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Pediatric Surgery and Orthopedics

Course Manual

MANUALE

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PEDIATRIC SURGERY

Chapter 1. CLEFT LIP AND PALATE

Definition. Orofacial clefts are a group of congenital fusion anomalies that include cleft lip (cheiloschisis), cleft palate (palatoschisis), or a combination of both cleft lip, alveolus and palate (cheilognathopalatoschisis). These anomalies may exhibit various clinical forms and combinations.

The **incidence** is approximately 1.7 per 1,000 newborns. Cleft lip and cleft palate affect males more frequently, while cleft palate affects females more frequently.

Etiopathogenesis of the condition is multifactorial (genetic and environmental factors). Genetic factors play the main (major) role, these anomalies occur isolated (either in isolation) or associated (in association) with other types of malformations in more than 60 known syndromes (Pierre-Robin, Patau, Optiz, Klippel-Feil, Edwards, Van der Woude, etc.). If one parent and the first child have a cleft, the probability that the second child will also have the anomaly is 17-20%. Environmental factors implicated in the occurrence of cleft lip and palate malformations are medications (anticonvulsants, paracetamol, thalidomide), alcohol abuse and smoking during pregnancy, vitamin deficiencies (zinc, folic acid), infections (rubella), oligohydramnios, and radiation. Environmental factors act as triggers for the development of anomalies in a genetically susceptible background.

Embryology. From the fourth week of embryonic life, five facial prominences (frontonasal, paired maxillary, paired mandibular prominences) appear at the cephalic end, contributing to the development of the face. These

are arranged around a superficial ectodermal depression called the stomodeum and are composed internally of mesenchymal tissue - to which cells detached from the neural crests migrate - and externally delimited by the ectoderm. Starting in the fifth week of intrauterine life, the frontal prominence descends vertically between the upper maxillary prominences and divides into four parts (Figure 1.1).

The union of these processes occurs around week 8 through the process of mesodermisation. Failure of fusion between the frontonasal bud (medial nasal process) and the maxilla causes cleft lip and premaxilla. Next, the secondary palate is formed, consisting of the hard and soft palate, which is located posterior to the incisive foramen. It is formed by the growth and fusion of two palatal processes at the level of the maxillary buds. The palatal processes or plates initially have a vertical orientation, subsequently arranging themselves transversely and fusing from anterior to posterior. The union of these processes occurs around week 8 through mesodermisation. Failure of this process leads to the appearance of palatal clefts.

Classification. The phenotypic manifestation of clefts is variable, and thus a multitude of classification systems have been developed. Depending on the anatomical, embryological, and pathophysiological characteristics, these malformations are classified according to a key anatomical landmark - the incisive foramen.

With a different origin than the posterior structures, the structures that develop anterior to the incisive foramen form first and eventually give rise to the nose, lip, and dental alveolus. Embryologically, they will be classified as the primary palate. The structures that develop posterior to the incisive foramen will be defined as the secondary palate—the hard and soft palate. In practice, we may encounter complete/incomplete, unilateral or bilateral clefts.

Diagnostic. Prenatal diagnostic is possible through fetal ultrasound with a detection rate of up to 80%, but also with false positive results (Fig. 1.2). Although it does not benefit the affected newborn, it greatly benefits the parents in terms of their psychological preparation for the future child.

Postnatal diagnostic is clinical. The clinical presentation of clefts varies, with the degree of dysmorphism and dysfunction depending on the severity of the malformation. The positive diagnostic of the condition is primarily clinical, evident at birth in most cases, but there may also be mild forms that go unnoticed, represented as an indentation in the red of the lip or a submucosal palatal cleft. The least severe forms are incomplete, simple unilateral clefts. (Figure 1.3a). Cleft lip is often associated with cleft alveolus and palate (Figure 1.3b).

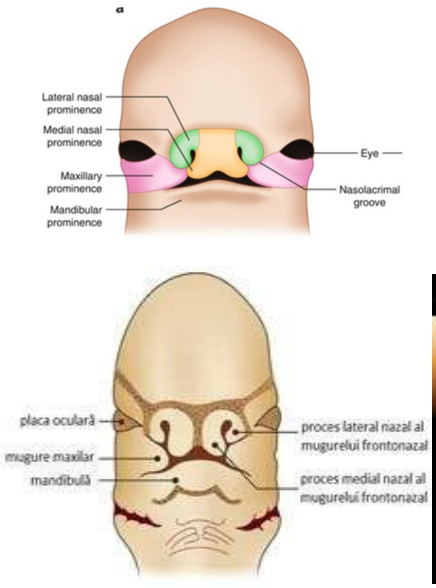


Fig. 1.1. Development of frontonasal and maxillary buds in week 8

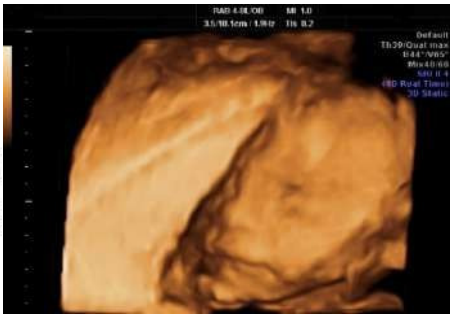


Fig. 1.2. Unilateral cleft lip, intrauterine ultrasound

Bilateral cleft lip is the most severe form of lip malformation. Cleft palate may accompany cleft lip or may be isolated. The most severe and disfiguring malformation is complete bilateral cleft, affecting the nose, upper lip, alveolus (primary palate), secondary palate (Figure 1.3c).

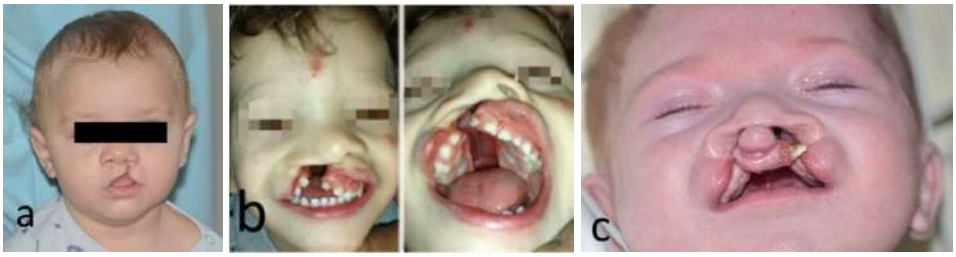


Fig. 1.3. a. Simple unilateral cleft lip; b. Complete unilateral cleft lip associated with complete cleft alveolus and palate; c. Bilateral cleft lip associated with complete cleft palate

Treatment. The disorders caused by these conditions vary depending on the severity of the malformation but generally include feeding difficulties with secondary growth and weight deficits, ENT infections, impaired speech development, and as well as affective, psychological, and psychosocial disorders. Immediately after birth, these patients struggle with respiratory problems caused by poor drainage of secretions. Routine aspiration of the newborn's secretions is usually sufficient to clear the airways. Breastfeeding is usually possible. Sometimes, adjuvant measures such as special nipples (the pigeon cleft palate feeding bottle) or palatal plates are required.

The management of cleft lip and palate involves multiple surgical interventions and a multidisciplinary approach (oral and maxillofacial surgery, pediatric surgery, ENT, plastic surgery, dentistry, orthodontics, speech therapy). The appropriate time to begin surgical correction of the anomaly follows the "rule of 10": 10 weeks of life, 10 g/dl of hemoglobin, and 10 pounds (approximately 4.5 kg) of weight. The sequence of

interventions begins with lip and alveolar process repair, around the age of 3-4 months. This is followed by correction of the cleft palate at 6-9 months of age. At 7 and 9 years of age, permanent tooth eruption is facilitated by an adjuvant alveolar bone grafting procedure at the site of the existing defect. Finally, at skeletal maturity (17 years in girls and 19 years in boys), maxillary/mandibular osteotomies can be performed to compensate for the maxillomandibular skeletal deficit and other corrections. The cleft rhinoplasty is performed at different stages of growth (primar, intermediate and secondary) of pacient to correct nasal deformity associated with cleft lip and palate.

Principles of postoperative care. Feeding is individualized through gavage, breastfeeding, or bottle feeding, depending on the surgical technique used, type of incision, and local conditions. Medication consists of administering analgesics, anti-inflammatories, and antibiotics. For optimal healing, it is very important to properly clean the postoperative wound daily and protect it from trauma. Frequent washing and aspirating secretions throughout the day are necessary to maintain a clean local environment. The skin sutures are removed 7- 10 days after surgery. The sutures in the oral cavity do not require removal, as they are absorbable.

Cleft lip and palate constitute a significant aesthetic and functional handicap for the child. Repairing these defects requires a multidisciplinary, coordinated effort and long-term collaboration between the healthcare team and patient.

Chapter 2. THYROGLOSSAL DUCT CYST

Definition. It is a cystic tumor located anteriorly on the midline of the neck, resulting from the persistence of the thyroglossal duct - an embryonic structure that connects the pyramidal lobe of the thyroid gland to the foramen cecum.

Embryology. The thyroid gland develops between weeks 4-7 of intrauterine life and descends from the base of the tongue through a canal called the thyroglossal duct. Once the descent of the thyroid is complete (weeks 5-8), the thyroglossal duct disappears. Persistence of this duct leads to the formation of a thyroglossal duct cyst. Concurrently with the formation of the thyroid, the hyoid bone (derived from the second branchial arch) is formed, allowing the thyroglossal duct to pass either anteriorly, or posteriorly to the hyoid bone. The duct may be partially obliterated, so its remannats can be located anywhere along the migratory path of the thyroglossal tract (most commonly beneath the hyoid bone). It is lined with a cylindrical epithelium, sometimes ciliated, presenting a mucoid secretion.

