.4 mval K^+ 800 mg tbl = 8.05 mval K^+			
Ketamine	10 $mg \cdot ml^{-1}$ 50 $mg \cdot ml^{-1}$	1 (–2) $mg \cdot kg^{-1}$ 3 (–4) $mg \cdot kg^{-1}$ 0.5 (–3) $mg \cdot kg^{-1} \cdot h^{-1}$	i.v., i.m. 0.1 $ml \cdot kg^{-1}$ 0.02 $ml \cdot kg^{-1}$	RR ↑, HR ↑, salivation ↑ cerebral pressure ↑, contraindicated in newborns
Lidocaine	100 mg/5 ml = 20 $mg \cdot ml^{-1}$	1 $mg \cdot kg^{-1}$ 20 $\mu g \cdot kg^{-1} \cdot min^{-1}$	0.05 $ml \cdot kg^{-1}$ 0.1 $ml \cdot kg^{-1} \cdot h^{-1}$ if diluted	Dilution: 30 ml + 20 ml G 5% = 12 $mg \cdot ml^{-1}$ In ventricular extrasystoles repeat 3×

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Magnesium	0.54 mmol·ml^{-1} = 1.08 mval·ml^{-1}		0.2 ml·kg^{-1} parenteral alimentation: ca. 0.25 ml·kg^{-1}·day^{-1} = 1 ml·4 kg^{-1}·day^{-1}	Hypotonic muscles Apnea
Mannitol 20%	20 g·100 ml^{-1}	0.25 g·kg^{-1}	i.v. 1.25–2.5 ml·kg^{-1}	Max. 1–2 g·kg^{-1} in 2–6 h
Metamizole	1 g/2 ml = 500 mg·ml^{-1} 20 mg·dr^{-1}	10–20 mg·kg^{-1}	1 dr·kg^{-1}	Allergic reaction, agranulocytosis
Metildigoxin	0.2 mg/2 ml = 0.1 mg·ml^{-1} 0.6 mg /45 dr = 0.013 mg/dr		kg dr/day 3–5 3 6 4 7–8 5 9–10 6 11–12 7 13–14 8 15–16 9 17–18 10 19 11 20–21 12	w

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Midazolam	15 mg/3 ml = 5 $mg \cdot ml^{-1}$	0.1 $mg \cdot kg^{-1}$ 0.18 $mg \cdot kg^{-1} \cdot h^{-1}$	0.02 $ml \cdot kg^{-1}$ 0.1 $ml \cdot kg^{-1} \cdot h^{-1}$ if diluted	Dilution: 18 ml + 32 ml G 5% = 1.8 $mg \cdot ml^{-1}$ Occasional paradoxical reaction
Morphine	10 $mg \cdot ml^{-1}$	0.1 $mg \cdot kg^{-1}$ 10 $\mu g \cdot kg^{-1} \cdot h^{-1}$ = 0.024 $ml \cdot kg^{-1} \cdot day^{-1}$	0.01 $ml \cdot kg^{-1}$ 0.1 $ml \cdot kg^{-1} \cdot h^{-1}$ if diluted	Dilution: 0.5 ml + 49.5 ml G 5% = 0.1 $mg \cdot ml^{-1}$ Sedation, algogenic administration, long EHL, nausea/vomiting, broncho-spasm, constipation
Naloxone	0.04 mg·2 ml^{-1}	0.01 $mg \cdot kg^{-1}$	0.5 $ml \cdot kg^{-1}$	Not for premature infants, relative short EHL (1 h)
Sodium hydrogen carbonate 8.4%	1 $mmol \cdot ml^{-1}$ Na^+	ml sodium hydrogen carbonate = BE×kg×0.3		
Sodium solutions	NaCl 0.9% NaCl 10% Sodium glycero-phosphate	0.154 $mol \cdot ml^{-1}$ 1.7 $mmol \cdot ml^{-1}$ 2.0 $mmol \cdot ml^{-1}$		
Neostigmine	0.5 $mg \cdot ml^{-1}$			Cholinergic
Nifedipine	20 $mg \cdot ml^{-1}$ (20 dr)		ca. 1 $dr \cdot kg^{-1}$	

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Nifedipine	5 mg/50 ml = 0.1 mg·ml^{-1} 5/10/20 mg·cps^{-1}	0.3–0.7 mg·kg^{-1} (0.5–6 mg·kg^{-1}·h^{-1}) Prf: 0.05 mg·kg^{-1}·h^{-1} = 1.2 mg·kg^{-1}·day^{-1}		Headaches Feel hot Shiver
Sodium nitroprusside	60 mg/3 ml = 20 mg·ml^{-1}	0.5–5 µg·kg^{-1}·min^{-1}	0.1 ml·kg^{-1}·h^{-1} if diluted	Dilution: 1.5 ml + 48.5 ml G 5% = 600 µg·ml^{-1}
Omeprazole	40 mg/10 ml 4 mg·ml^{-1}	0.25 mg·kg^{-1}	i.v. 0.25 ml·4 kg^{-1}	Proton pump inhibitors in ulcer, esophagitis, gastric acid ↓ Slow injection
Orciprenaline	0.5 mg·ml^{-1}	0.01 mg·kg^{-1} 0.1 µg·kg^{-1}·min^{-1} – 0.5 µg·kg^{-1}·min^{-1}	0.02 ml·kg^{-1} = 0.1 ml/5 kg 0.1 ml·kg^{-1}·h^{-1} if diluted	Dilution: 6 ml + 44 ml G 5% = 60 µg·ml^{-1}
Pethidine	50 mg·ml^{-1}	0.5–1 mg·kg^{-1}	0.01–0.02 ml·kg^{-1}	Vomiting/nausea, bronchospasm
Phenobarbital	200 mg·ml^{-1} Luminalete: 15 mg	10–20 mg·kg^{-1} Md: 5 mg·kg^{-1}·day^{-1}	0.1 ml·kg^{-1}	10 mg = 0.5 ml 1:10 diluted Level 20–80 mg·l^{-1}
Phenprocoumon	3 mg·tbl^{-1}	0.8 mg·kg^{-1} in 3 days (50/25/25%) Md: 0.08 mg·kg^{-1}·day^{-1}	ca. ¼–½ tbl·day^{-1} then ca.1–1½ tbl·day^{-1}	Prothrombin time 20%–30%

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Phenytoin	750 mg/50 ml = 15 $mg \cdot ml^{-1}$	15–20 $mg \cdot kg^{-1}$ Md: 5 $mg \cdot kg^{-1} \cdot day^{-1}$	i.v. 0.3–0.4 $ml \cdot kg^{-1} \cdot day^{-1}$ 2×	1:10 with G 5% diluted Level: 5–20 $mg \cdot l^{-1}$
Promethazine	1 mg/dr 25 $mg \cdot ml^{-1}$	1 $mg \cdot kg^{-1}$ Prf: 6 $mg \cdot kg^{-1} \cdot day^{-1}$	p.o.: 1 $dr \cdot kg^{-1}$ i.v.: 1 ml·25 kg^{-1}	Cardiotoxic, vomiting, nausea Addiction
Propofol	10 $mg \cdot ml^{-1}$	5–20 $\mu g \cdot kg^{-1} \cdot min^{-1}$	0.2 $ml \cdot kg^{-1}$ = 1 ml·5 kg^{-1} $0.6^{-1} \cdot 2$ $ml \cdot kg^{-1} \cdot h^{-1}$	Bronchospasm EHL 3 min Fever RR ↓, HF ↓
Ranitidine	10 $mg \cdot ml^{-1}$ 150, 300 $mg \cdot tbl^{-1}$	i.v. 1 $mg \cdot kg^{-1} \cdot day^{-1}$ p.o. 2 $mg \cdot kg^{-1} \cdot day^{-1}$	0.1 $ml \cdot kg^{-1} \cdot day^{-1}$ in 3 doses i.v.	
Salbutamol	5 $mg \cdot ml^{-1}$ = 0.2 $mg \cdot dr^{-1}$		1 $dr \cdot kg^{-1}$, max. 1 ml	Diluted in 2–5 ml NaCl 0.9% Inhalation
Sotalol	40 mg/4 ml = 10 $mg \cdot ml^{-1}$	20 mg in 5 min, max. 1.5 $mg \cdot kg^{-1}$		Repeat every 6 h
Spironolactone	25.50 $mg \cdot cps^{-1}$ 20 $mg \cdot ml^{-1}$	1–5 $mg \cdot kg^{-1} \cdot day^{-1}$	Initially 5 $mg \cdot kg^{-1} \cdot day^{-1}$ then 3 $mg \cdot kg^{-1} \cdot day^{-1}$	First effect within few days