Incidence. It is the most common cervical cystic formation and occurs in 7% of the population. It occurs in both sexes after the age of 2-3 years. In 2/3 of cases, it is discovered in the first 3 decades of life. In 1% of cases, it is associated with malignancy, most commonly papillary carcinoma.

Diagnostic. Symptoms are most commonly observed in preschool-aged children. Clinically, a round tumor is detected on the midline, anterior to the neck, which is elastic, painless, and mobile. The pathognomonic sign is the upward movement of the cyst during tongue protrusion and swallowing, doe to its connection to the base of the tonguevia a fistulous tract. Sometimes, the cyst becomes inflamed, painful, and may fistulate secondarily to the skin (Figure 2.1).

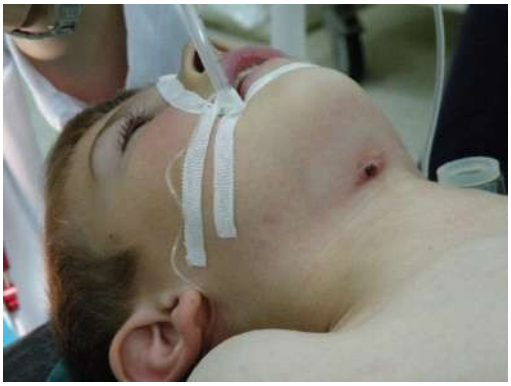


Fig. 2.1. Fistulated thyroglossal duct cyst

Paraclinical picture. If clinical and anamnestic signs of thyroid hypofunction (chronic constipation, delayed growth and development, excessive sleepiness) are present, additional paraclinical investigations are performed, such as ultrasound, thyroid function assessment, radioactive iodine scan, CT, MRI.

The **differential diagnostic** include ectopic thyroid nodule, dermoid cyst, cervical adenopathy.

Treatment is surgical and consists of excision of the cyst and the entire fistulous tract to the base of the tongue, including the central portion of the hyoid bone (Sistrunk procedure). The intervention is not recommended during an inflammatory flare-up of the cyst, as the risk of recurrence is higher. The infection is treated first, and surgical excision is performed later, once the infection has subsided.

Prognosis. If left untreated, the thyroglossal duct cyst can be serve as a precursor to papillary thyroid carcinoma. Failure to excise the central portion of the hyoid bone leads to recurrence in 70% of cases. Complete surgical excision, performed correctly, ensures a good cure without sequelae.

Chapter 3. BRANCHIAL ARCHES CYSTS AND FISTULAS

Definition. Congenital fistulas and cysts of the neck are remnants of embryonic branchial arches that have been incompletely resorbed.

Embryology. During weeks 4-8 of gestation, six pairs of branchial arches develop at the cranial end of the embryo. These arise from a thickening of the mesoderm, are externally covered by the ectoderm and internally by the endoderm, and each contains an artery, a vein, cartilage, and a muscle. The last two pairs of gill arches involute, as they are not important for the further development of the embryo. The first four pairs of branchial arches develop into structures of the cephalic extremities and neck. Normally, the sinuses (or clefts) separating these branchial arches gradually close; failure to do so results in the formation of branchial sinuses or cysts.

Pathological anatomy. Persistence of the first groove results in the presence of a fistula that opens either posteriorly or inferiorly to the ear. Cysts and fistulas derived from the first branchial arch are rare and are located near the external auditory canal, the facial nerve, and the parotid gland.

Fistulas and cysts derived from the second branchial groove are most common and account for 95% of all branchial anomalies. They can be located unilaterally or bilaterally, with the fistulous tract starting at the pharynx, passing either adjacent to or through the carotid bifurcation, and opening on the anterior edge of the sternocleidomastoid muscle.

Arches III and IV fistulas are extremely rare. Arch III fistula starts at the pharynx and follows a posterior course relative to the carotid artery. Arch IV fistula descends to the level of a thyroid lobe.

Fistulization to the skin and externalization of the contents occur secondary to infection, with all cystic lesions being initially located beneath the skin.

Diagnostic. Clinically, a cyst or fistula through which purulent fluid is externalized is evident. Cysts appear as tumor formations of varying sizes, with a smooth surface, elastic consistency, mobile, painless, located in the subcutaneous tissue. Fistulization occurs secondarily, and clear or milky drops of fluid are intermittently eliminated through the external opening of the fistula. The evolution is cyclical: after an inflammatory flare-up of the lesion, fistulization occurs with the elimination of purulent content, followed by the extinction of the inflammatory focus. The process repeats at variable intervals.

The lesions derived from the first branchial arch are located inferiorly or medially to the auricle or beneath the angle of the mandible. Cysts or fistulas of the second branchial arch are usually located on the anterior edge of the sternocleidomastoid muscle.

Diagnostic is usually based on the clinical picture, but soft tissue ultrasound or fistulography may be performed for confirmation.

Treatment is surgical and should be performed in the absence of inflammation. The entire fistulous tract up to the pharynx is excised. To minimize aesthetic concerns, staggered transverse incisions are performed. Due to the proximity of important vascular and nervous structures, the dissection should be performed as close to the fistula as possible.

Chapter 4. VASCULAR ANOMALIES

In the past, the word "hemangioma" was misused and incorrectly used to describe reddish-purple lesions on the skin. Currently, there is a clear distinction between vascular tumors (hemangiomas) and vascular malformations.

Definition. Hemangiomas are benign vascular tumors characterized by initial cell proliferation with subsequent involution. Vascular malformations are congenital lesions that occur due to errors in embryonic development and have normal cell turnover. Vascular malformations do not proliferate; they grow as the patient grows.

Classification. The most used nowadays is the ISSVA (International Society for the Study of Vascular Anomalies) classification, revised in 2018. It divides vascular anomalies into vascular tumors and vascular malformations (Table 4.1).

4.1. VASCULAR TUMORS

Vascular tumors are common in the pediatric population, being diagnosed in 10% of cases in the first year of life. Of these, infantile hemangioma is the most common.

INFANTILE HEMANGIOMA

Infantile hemangioma is the most common tumor found in children, with majority located on the head and neck. They are characterized by rapid initial growth of endothelial cells followed by slow involution.

Epidemiology. The incidence of these tumors in the first 3 days of life is between 1.1% and 2.6% and increases to 8.7% to 12.7% between 1

month and 1 year of age. Caucasian individuals are more commonly affected, with an incidence of 1% in the black race. It occurs more frequently in girls, with a female: male ratio of 2.4:1. The incidence increases with prematurity.

Table 4.1. Vascular anomalies – 2018 ISSVA classification

Vascular tumors	**Vascular malformations**
1. Infantile hemangioma 2. Congenital hemangioma (RICH, PICH, NICH) 3. Tufted angioma (with or without Kasabach Merritt syndrome) 4. Kaposiform hemangioendothelioma (with or without Kasabach Merritt syndrome) 5. Spindle cell hemangioendothelioma 6. Other rare hemangioendotheliomas (epithelioid, mixed, retiform, polymorphic, Dabska tumor, lymphangioendotheliomatosis) 7. Acquired dermatological vascular tumors (pyogenic granuloma, targetoid hemangioma, glomeruloid hemangioma, microvenular hemangioma) 8. Malignant tumors—angiosarcoma	I. Low-flow vascular malformations 1. Capillary malformations • Salmon patch • In "wine stain" • Telangiectasia • Angiokeratoma 2. Venous malformations 3. Lymphatic malformations II. High flow vascular malformations • Arterial malformation • Arteriovenous malformation III. Complex vascular malformations • Capillary-venous malformations • Capillary-lymphatic malformations • Venous-lymphatic malformations • Venous-lymphatic-capillary malformations • Arterio-venouslymphatic malformations • Arterio-venous capillary malformations • Syndromes: Klippel- Trenaunay, Parkes-Weber, Servelle-Martorell, Cutis marmorata telangiectatica congenita, Adams-Oliver, Ataxia Telangiectasia, etc.

Etiopathogenesis. The placental trophoblastic origin has been suggested by some authors who have shown that the incidence increased to 21% among children born from pregnancies in which chorionic villus sampling was performed.

Clinical picture. The clinical appearance varies depending on the depth of skin involvement; it can present as a superficial, bright red lesion resembling a "strawberry" located on the skin and affects the superficial dermis (Figure 3.1a), while others are located deep in the hypodermis and present as a bluish arch shape (Figure 3.1b). The combination of a superficial component with a subcutaneous one results in a mixed infantile hemangioma (Figure 3.1c). Thus, infantile hemangiomas are classified into three types: superficial, deep, and mixed. Infantile hemangioma has a particular evolutionary history: it is present from the first month of life, grows until the 10th-12th month when it reaches a phase called plateau, which will last several years, then regresses spontaneously, disappearing completely before the age of 7.

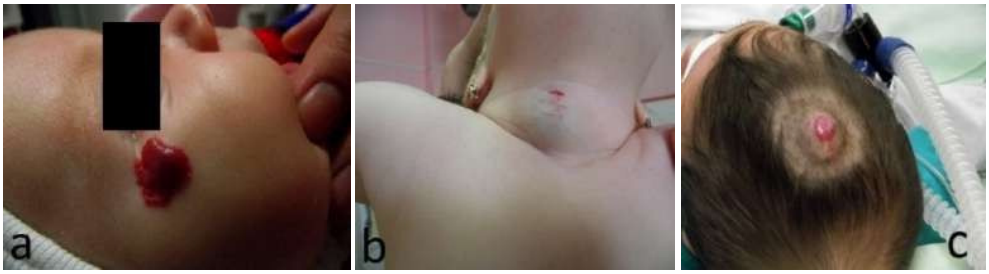


Fig. 3.1. Infantile hemangioma: **a.** Superficial **b.** Deep **c.** Mixed

Diagnostic. The diagnostic is usually clinical, based on simple inspection. Depending on the location, imaging investigations (ultrasound, CT, MRI) may be necessary.

Differential diagnostic include both vascular malformations and vascular tumors (congenital hemangioma) and soft tissue tumors (myofibromatosis, lipoblastoma, fibrosarcoma, rhabdomyosarcoma).

Prognostic and complications. Although most infantile hemangiomas disappear completely without sequelae, several potential

complications may occur, most commonly during the proliferative phase. Complications include hemorrhage, ulceration, infections, obstruction (visual axis, auditory canal, airways), congestive heart failure, skeletal deformity, and cosmetic impairment.

Treatment of vascular tumors depends on the type of tumor, clinical evolution, location, size of the lesion, and age of the patient. In the last decade, the first line of treatment for infantile hemangioma has been β-blockers, mainly Propranolol. Other systemic treatments include corticosteroids, interferon alpha, bleomycin, cyclophosphamide, thalidomide, and vincristine, but these have multiple and serious side effects compared to β-blockers. Locally, cryotherapy, sclerotherapy (injection with corticosteroids, bleomycin), and LASER can be used. Local topical corticosteroids or β-blockers can also be used. If local or systemic drug treatment is ineffective or contraindicated, surgery intervention is employed.

4.2. VASCULAR MALFORMATIONS

Vascular malformations are structural abnormalities resulting from defective vascular or lymphatic morphogenesis. In contrast to hemangiomas, these lesions generally have normal levels of endothelial cells and are present at birth. Vascular malformations can be divided into groups based on their vascular component and blood flow characteristics. They can be capillary, venous, low-flow lymphatic, high-flow arterial, or a combination thereof. They can be located anywhere but are commonly found in soft tissues. They are generally present at birth, persist throughout life, and in most cases worsen with time.

I. Slow-flow malformations

Capillary malformations. These lesions affect the capillary network of the skin and mucous membranes, and may occasionally extend into deeper structures, especially in the facial area. Capillary malformations, telangiectasias, and "wine stains" (flat angioma) represent the most common types of vascular malformations. They are present at birth, usually appear sporadically, although familial cases have also been described.

- "Salmon patches" are present in approximately 44% of all newborns, are brick-red or pink in color, or completely white, flat, and darken during intense activity such as crying, defecation or or in response to changes in ambient temperature. They are most commonly located on the back of the head, glabella, and eyelids, within the nasolabial folds, on the lips, and in the sacral area. They are symmetrical, with lesions on both eyelids or on one side of the midline (Figure 4.2a). They tend to regress significantly over time, with those on the eyelids and glabella usually disappearing by the age of 2-3 years, and those on the back of the neck and sacral area tending to persist longer, up to the age of 6 years.
- "Port-wine stains" appear in approximately 0.3% of all newborns, are present at birth as red macules, become purple with age, and do not disappear. They can appear anywhere on the body but are commonly found on the face or a limb. "Wine stains" can be markers of certain syndromes (Klippel-Trenaunay, Sturge-Weber). (Figure 4.2 b, c)

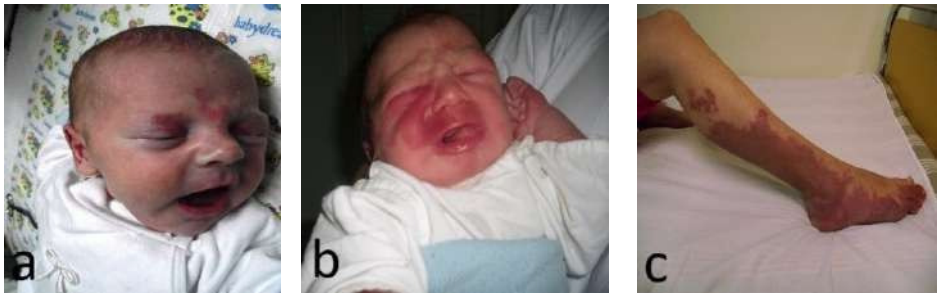


Figure 4.2. Capillary malformations: **a.** "Salmon patch." **b** and **c**. "Port-wine stain."

Venous malformations. Most are visible at birth as bluish spots or patches and progressively enlarge from childhood through puberty. Clinically, they present as soft, compressible blue masses that increases in size when the area is dependent and subjected to increase venous pressure.

The blue color is pathognomonic. There is no increase in local skin temperature or thrill on palpation. Venous malformations may occure throughout the body and can present as either localized lesions or extensive, diffuse malformations. The most common locations are the scalp and neck. These lesions may be superficial (intradermal or subcutaneous) or deep (intramuscular or intraosseous). Most venous malformation are asymptomatic; however, pain may occur in lesions located in the extremities. Phlebitis and hemorrhagic diathesis are complications of venous malformations. Treatment depends on the location and extent of the lesion. If the lesion is localized and accessible, surgical excision is performed, with good results. For extensive lesions that are not amenable to surgical excision, sclerotherapy or embolization is performed. Patients with extensive lesions in the limbs will be instructed to wear compression garments.

Lymphatic malformations. These are benign vascular lesions that occur due to a developmental disorder of the lymphatic system during embryogenesis. They can be superficial, focal, or diffuse, subcutaneous, deep,

but can also occur in muscles or organs. Lymphatic malformations can occur anywhere in the body, are present at birth, but sometimes become visible during puberty.

There are two types of lymphatic malformations: microcystic and macrocystic. Microcystic malformations appear as small, clear vesicles that penetrate the subcutaneous tissue and muscles. Macrocystic malformations are large, compressible or non-compressible, smooth, translucent under normal skin or bluish in color.

Possible complications include infection, intralesional bleeding, overgrowing of the affected limb, excessive bone growth, malocclusion, airway obstruction, visual impairment, chylothorax, chylopericardium, chylous ascites, and disfigurement.

Once the diagnostic has been established, treatment depends on the clinical appearance, size of the lesion, anatomical location, and complications present. Complete excision of the lesion is the preferred treatment, where possible. LASER is used in certain cases. An alternative to surgical treatment is sclerotherapy, which is effective in cases of macrocystic malformations. A variety of sclerosing agents have been used such as ethanol, bleomycin, doxycycline, acetic acid, Ethibloc, and OK-432 (Picibanil). Currently, various types of drug therapies (Sirolimus) and individualized molecular therapies are used.

II. Rapid-flow vascular malformations

Arteriovenous malformations belong to the category of high-flow lesions, consisting of dysmorphic arterial and venous vessels directly connected to each other, without a capillary bed. They are equally distributed between the sexes. Approximately 40-60% are visible at birth and progress

with age. Treatment is difficult. For a long time, surgical resection was considered the gold standard of treatment, but complications and recurrence rates were high. Complete eradication of the nidus is necessary for the intervention to be curative, but this is often not possible. Endovascular therapy is beneficial in patients with increased surgical risk and in cases of lesions located in inaccessible areas. In extremely rare cases, amputation may be necessary for lesions located in the extremities (lower and upper limbs). To resolve cases as accurately as possible and with the best results, a multidisciplinary team is required, consisting of vascular surgeon, pediatric surgeon, plastic surgeon, maxillofacial surgeon, anesthesiologist, pathologist, pediatrician, radiologist, interventional radiologist, dermatologist, neurologist, psychiatrist, and nuclear medicine physician.

Chapter 5. CHEST DEFORMITIES: PECTUS EXCAVATUM, PECTUS CARINATUM

5.1. PECTUS EXCAVATUM

Pectus Excavatum (PE) (sunken chest) is a deformity of the anterior chest wall consisting of posterior curvature of the sternum and costal cartilages (Fig. 5.1).

Incidence. It is the most common deformity of the chest wall, accounting for 90% of cases. The incidence is approximately 1 in 1000. It is more common in boys, with a ratio of 3:1.

Etiopathogenesis. The exact cause of Pectus Excavatum is not fully understood. Most studies have shown that an abnormality in the growth of the costal cartilage leads to deformation of the anterior wall of the chest and sternum; resulting anteriorly in Pectus carinatum or, more commonly, posteriorly in Pectus excavatum. The costal cartilage anomaly is most likely genetically determined, with the condition being familial in approximately 45% of cases.

Associated conditions. Pectus excavatum is frequently associated with scoliosis (15-20%), cardiac malformations (1.5%), asthma (5.2%), Ehlers-Danlos syndrome (2%), and Marfan syndrome (3%). Almost one-third of patients with Marfan syndrome have a sunken chest.

Pathological anatomy. Usually, the sternal depression is in the lower part of the anterior chest, with the maximum point of depression at the xiphoid process and has relatively limited extent (Fig. 5.1). The general appearance and posture of children with PE are characteristic. Patients are usually long-limbed with a narrow chest, marked kyphosis, and frequent scoliosis. The abdomen is often protruding in contrast to the patient's general asthenic appearance. Consequently, the deformation of the anterior chest wall reduces the sagittal diameter of the chest cavity, and the heart is most often deviated

to the left and is in contact with the inner surface of the sternum. The lungs are affected by the reduction in the capacity of the rib cage , resulting in a corresponding reduction in vital capacity. The deformation may be visible at birth or during the first year of life, but it is mild or moderate and remains so until puberty. At puberty, when the child exhibits rapid skeletal growth, the deformity progress significantly over a period of 1-2 years. After puberty, when the growing stops, the deformity tends to stabilize.