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Succinylcholine chloride	1%: 10 mg·ml^{-1} 2%: 20 mg·ml^{-1} 3%: 50 mg·ml^{-1}	1–2 mg·kg^{-1}	0.1–0.2 ml·kg^{-1}	
Sucralfate	5 ml/bag	2 ml·kg^{-1}·day^{-1}	p.o. 4×	5 ml = 190 mg aluminum
Theophylline	1 mg·dr^{-1} 100/200/350 mg·cps^{-1} 0.24 mg·ml^{-1}	6 mg·kg^{-1} Md: 4–6 mg·kg^{-1}·day^{-1} Initially 4–7 mg·kg^{-1} then 15–20 mg·kg^{-1}·day^{-1}	2× 3 dr·kg^{-1} in 2 h Md: 4–6 × 1 dr·kg^{-1} per day 0.1 ml·kg^{-1}·h^{-1} if diluted	HR↑, Bg ↑ , diuresis ↑ Prf: 1 mg·kg^{-1}·h^{-1} Dilution: 26 ml + 24 ml G 5% = 10 mg·ml^{-1}
Thiopental	0.5 g/20 ml = 25 mg·ml^{-1}	2–5 mg·kg^{-1}	0.08–0.2 ml·kg^{-1}	
Tramadol	50 mg·ml^{-1} 50 mg·20 dr^{-1}	1 mg·kg^{-1} 0.2–0.4 mg·kg^{-1}·h^{-1}		Max. 1 mg·kg^{-1}·h^{-1}
Triflupromazine	10 mg·ml^{-1}	0.1 mg·kg^{-1}	i.v./i.m. 0.1 ml·10 kg^{-1}	Antiemetic Tranquilizing effect Anxiolytic
Urokinase	50,000 IU/2 ml = 25,000 IU·ml^{-1}	Initially 4000 IU·kg^{-1} then ≥1000 IU·kg^{-1}·h^{-1}	0.16 ml·kg^{-1}	In addition heparin 400 IU·kg^{-1}·day^{-1}

Table 45.1 *(continued)* Commonly used pharmaceuticals. (*amp* Ampoule, *Bc* blood count, *Bg* blood glucose, *BM* bone marrow, *BPD* bronchiopulmonary dysplasia, *CVA* central vein access, *cps* capsules, *dr* drop, *EHL* effective half-life, *G5%* glucose 5%, *GFR* glomerular filtration rate, *HR* heart rate, *i.v.* intravenous, *IU* international unit, *Md* maintenance dose, *Nb* newborn, *Pm* premature, *p.o.* per os, *Prf* perfusor, *Pw* pregnancy week, *Rect* rectal, *RR* Blood pressure, *s.c.* subcutaneous, *syr* syrup, *tbl* tablet, *Temp* body temperature, *U* urine)

Generic	Concentration	Dosage	Administration	Comment
Vecuronium	4 $mg \cdot ml^{-1}$	50–100 $\mu g \cdot kg^{-1} \cdot min^{-1}$ 1.6 $\mu g \cdot kg^{-1} \cdot min^{-1}$	0.1 ml/5 kg 0.1 $ml \cdot kg^{-1} \cdot h^{-1}$ if diluted	Dilution: 12 ml + 38ml G 5% = 0.96 $mg \cdot ml^{-1}$ Effect amplified by: $K^+ \downarrow$ $Ca^{2+} \uparrow$, $Mg^{2+} \uparrow$, pH↓, $CO_2 \uparrow$ Antagonist: prostigmin, Mestinon®
Verapamil	5 mg/2 ml = 2.5 $mg \cdot ml^{-1}$	0.3 $mg \cdot kg^{-1}$ in 5 ml G 5% then 1 $ml \cdot min^{-1}$	Slowly i.v.	ECG Stop if HR↓
Vitamin E	100 $mg \cdot 2\ ml^{-1}$ 10 $mg \cdot dr^{-1}$	20 $mg \cdot kg^{-1} \cdot week^{-1}$ 10–20 $mg \cdot kg^{-1} \cdot day^{-1}$	i.m. 0.4 $ml \cdot kg^{-1} \cdot week^{-1}$ 1–2 $dr \cdot kg^{-1} \cdot day^{-1}$	Premature newborns at 10th day until about 2200 g
Vitamin K1	1 $mg \cdot 0.5\ ml^{-1}$ = 0.2 $mg \cdot 0.1\ ml^{-1}$	0.1–0.2 mg	i.m./i.v. 0.05 ml	Perinatal for newborns and premature infants