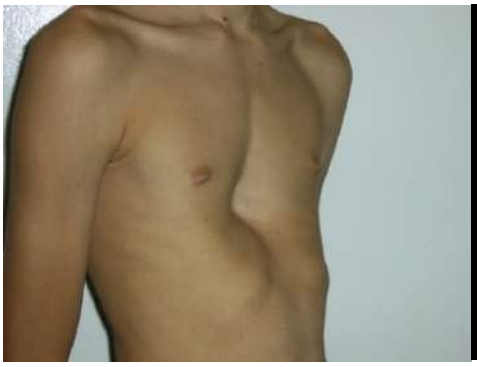


Fig. 5.1. Pectus excavatum

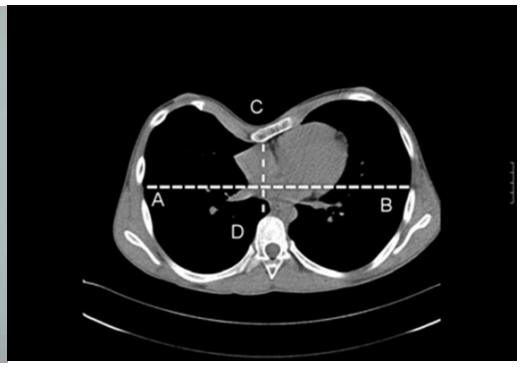


Fig. 5.2. CT, Haller index calculation

Clinical picture. In early childhood, patients are asymptomatic. The most common symptom is a progressive decline in exercise tolerance, primarily due to impaired cardiac function. Other less common signs and symptoms include pain in the deformed cartilage, palpitations, systolic heart murmurs, and frequent respiratory tract infections. At the same time, deformity has a major psychological impact on most patients.

Functional assessments. Spirometry and plethysmography can demonstrate a decrease in vital capacity (VC), forced vital capacity (FVC), forced expiratory volume in one second (FEV1), and forced expiratory flow (FEF25-75%). Cardiac function is affected by decreased stroke volume caused by direct effect of sternum compression, as well as mitral valve prolapse, and cardiac arrhythmias.

Paraclinical picture. Laboratory tests are unchanged, and chest X-ray does not provide useful information. Chest CT (Figure 5.2) is the imaging modality of choice and can assess the severity of the deformity by calculating the Haller index (transverse diameter of the chest divided by the sagittal diameter), normal: 1.75 - 2.6, pathological > 3.1.

Treatment. Patients with minor, asymptomatic deformity do not require treatment. Physical therapy is useful for correcting posture and spinal deformities. Deformity correction can only be achieved by surgical methods. The gold standard treatment is minimally invasive Nuss sternochondroplasty (Figure 5.3). The optimal age for surgical treatment is puberty. The procedure involves inserting a convex metal bar into the chest under thoracoscopic control. The bar passes transversely from the right pleural cavity to the left, anterior to the heart and behind the sternum. The bar will push the sternum anteriorly, correcting the deformity. It is kept in the chest for a period of 2 to 4 years.

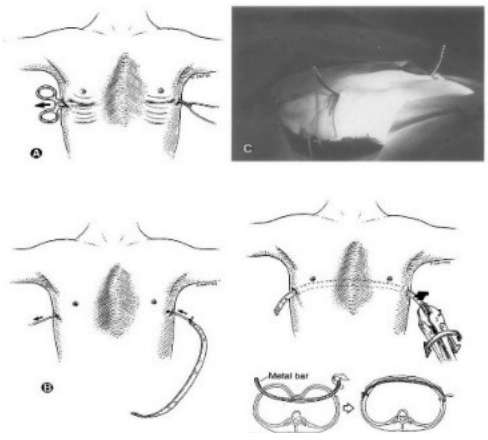


Fig. 5.3. Minimally invasive procedure Nuss (Nuss et al. 1998)

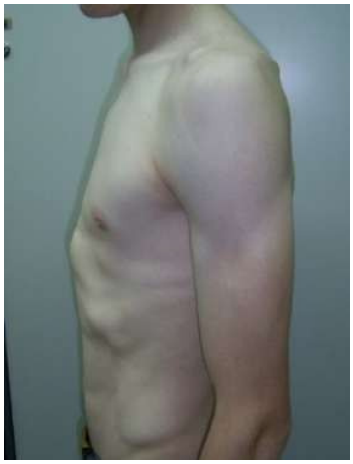


Fig. 5.4. Pectus carinatum

5.2. PECTUS CARINATUM

In Pectus Carinatum, the anteriorl chest wall is deformed, with the sternum and part of the costal cartilages protruding (Figure 5.4). It accounts for approximately 5% of congenital chest wall malformations, with 80% of cases occurring in boys. As with Pectus Excavatum, the etiology of the condition has not been fully clarified, but its origin most likely being similar to sunken chest. Clinical manifestations are less common than in the sunken chest, most complaints primarily related to aesthetic concern. Symptoms such as decreased exercise capacity are explained by lower chest wall compliance or the presence of concomitant cardiac conditions such as mitral valve prolapse and arrhythmias. As the consequences are cosmetic for most patients, treatment is less frequently indicated. For most patients, physical therapy is sufficient to correct poor posture. Corset-type devices can be used. Surgical treatment is exceptional, with the use of classic open surgical techniques or minimally invasive treatment options.

Chapter 6. CONGENITAL DIAPHRAGMATIC HERNIA

Definition. Congenital diaphragmatic hernia is a malformation caused by the failure of the diaphragm muscle to properly form. The diaphragm muscle is the structure that separates the thoracic cavity from the peritoneal cavity.

The **incidence** is approximately 1/5000 births.

History. The first description of diaphragmatic hernia was made by Ambroise Pare (1575). Bochdalek clarified the etiopathogenesis of the malformation (1848). Ladd and Gross reported the first therapeutic successes in 1940.

Embryology. The diaphragm, also known as transverse septum, begins to form during the eighth week of embryonic development, separating the thoracic and abdominal coelomic cavities. It develops laterally through the progression of pleuro-peritoneal folds in a central direction. Formation of the diaphragm ends paravertebrally on the left at the Bochdalek foramen, where approximately 85% of congenital diaphragmatic hernias occur here. On the right side, hernias occur less frequently due to the protective presence of the liver. More rarely, they occur through a retrosternal defect known as Morgagni foramen or Larrey cleft.

Pathological anatomy. It can occur isolated or as part of Cantrell syndrome (sternal cleft, omphalocele, cardiac ectopia, malformations of the large vessels, and congenital diaphragmatic hernia). The peritoneal cavity is underdeveloped, while the lung is collapsed, hypoplastic, and pushed toward the pleural dome. Additionally, the heart and mediastinal structures are shifted to the right. The hypoplasia of the left lung and sometimes heart malformations may further worsen the prognosis. Abdominal viscera-

including small intestine, ascending and transverse colon, spleen, partially the stomach and sometimes the left hepatic lobe- herniate into pleural cavity.

Pathophysiology. The herniation of the abdominal viscera into the thorax exerts compression on the fetal lung, leading to its underdevelopment on the affected side. Thus, at birth, the lung will have reduced gas exchange area, which will lead to respiratory distress in the newborn. Also, pulmonary vascular resistance is increased, leading to pulmonary hypertension.

Diagnostic. Prenatal diagnosis is possible using ultrasound and MRI from the 20th week of gestation. The lung-to-head ratio (LHR) can be calculated—the lower it is, the lower the chance of survival. The presence of the liver in the abdomen or chest correlates with a survival rate of 93% and 43%, respectively. Alpha-fetoprotein testing and assessment of the degree of alveolar surfactant maturation help to determine the appropriateness of labor induction.

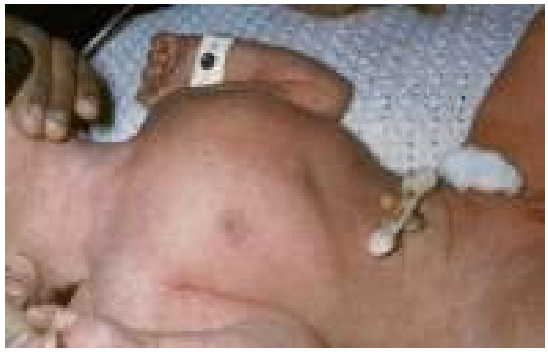


Fig. 6.1. Congenital diaphragmatic hernia,
excavated appearance of the abdomen

Postnatal diagnostic - immediately after birth, the clinical picture of respiratory distress frequently appears, with APGAR < 5, dyspnea, cyanosis, polypnea, intercostal retractions, globular shaped chest, and excavated abdomen (Figure 6.1). Percussion reveals tympanism in place of normal pulmonary resonance, while auscultation demonstrates absent breath sounds

and the presence of intestinal peristaltic sounds within thethoracic cavity, withcardiac sounds ausculted on the right side.

Chest X-ray ± with contrast medium (risk of aspiration) indicates the presence of intestinal loops in the thoracic cavity (Figure 6.2).

Differential diagnostic include pneumothorax, pulmonary atelectasis, esophageal atresia, and cyanogenic cardiac malformation.

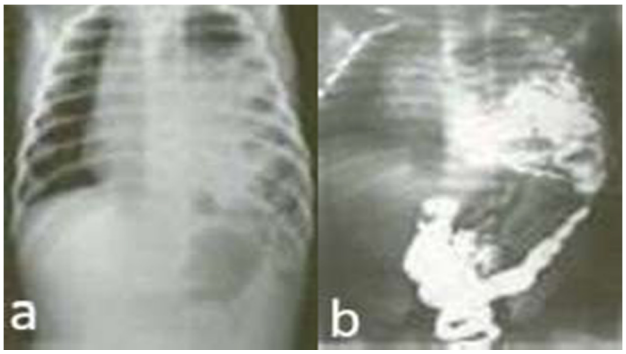


Fig. 6.2. Congenital diaphragmatic hernia.
Chest X-ray: **a.** Native and **b.** With contrast

Treatment. In selected cases (lung volume below 20 ml, liver-up), endoscopic balloon occlusion of the trachea (FETO) may be initiated in some specialized centers in the 34th week of pregnancy, followed by scheduled delivery by cesarean section in the 38th week of pregnancy. The results are contradictory. Delivery is recommended to be carried out in a specialized center. In fetuses with LHR >1.8, natural delivery followed by orotracheal intubation and conventional mechanical ventilation ("gentle ventilation" with PIP<28 cm H2O). For fetuses with LHR <1.5, cesarean section is recommended starting in week 38 of pregnancy.

Postnatally, the newborn is placed in an incubator and positioned in a lateral decubitus position on the affected side (usually the left). A nasogastric tube is inserted to prevent massive gastrointestinal pneumatization, and an

infusion line is placed to restore hydro-electrolytic and acid-base balance. In some cases, IOT= ETI ?? (endotracheal intubation) with O2 administration via the intubation tube is required. Some cases require preoperative arterio-venous or veno-venous ECMO (extracorporeal membrane oxygenation), HFOV (high-frequency oscillatory ventilation), inhalations with NO, administration of Sildenafil (for combating pulmonary hypertension pulmonary hypertension), surfactant, etc. Oral feeding is prohibited. It is also important to identify possible associated malformations.

From a surgical point of view, diaphragmatic hernia is a delayed emergency. The purpose of the operation is to return the herniated viscera into the abdomen cavity and to close the diaphragmatic defect. Approaches for left diaphragmatic hernia: abdominal, thoracic, thoraco-phreno-laparotomy, supra-umbilical median laparotomy, or usually left subcostal transverse laparotomy. The surgical approach can also be minimally invasive (thoracoscopic). For right diaphragmatic hernia, right thoracotomy, thoracoscopic approach, or laparotomy are suitable. After repositioning the herniated viscera, the edges of the diaphragmatic defect are approximated and closed with non-absorbable sutures in a "U" shape (horizontal matress sutures ??

). Pleural drainage is a controversial measure; some authors recommend it as mandatory, while others use it only in cases of significant intrapleural effusion. Sometimes the diaphragmatic defect is very large and does not allow primary closure, so a Goretex patch is usually used to augment and close the defect. The surgery is followed by care in the intensive care unit, with some patients requiring mechanical ventilation for an extended period. The aims are to stabilize any changes in cardiopulmonary function parameters,

gradually reintroduce oral feeding, and treat any associated pulmonary infections.

Complications: bleeding at the surgical site, local hematoma, infection, recurrence of diaphragmatic hernia, sepsis, respiratory failure, death.

Prognosis is influenced by the early diagnostic, the degree of pulmonary hypoplasia, and the early treatment. The postoperative evolution is better in the absence of associated malformations; mortality is 15-20%.

Chapter 7. ESOPHAGEAL ATRESIA

Definition. Esophageal atresia is the interruption of the continuity of the esophageal lumen due to changes in the normal embryogenesis of the esophagus during the intrauterine period, with or without the presence of abnormal communication with the trachea.

Incidence is approximately 1 case per 3,000-4,000 births.

The **etiology** is still unknown, but familial aggregation should be emphasized. At 20 days of gestation, a diverticulum develops on the midline of the ventral wall of the digestive tract, which subsequently separates into the esophagus and trachea by joining the proliferative edges of the cells in the endothelial layer. The separation of the trachea from the esophagus first occurs at the carina and proceeds cranially. It is possible that too rapid elongation reduces the tissue necessary for the esophagus and causes an imbalance in the growth of the trachea and esophagus.

Pathological anatomy. Ladd and Roberts (1944) proposed one of the most widely used classifications (Figure 7.1):

- Type I: Esophageal atresia without tracheoesophageal fistula (5-8%)
- Type II: Esophageal atresia with proximal tracheoesophageal fistula (1%)
- Type III: Esophageal atresia with distal tracheoesophageal fistula (85-88%)
    - Type IIIa: with a large distance between the esophageal stumps
    - Type IIIb: with a short distance between the esophageal stumps
- Type IV: Esophageal atresia with double tracheoesophageal fistula (5-8%)
- Type V: Tracheoesophageal fistula without atresia (H-shaped fistula)

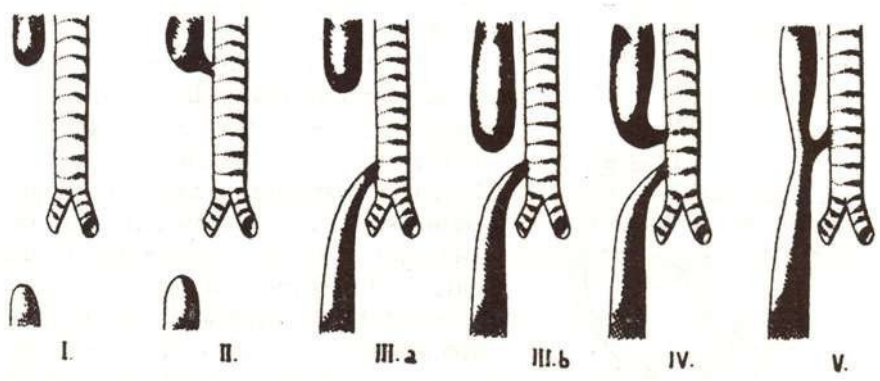


Fig. 7.1. Ladd and Roberts classification of esophageal atresia

Approximately 50% of newborns with esophageal malformations present with major congenital anomalies affecting other organ systems. To categorizfe the spectrum of the malformation associated with esophageal atresia, the literature has proposed the VACTERL association, which includes Vertebral and Sacral anomalies, Anorectal malformations, Cardiovascular malformations, Tracheoesophageal fistula, Renal malformations, Limb malformations. Esophageal atresia may be associated with Down syndrome and CHARGE syndrome—coloboma (microcephaly or anencephaly), congenital heart disease, choanal atresia, mental retardation, genital malformations, and ear malformations.

Pathophysiology. After birth, salivary secretions that either stagnate at the bottom of the upper esophageal sac or drain through the fistula reach the lungs, causing bronchoalveolar flooding and subsequent bronchopneumonia through microbial colonization. Additionally, in type III or IV atresia, some of the air breathed enters the stomach and the rest of the digestive tract through the fistula. Abdominal distension occurs with diaphragmatic elevation and reduces respiratory mechanics. At the same time, gastric distension causes gastrointestinal reflux, so that gastric juice with a corrosive effect on the alveolar epithelium is expelled into the lungs along

with the air. The combination of these disorders is very suggestive by Mallet in the formula: "the newborn with esophagal atresia and tracheoesophageal fistula swallows into the trachea and breathes into the abdomen.

Diagnostic. Antenatal ultrasound performed on the mother can reveal polyhydramnios in 85% of cases of esophageal atresia without fistula but cannot establish a definitive diagnostic of esophageal atresia.

Postnatally, the clinical picture is dominated by the inability to swallow, hypersalivation, and respiratory disorders. Before the first feeding, a radiopaque nasogastric tube must be inserted into the newborn's esophagus. In esophageal atresia, the nasogastric tube does not reach the stomach and stops at approximately 10-12 cm from the dental arch. To confirm the diagnostic, a chest and abdominal X-ray is performed with the radiopaque nasogastric tube placed in the upper esophageal stump (Figure 7.2).

In 1962, Waterston stratified patients with esophageal atresia into three risk groups as follows:

- group A - BW > 2500g, no associated malformations, no pulmonary changes
- Group B -BW 1800-2500g or GN > 2500 g but with malformations and/or moderate pneumonia
- Group C, BW < 1800g with severe malformations and pneumonia

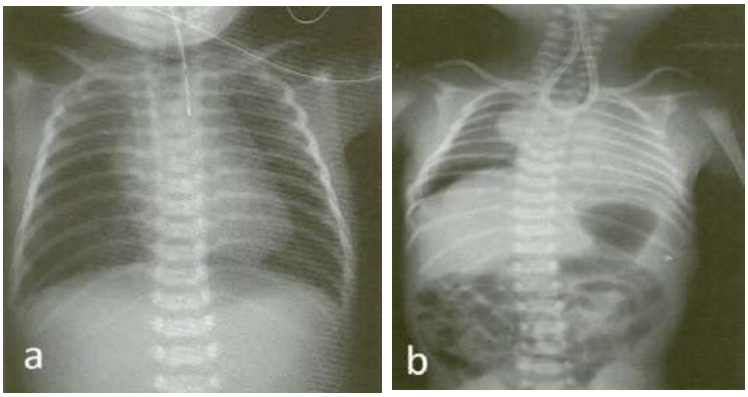


Fig. 7.2. Thoracoabdominal radiograph, esophageal atresia:
a. Type I without trachea-esophageal fistula and **b.** Type III with fistula

Differential diagnostic is made with traumatic perforation of the hypopharynx, craniocerebral trauma caused during birth, swallowing disorders encountered in premature infants, laryngotracheal clefts, congenital pharyngeal pseudodiverticula, choanal atresia, congenital diaphragmatic hernia.

Postnatal **treatment** is an emergency. The newborn will be placed in a prone position in a slight Trendelenburg position to avoid aspiration of saliva into the respiratory tract. The esophageal stump is aspirated every 5-10 minutes. Radiological diagnostic should not delay emergency transport to specialized clinics. Transport will be carried out under maximum safety conditions, assisted by a doctor.