Important note

Information from the manufacturers indicates that some of these medicines are not licensed for administration to children. This is because clinical trials are performed primarily on adults and separate trials for prescription in children must be carried out. As these studies are very expensive and the market for children is not very big, these studies are not performed for all pharmaceuticals.

Pharmaceuticals should only be prescribed to children if the benefit of doing so is greater than the risk of not prescribing them. Although this table has been prepared very meticulously, we advise readers to check the information leaflet produced by the manufacturer for the drug's indications, contraindications, and dosage.

46 Syndromes

Syndrome	Explanation
Abt-Letterer-Siwe	Malignant reticulosis with high fever, skin petechiae, bone lesions, enlarged glands, hepatosplenomegaly, and anemia
Adrenogenital (AGS)	Hereditary or acquired hyperadrenocortism
Albright	Fibrous dysplasia, pigmentation defect, pubertas praecox
Amazon	Unilateral syndactyly and brachydactyly, pectoral aplasia (Poland syndrome), and mammillary hypo- or aplasia
Apert	Acrocephalic syndactyly, turricephaly, small bowel atresia
Apple peel	Distal small bowel twisted along a central vessel
Bang disease	Brucellosis, undulant fever
Battered child	Every physical or psychological impairment of a child, which is induced deliberately or by negligence. Somatic abuse, malnourishment, sexual abuse, toxication, mental abuse
Beckwith–Wiedemann	See Wiedemann–Beckwith syndrome
Bland–White–Garland	The origins of the coronary artery origins are in the pulmonal trunk
Bonnevie–Ullrich	Malformation of the extremities, formation of pterygium, loss of the function of cerebral nerves, cerebral malformations, laced skin, disturbances of growth and ossification, earlap dysplasia, intelligence deficiency
Bornholm disease	Epidemic acute myalgia: pleurodynia, pseudo appendicitis, pseudo paresis, meningitis
Budd–Chiari	Clinical picture after occlusion of the liver veins
Calvé	Osteonecrotic planar vertebra: aseptic necrosis of the epiphyseal plate of a vertebral body
Calvé–Legg–Perthes	See Perthes syndromes

Syndrome	Explanation
Cantrell	Malformation pentalogy with defects of the thoracic wall, the abdominal wall, the pericardium, the diaphragm, and the heart
Caroli	Inborn segmental enhancement of the intrahepatic bile ducts with (sub-)icterus, hepatomegaly, eventually fibroangiomatosis of the bile ducts with primary cholangiolithiasis. Additional multiple kidney cysts
Cat scratch disease	Benign inoculation lymphoreticulosis
Chagas disease	Infection with Trypanosoma cruzi
Chatterbox	Mental retardation coupled with remarkable language skills, Cardiovascular diseases
Chilaiditi	Interposition of the colon between liver and diaphragm
Christmas tree	See Apple peel syndrome
Cocktail party	See Chatterbox syndrome
Collins	See Franceschetti syndrome
Conn	Hyperaldosteronism with adenoma of the suprarenal gland cortex
Crohn's disease	Regional enteritis
Crouzon	Craniofacial dysostosis
Cushing	Clinical picture of a glucocorticoid surplus
Down	Trisomy 21
Dysplasia linguofacial Grob	Scanty hairiness on the head with a forehead dome, epicanthus, broad nose bridge with flat tip and small nostrils, small median upper lip cleft, palate cleft, brachydactyly with clinodactyly of the small finger
Ebstein anomaly	Malformation of the tricuspid valve
Exomphalos -macroglossia-gigantism (EMG)	See Wiedemann–Beckwith syndrome
Franceschetti	Mandibulofacial dysostosis
Gardner	Combination of skin, bone tumors, and adenomatous colon polyps
Gaucher	Familiar sphingolipidosis with storage of cerebrosides in the reticulo-endothelial system
Golden	Follicular ileitis; terminal ileum ileitis in small children (5–6 years) with gastrointestinal bleeding