Surgical treatment is performed under general anesthesia with oro-tracheal intubation. The first step is usually to perform a bronchoscopy to visualize the tracheoesophageal fistula(s) and mark them by placing thin catheters, which will help identify them intraoperatively. The classic surgical approach is performed through a right extra-pleural thoracotomy in the IV/V intercostal space. Alternatively, a thoracoscopic approach can be used (Figure 7.3). The

fistula is ligated and, if the distance between the stumps is not too great, a termino-terminal (end-to-end) esophageal anastomosis is performed. When the esophageal ends are more than 4 cm apart or 2 vertebral spaces apart (long gap esophageal atresia – LGEA), primary anastomosis cannot be performed. In this situation, after ligating the trachea-esophageal fistula, procedures are performed to lengthen the esophageal stumps (by progressive traction with sutures or magnets). Other times, a cervical esophagostomy and gastrostomy are performed, followed by esophagoplasty around the age of 12 months, using the stomach, small intestine, or colon as an esophageal substitute.

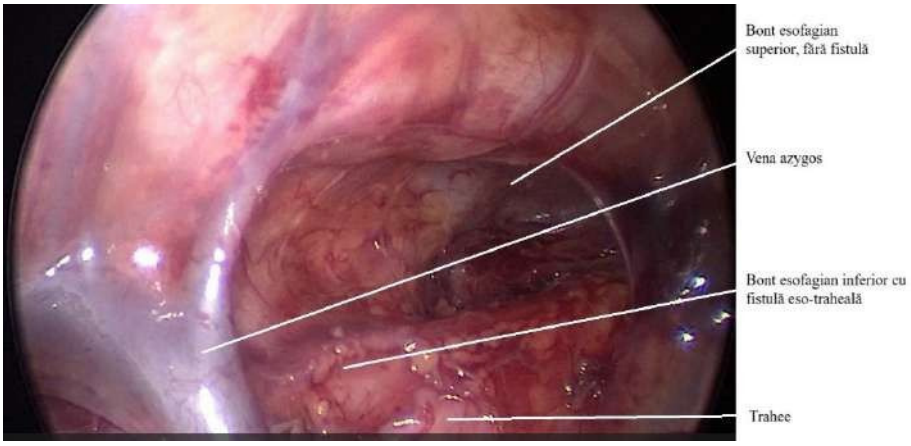


Fig. 7.3. Esophageal atresia with distal esophagotracheal fistula (type IIIb)

Complications may be nonspecific, common to any surgical procedure (bleeding, infection), or specific: anastomotic dehiscence, fistula recurrence, anastomotic stricture, gastroesophageal reflux with aspiration pneumonia, esophageal motility disorders, irritative cough, tracheomalacia, chest deformities.

Prognosis. In cases classified as Waterson risk groups A and B, the cure rate is up to 90%, and in patients in risk group C, mortality has decreased from 90% to approximately 20%.

Chapter 8. HYPERTROPHIC PILORIC STENOSIS (HPS)

Definition. A condition in newborns and infants characterized by hypertrophy of the circular and longitudinal muscle layers of the pylorus, which causes an obstruction in gastric emptying.

Incidence. Hypertrophic pyloric stenosis is one of the most common causes of gastric obstruction in newborns. The prevalence of cases varies from 1.5-4 per 1,000 newborns, with a higher prevalence in Caucasians. Males are more commonly affected, with a ratio of 4:1.

Etiology. The cause of hypertrophic pyloric stenosis remains unknown. Both genetic and environmental factors have been described in the etiopathogenesis of this condition. The increased incidence in Caucasians, the preponderance among males, and the increased risk for firstborn newborns with a positive family history support the theory of genetic predisposition.

Environmental factors such as exposure to erythromycin and pesticides, artificial feeding of the newborn, and seasonal variability have been implicated as predisposing factors in HPS. Recent studies have raised the possibility of an association between hypertrophic pyloric stenosis and a deficiency of certain gastrointestinal peptides and growth factors in newborns (low neurotrophins, deficiency in nitric oxide synthesis) or hypersecretion of others such as substance P or gastrin.

Clinical and paraclinical picture. Pathognomonic for this condition are white, non-bilious, explosive (projectile) vomiting, with sudden onset in a newborn aged 2-8 weeks, after a symptom-free interval since birth. The expelled gastric contents are white and appear immediately after feeding or 10-15 minutes after a meal. This interval increases as gastric dilatation progresses. Occasionally, bloody streaks may be present due to associated

gastritis. The newborn's appetite is preserved. The weight curve is initially stationary, then becomes descending.

Depending on the time of diagnostis, the clinical presentation may range from a hemodynamically stable newborn to a lethargic, somnolent, dehydrated newborn. In general, the newborn has constipation. Diarrhea (hunger stools) may also occur.

Clinical examination may reveal gastric peristalsis (contractile waves) visible in the epigastrium. Palpation of the pyloric olive (in the right hypochondrium, subhepatic) is possible in 70-90% of cases. To palpate the olive, the newborn must be relaxed.

Lab exam usually reveals dehydration, hypochloremia, hypokalemia metabolic alkalosis.

Positive diagnostic. Although the diagnostic of hypertrophic pyloric stenosis can be made based on clinical presentation and laboratory investigations (non-bilious projectile vomiting, peristaltic wave present in the epigastrium, hypochloremic metabolic alkalosis, hypokalemia), the gold standard in diagnostic is abdominal ultrasound (muscle layer thickness ≥4 mm and pyloric length ≥16 mm). This is non-invasive, non-irradiating but operator-dependent (Figure 8.1). If this method is not available, barium passage is the alternative method of diagnostic.

Differential diagnostic is made with non-bilious vomiting of medical causes such as food allergies, gastroesophageal reflux disease, intolerance to certain milk formulas, gastroenteritis, pylorospasm, pyloric duplication, ectopic pancreatic tissue at the pyloric level, adrenogenital syndrome, metabolic disorders, increased intracranial pressure, gastric tumors, or tumors causing gastric compression.

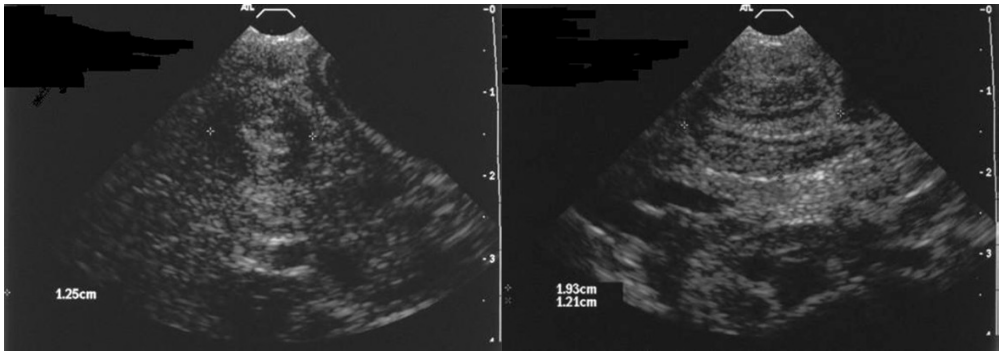


Fig. 8.1. Hypertrophic pyloric stenosis – ultrasound: A cross-section; B longitudinal section

Preoperative treatment consists of correction of fluids, electrolytes and acid-base imbalance through intravenous administration of appropriate fluids, ensuring stabilization of the infant before surgical intervention. Surgical treatment consists of Fredet extramucosal pyloromyotomy, which can be performed either conventionally or laparoscopically (Figure 8.2). Postoperatively, analgesic and anti-inflammatory therapy is instituted, and feeding can be resumed 4 hours after the procedure.

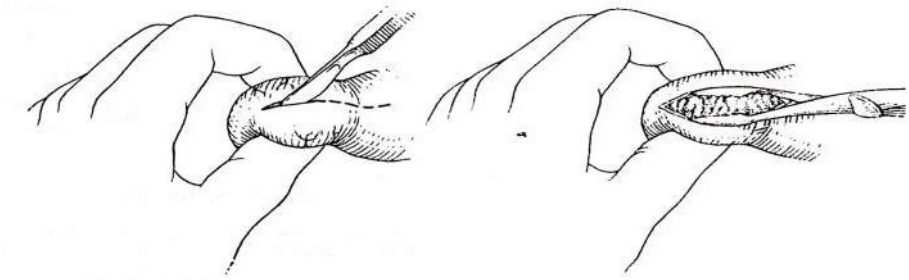


Fig. 8.2. Fredet extramucosal pylorotomy

Complications associated with pyloromyotomy include perforation of the duodenal mucosa, incomplete myotomy and duodenal lesions, prolonged postoperative vomiting, wound infection, and incisional hernia.

Prognosis. Without surgical treatment, frequent vomiting dehydrates the infant, leading to weight loss and death due to fluid and electrolyte

disorders, severe electrolyte imbalances, tetany, malnutrition, aspiration bronchopneumonia. In patients who undergo surgical treatment, the outcome is favorable in most cases, with normal subsequent growth and weight gain in operated infants. The prognosis is good in both the short and long term, with minimal post-operative mortality and morbidity.

Chapter 9. DUODENAL ATREZIA AND STENOSIS

Congenital duodenal malformations are some of the most common causes of high intestinal obstruction in newborns and are divided into three main categories:

1. Duodenal atresia
2. Duodenal stenosis, with its two forms, intrinsic and extrinsic
3. Mixed duodenal obstruction, represented by the annular pancreas

9.1. DUODENAL ATREZIA

Definition. Duodenal atresia is a congenital malformation of the duodenum characterized by complete luminal obstruction, causing high intestinal obstructuion in neonatal period.

Incidence. Global incidence of duodenal atresia has been estimated at 1/6,000-10,000 births. It is the most common cause of duodenal obstruction in newborns.

Etiopathogenesis. It is assumed that there are certain deficiencies in the recanalization of the small intestine mucosa during the embryonic development of the fetus, but the mechanism is not fully understood. The site of duodenal atresia is more common at the D2-D3 junction. Approximately 50% of newborns with duodenal atresia or stenosis present with associated anomalies or malformations such as Down syndrome, annular pancreas, congenital heart defects, and intestinal malrotation.

Pathological anatomy. Duodenal atresia is classified into three categories:

I. Type I atresia is the most common (92%) and is caused by a septum formed from the duodenal mucosa and submucosa without involvement

of the muscularis, resulting in complete obstruction. A variant of type 1 atresia may be obstruction of the lumen due to an elongated, thin mucosa that collapses into the duodenal lumen. If the septum has a central opening, type 1 atresia can cause incomplete obstruction.

II. Type II atresia (1%) is due to a non-luminal fibrous cord connecting the dilated and hypertrophied proximal segment to the narrowed distal duodenum. The mesentery is intact.

III. Type III atresia (complete – 7%). The proximal segment is significantly dilated and hypertrophied. In this type of atresia, there is no connection between the proximal and distal segments. (Figures 9.1, 9.2)

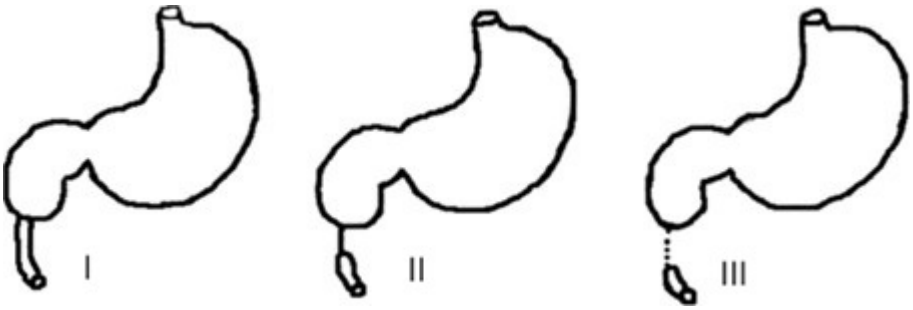


Fig. 9.1. Type I, II, and III duodenal atresia

9.2. DUODENAL STENOSIS

Definition. Duodenal stenosis is a congenital malformation of the duodenum that causes partial obstruction of the duodenum. It can be classified as intrinsic or extrinsic stenosis depending on the mechanism of occurrence.

Intrinsic stenosis is caused by a diaphragm that obstructs the lumen, presenting a narrow opening that does not allow normal passage of gastric contents and causes dilation and hypertrophy of the proximal duodenal segment and stomach (Figure 9.3).

Extrinsic duodenal stenosis is caused by external compression like Ladd ligament -congenital fibrous tissue peritoneal band that forms due to intestinal malrotation, extending from the abnormally located cecum (often located in the epigastric area) to the posterior abdominal wall, potentially causing intestinal obstruction by compressing the duodenum (Figure 9.4). Extrinsic stenosis can also be caused by the preduodenal portal vein, aorto-mesenteric clip, or duodenal duplication.

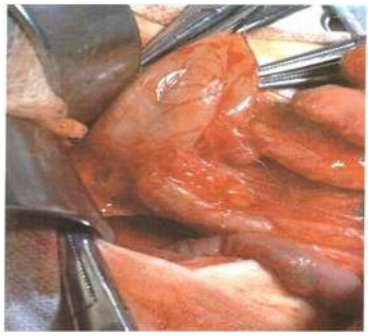


Fig. 9.2. Duodenal atresia.

Fig. 9.3. Duodenal stenosis

5.3. ANNULAR PANCREAS

Definition. The annular pancreas is a congenital malformation resulting from a developmental defect of the pancreas, consisting of a band of pancreatic tissue surrounding and intersecting the second portion of the duodenum. This anomaly is often associated with duodenal atresia or intrinsic duodenal stenosis, resulting in a mixed (extrinsic and intrinsic) duodenal obstruction (Figure 9.5).

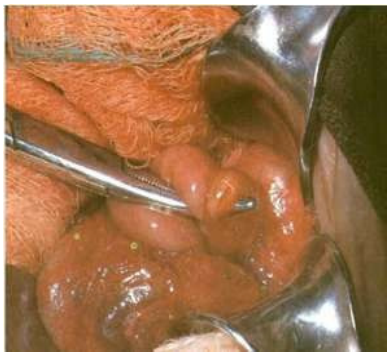


Fig. 9.4. Ladd's ligament

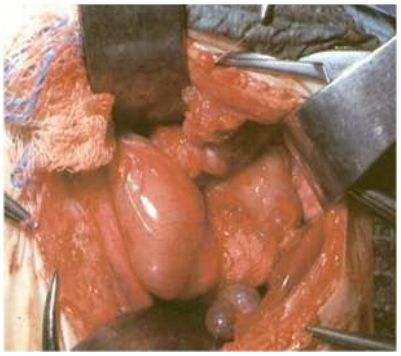


Fig. 9.5. Annular pancreas

Clinical and paraclinical picture. Advances in prenatal medicine have made it possible to diagnose this pathology by fetal ultrasound. In general, the diagnostis can be made in the 7th to 8th month of pregnancy, especially in the context of a pregnancy that has developed with hydramnios.

Newborns with duodenal atresia present with a scaphoid abdomen, bilious vomiting within the first hours after birth, and failure to pass meconium. In 15% of cases, newborns may present with non-bilious vomiting due to the pre-ampullary location of the duodenal atresia. Nasal-gastric tube aspiration of the newborn may suggest the presence of obstruction. A volume of more than 20 ml of gastric aspirate is suggestive of obstruction, whereas volume under 5 ml are considered within the normal range for neonates.

Abdominal radiography reveals the "double bubble" sign and the absence of intestinal gas distal to the defect (Figure 9.6). Barium passage can be used to rule out malrotation and intestinal volvulus.

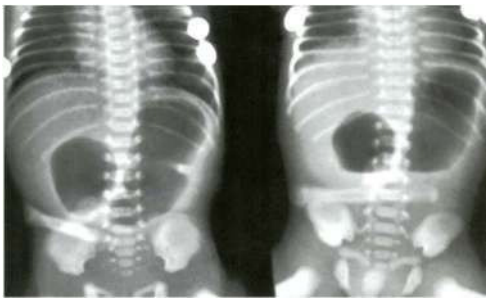


Fig. 9.6. Duodenal atresia – X-ray showing a "double bubble" appearance.

Laboratory tests such as complete blood count with leukocyte formula, blood electrolytes, and ASTRUP are also necessary to determine the status of the newborn. Typically, patients with multiple vomiting episodes present with hypokalemia hypochloremia, metabolic alkalosis. Cardiac ultrasound is necessary due to the increased risk of associated congenital heart malformations. In duodenal stenosis, obstructions are usually incomplete, with some passage of gastric content.

Patients with duodenal stenosis typically present with more subtle symptoms including intermittent vomiting, and feeding difficulties rather than the persistent, high-volume vomiting seen in complete atresia. Radiological examination reveals air in the intestine and the "double bubble" sign is not present.

Positive diagnostic. The postnatal diagnostic is established immediately after birth based on clinical data and imaging investigations. If the pregnancy was monitored, a positive diagnostic of obstruction is made antenatally with radiological confirmation immediately after birth. For newborns who presented with bilious vomiting at birth and had more than 20 ml on nasogastric aspiration, 40-60 ml of air can be introduced to reproduce the "double bubble" sign on radiograph and confirming the diagnostis.

Differential diagnostic. Includes malformations of the small bowel, volvulus, meconium ileus, meconium peritonitis, Hirschsprung's disease.

Treatment. Preoperative gastric decompression, correction of the newborn's hydro-electrolytic imbalance is required. In general, these patients are premature or newborns with very low birth weight, which requires careful preoperative preparation. Surgical management involves correction of duodenal malformation. The most employed surgical technique for correction of duodenal atresia, stenosis, or annular pancreas is a diamond-shape duodeno-duodenal anastomosis, which can be performed via either an open (conventional) or laparoscopic approach.

Complications. Anastomotic fistula, duodenal obstruction, and local infection are the most common complications. Long-term complications may include gastroesophageal reflux, peptic ulcer, anastomotic ulcer, megaduodenum, gastritis, and peritoneal adhesions.

Prognosis. Early postoperative mortality is 3-5%. Many deaths occur due to associated malformations. Long-term survival of these patients is 90% and was improved by the new antenatal diagnostic techniques as well as preoperative management and surgical techniques.

Chapter 10. INTESTINAL ATRESIA AND STENOSIS

Definition. Intestinal atresia and stenosis are the most common malformations of the small intestine and are also the most common causes of intestinal obstruction in newborns.

According to Grosfeld's classification, congenital intestinal malformations are divided into:

- Stenosis, defined as a localized narrowing of the intestinal lumen without disruption of the intestinal wall. Often, the musculature is irregular, and the submucosa is thickened.
- Atresia

I. In type I jejunoileal atresia, intestinal obstruction occurs secondary to a membrane made of the mucosa and submucosa, while the muscularis and serosa remain intact, causing dilation of the proximal intestine and collapse of the distal intestine. The intestine has a continuous wall, without mesenteric defect, with normal intestinal length.

II. Intestinal obstruction is due to a fibrous cord. The mesentery is intact.

III. There are two subcategories:

- IIIa: In this type of atresia, the proximal intestinal loop ends blindly, with no continuity between the proximal and distal intestine. The mesentery has a V-shaped defect. The length of the intestine is generally shorter than normal.
- IIIb (apple peel, Christmas tree): consists of jejunal atresia accompanied by anatomical changes in the mesenteric arteries, and the length of the intestine is significantly reduced. The decompressed distal small intestine lies freely in the abdomen and

takes on a helical configuration around a single vessel originating from the ileocolic artery or the right colic arches.