Syndrome	Explanation
Haglund–Sever	Calcaneal apophysitis: aseptic necrosis of the heel at the onset of the Achilles tendon
Hand–Schüller–Christian's disease	Cholesterol lipidosis (lipoid granulomatosis), exophthalmia, diabetes insipidus and cranial changes with irregular bone defects, stomatitis, splenomegaly, hepatomegaly
Hashimoto thyroiditis	Immune thyroiditis with progressive destruction of the thyroid parenchyma and diffuse goiter with hormonal dysfunction
Hemolytic-uremic	Acute hemolytic anemia with simultaneous acute renal failure
Henoch–Schoenlein	See Schoenlein–Henoch syndrome
Hippel–Lindau	Angiomatosis of the cerebellum and the retina, commonly combined with multiple angiomas
Hirschsprung's disease	Congenital megacolon
Horner	Narrow palpebral fissure, miosis, enophthalmos
Hutchinson	See Horner syndrome
Hyaline membrane disease	See Respiratory distress syndrome
Inspissated bile	Blockade of the bile ducts with bile thrombus after severe hemolysis, especially in fetal erythroblastosis
Kasabach–Merritt	Inborn large cavernous hemangiomas with thrombosis and consumptive coagulopathy
Kienböck	Malacia of the lunatum bone
Klippel–Feil	Familial inherited developmental error with short neck, low hairline at the neck and restricted mobility of the upper spine. Associated abnormalities of the head, face, skeleton, sex organs, muscles, brain and spinal cord are possible
Klippel–Trenaunay	Partial angiectatic gigantism
Köhler	I: Aseptic bone necrosis of the navicular bone II: Aseptic necrosis of the metatarsal heads
Laurence–Moon–Biedl	Diencephalon-retinal degeneration: symmetric obesity, retinitis pigmentosa, developmental delay, hypogonadism, poly- and syndactyly, renal anomalies
Letterer–Siwe	See Abt–Letterer–Siwe syndrome
Louis–Bar	Ataxia telangiectasia. Disease starts at about 2 years of age with increased clumsiness, teleangiectatic red spider veins in the conjunctiva and face, hypersalivation, hearing loss, scoliosis

Syndrome	Explanation
Maffucci	Dyschondroplasia hemangiomatosa
Maladie des griffes du chat	See Cat scratch disease
Mayer–Rokitansky–Küster–Hauser	Congenital absence of the vagina, rudimentary cornua uteri, amenorrhea
Meckel–Gruber	Polycystic kidney degeneration
Meconium plug	Clinical picture of the obstruction of the colon by a ileocolic or anorectal meconium plug
Melkersson–Rosenthal	Recurring facial paralysis, swelling of the upper lip, development of folds and furrows in the tongue
Membrane	See Respiratory distress syndrome
Mendelson	Bronchiopulmonary reaction following aspiration of gastric contents
Ménétrier	Gastropathia hypertrophica gigantea; enlargement of the gastric mucosa folds with massive secretion of mucus resulting in low plasma protein levels with peripheral edema and ascites
Mesenteric artery	Gastrointestinal pain caused by pressure of the isthmus of a horseshoe kidney on the inferior mesenteric artery
Milroy	See Nonne–Milroy–Meige syndrome
Niemann–Pick's disease	Sphingolipidosis with sphingolipid accumulation in cells especially in reticuloendothelial cells throughout the body
Nonne–Milroy–Meige	Familial, chronic, pain-free lymphedema of the limbs compared with acromicria, obesity, hypogenitalism, retardation, often associated with various ulcerations and secondary infections
Noonan	Pseudo-Turner syndrome
Oro–facial–digital	See Dysplasia linquofacialis Grob
Osgood–Schlatter	Uni- or bilateral aseptic necrosis of the apophysis of the tibial tuberosity
Panner disease	Necrosis of the epiphysis of the humerus capitulum
Papillon–Léage	See Dysplasia linguofacialis Grob
Parkes–Weber	Hemangiopathy with arteriovenous anastomoses
Perthes	Aseptic necrosis of the hip joint
Peutz–Jeghers	Intestinal hamartomatous polyps associated with mucocutaneous melanocytic macules

Syndrome	Explanation
Pierre–Robin	Swallowing and breathing problems due to micrognathia and retrognathia sometimes combined with palate cleft
Poland	Unilateral syndactyly and brachydactyly and aplasia of the major pectoral muscle
Potters	The result of oligohydramnios secondary to renal diseases such as bilateral renal agenesis, obstructive uropathy, autosomal recessive polycystic kidney disease, and renal hypoplasia
Proteus	Lipomatosis invading the muscles, development of hamartomas, acromegaly, hemihypertrophy of extremities
Prune belly	Dysplasia of the abdominal wall, with genitourinary malformations, often associated with skeletal malformations as well as intestinal anomalies
Pseudo-Hirschsprung's disease	Hypoganglionosis with low number of ganglia cells in the submucosal myenteric plexus as well as reduced density of the nerve fibers although acetylcholinesterase activity is normal
Recklinghausen	Neurofibromatosis
Reifenstein	Peripheral androgen resistance: phenotypically males with abnormal male genitals (cryptorchidism with hypotrophic testicle, perineoscrotal hypospadias, occasionally with a rudimentary vaginal sac, azoospermia and gynecomastia)
Respiratory distress	Cyanosis, tachypnea, apnea, edemas, acidosis, oliguria
Rossi	Congenital pterygoarthromyodysplasia: arthrogryposis with formation of pterygia on the joints affected
Roviralta	Hypertrophic pyloric stenosis in combination with a hiatus hernia and gastric ectopia
Salt-losing nephritis	Form of the AGS with blocked aldosterone synthesis
Sandifer	Spasmodic torsional dystonia associated with gastroesophageal reflux and/or hiatus hernia
Short bowel	Malabsorption, and other complications after excessive small bowel resection
Scheuermann	Osteochondritis deformans juvenilis dorsi, kyphosis in adolescent children
Schlatter	See Osgood–Schlatter syndrome

Syndrome	Explanation
Schoenlein–Henoch	Systemic vasculitis with skin purpura, arthralgia, abdominal pain, eventually bowel bleeding and glomerulonephritis
Scimitar	Right-sided lung hypoplasia with a sequester with individual arterial vascularization and abnormal venous drainage in the inferior cava vein
Silverman	See Battered child syndrome
Sinding–Larsen–Johansson	Aseptic necrosis of the distal patella tip
Sprengel deformity	Congenital high scapula
Sturge–Weber–Krabbe	Neuroangiomatosis encephalofacialis: congenital neuroectodermal dysplasia with hemangiomas in the face, meninges and chorioidea
Tardieu	See Battered child syndrome
Treacher–Collins	See Franceschetti syndrome
Triad	See Prune belly
Turner	Monosomy X: gonadal dysgenesis with female phenotype due to chromosome aberrations
Upper calyx	Isolated hydronephrotic increments of the upper calyx groups of the kidney due to inborn or acquired obstruction
Urethra	Pollakisuria and dysuria of unclear etiology
VACTERL	Additional malformations in esophageal atresia: vertebral, anal, cardiac, tracheal, esophageal, radial, limbs
VATER association	Additional malformations in esophageal atresia: vertebral, anal, tracheal, esophageal, renal
Whipple	Intestinal lipodystrophy
Wiedemann–Beckwith	Omphalocele, muscular macroglossia, microcephaly, visceromegaly
Wilson	Hepatolenticular degeneration with copper deposits in organs
Wiskott–Aldrich	Immune defect with thrombocytopenic bleeding, dermatitis with high infection potential
Wolff–Parkinson–White	Heart function disorder with tendency to paroxysmal tachycardia
Zuelzer–Wilson disease	Total aganglionosis of the intestine in congenital megacolon

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