IV. The intestine has multiple atretic areas or a combination of type I, II, or III atresia (Figure 10.1).

Incidence. The prevalence of congenital intestinal malformations varies worldwide. An approximate rate of 1 in 5,000 births can be estimated, with no significant sex predilection; however, this condition occurs more frequently in premature newborns , with a rate of 1 in 3.

Etiopathogenesis. Although these malformations are believed to occur sporadically, cases of familial intestinal atresia have been described. It is generally accepted that jejunoileal atresia occurs because of intrauterine ischemic injury to the mesentery. Intrauterine vascular disturbance can cause ischemic necrosis of the intestine, leading to resorption of the affected segment(s). The vascular theory remains a widely considered explanation for intestinal atresia. In the 10th week of intrauterine life, when the intestine returns to the coelomic cavity, or later in intrauterine life due to volvulus, intussusception, or strangulation resulting in ischemia, necrosis and subsequent atresia of one or more intestinal segments.

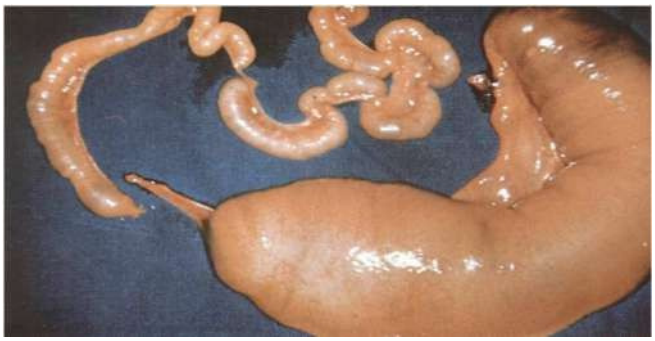


Fig. 10.1. Resected bowel with 2 atretic areas.

Clinical and paraclinical picture. The clinical presentation of intestinal atresia is consistent with intestinal obstruction, characterized by hallmark signs and symptoms. Bilious vomiting is the predominant symptom, occurring on the first day after birth in jejunal obstructions and after 24 hours in ileal obstructions. In low intestinal obstructions, vomitting occurs at longer intervals, is abundant, and it is initially bilious, later becoming fecaloid as stasis and bacterial overgrowth develop. Abdominal distension is a common finding in intestinal atresia, typically mild in high obstructions (e.g. proximal jejunum) and marked in lower obstruction. Usually there is no meconium passing. Mucus may be observed on rectal examination.

Compared to congenital duodenal malformations, where fetal ultrasound is extremely useful in diagnostic, in intestinal malformations fetal ultrasound may be relevant if the occlusion is more distal and greatly dilated intestinal loops with accelerated peristalsis are observed. Guidelines recommend the use of MRI in pregnant women in whom fetal ultrasound has revealed the above-mentioned changes. After birth, air enters the digestive tract, reaching the jejunum after 1 hour and the ileocecal valve after 3 hours.

Plain abdominal radiography can help approximate the location of the obstruction by assessing the degree of intestinal gas distension and identifying the air-fluid levels. Atresia located in the terminal ileum and colon can be confused with congenital megacolon = dull (unaerated) crescent in the small pelvis. (Figure 10.2)

In the case of stenoses, the clinical picture is one of sub-occlusive syndrome, and barium passage is useful for the diagnostic.

Positive diagnostic. It is based on the clinical triad (bilious vomiting, abdominal distension, absence of intestinal transit) and plain abdominal X-ray in the orthostatic position.

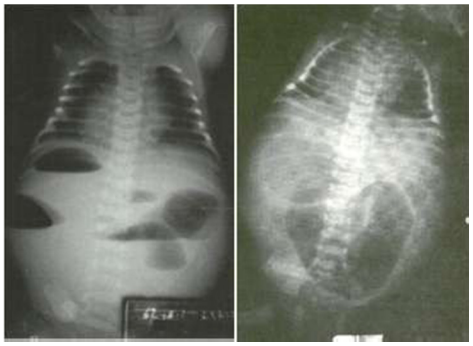


Fig. 10.2. Jejunoileal atresia

The differential diagnostic includes intestinal malrotation with or without volvulus, meconium ileus, intestinal duplication, internal hernia, colonic atresia, adynamic ileus secondary to sepsis, and total colonic aganglionosis.

Treatment. Delayed diagnostis can lead to intestinal damage with necrosis and perforation, electrolyte imbalances, and sepsis. Preoperative management should include gastric decompression (nasogastric tube), correction of electrolyte imbalances, and hypovolemia. Antibiotic therapy should be initiated if perforation or infection is suspected. Surgical management is tailored to the type of defect, location, and intestinal damage and typically involves an exploratory laparotomy to evaluate the lesions followed by restoration of intestinal continuity.

Infectious **complications** (peritonitis, pneumonia, sepsis), anastomotic failure (obstructions or anastomotic fistulas), and short bowel syndrome are the most common and feared postoperative complications. Infants with more distal ileal resections are prone to malabsorption (fats, biliary salts, vitamin B12, calcium, magnesium), diarrhea (steatorrhea), and increased bacterial proliferation.

Prognosis. Infections such as pneumonia, peritonitis, or sepsis are the most common causes of early death in infants with intestinal atresia. Before the introduction of parenteral nutrition, mortality and morbidity were high in this patient group. Recent studies show that newborns with congenital intestinal malformations and birth weight below 2 kg are at increased risk of prolonged hospitalization, with an associated mortality rate of approximately 3.3%.

Chapter 11. BOWEL MALROTATION AND VOLVULUS

Definition. Intestinal malrotation is characterized by disruption of the physiological process of rotation and fixation of the fetal intestine. Volvulus is a complication of intestinal malrotation, characterized by the twisting of one or more intestinal loops around the mesenteric axis which can compromise blood flow and lead to ischemia.

Incidence. The malformation may remain asymptomatic throughout life, making its true incidence difficult to determine. It is sometimes found incidentally during surgery performed for other diseases. The condition occurs more frequently in males, with a male to female ratio of approximately 2:1.

Etiology. The causes of malrotation could not be accurately identified. In the case of volvulus, it is assumed that the triggering factor for intestinal twisting around the mesenteric axis is exaggerated peristalsis or sudden distension of the small intestine. Various forms of intestinal malrotation occur through disruption of the physiological process of intestinal rotation and fixation, which takes place during the first 3 months of intrauterine life in 3 distinct consecutive phases (Figure 11.1):

I. Phase 1 - the primitive intestine is a uniform tubular structure, vascularized posteriorly by the superior mesenteric vessels, which elongates in the sagittal plane in its middle portion, laying in the coelomic cavity

II. Phase 2 - the primitive midgut now located outside the celomic cavity, rotates counterclockwise 180° around its mesenteric axis, then reintegrates into the abdominal cavity of the intestine. The end of the second phase is marked by a rotation of the duodenum under and to the left of the mesenteric vessels.

III. Phase 3 - the integration of the entire primitive middle intestine is completed, with the colon occupying the entire left hemiabdomen. The phase ends with a final counterclockwise rotation of 90°, causing the cecum and right colon to pass over the superior mesenteric vessels toward the right iliac fossa. After completing a full 270° rotation, the mesentery become fixed, and the coalescence of the peritoneal fasciae will form (by the end of the third month of gestation).

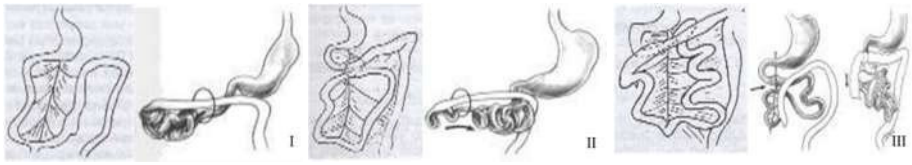


Fig. 11.1. Phases of the physiological process of intestinal rotation and coiling

The cecum develops and reaches the right iliac fossa at birth. Various attachments form during this time, securing the colonic framework and the root of the mesentery, thereby preventing any possibility of volvulus. The duodenum, the ascending and descending colon are secured to the posterior abdominal wall.

Anatomo-pathological forms:

- Absence of any rotation – the small bowel is in the right hemiabdomen, and the colon is in the left hemiabdomen. No intestinal fixation has occurred.
- Incomplete rotation – the process of intestinal rotation stops (usually at 180°, the cecum is located on the midline in the epigastric region).
- Reverse rotation (usually 90°)

Clinical manifestations:- Malrotation may be asymptomatic and remain asymptomatic throughout life. The most common clinical manifestations are:

- Midgut volvulus. This is the most common clinical manifestation (75% of cases). Due to the lack of bowel attachment, the entire small bowel and part of the colon twist (volvulate) around an axis represented by the superior mesenteric artery. It usually occurs in the neonatal period and is a dramatic event, with sudden onset. The child exhibits severe abdominal pain, excruciating crying, bilious vomiting, and rapidly progressive abdominal distension. As the condition progresses, vomiting may become fecaloid and occasionally bloody, and abdominal wall edema or even muscle contracture/ rigidity can develop. Patients may exihits absence of stool passage, or occasionally melena reflecting intestinal obstruction and mucosal compromise.
- Duodenal obstruction caused by Ladd's ligament. Ladd's ligament occurs in incomplete malrotations when the cecum is in the epigastric region and is a parietal-colonic fibrous band that exerts compressive pressure on the duodenum and causes its extrinsic obstruction.
- Chronic volvulus is a partial, intermittent twisting of intestine that typically occurs in childhood and adolescence. It usually manifests as recurrent colicky abdominal pain, intermittent bilious vomiting, and malabsorption of varying degrees.
- Malrotation in other congenital conditions: congenital diaphragmatic hernia, omphalocele, gastroschisis.

Positive diagnostic is made incidentally in asymptomatic cases or based on the clinical picture. For confirmation, an abdominal X-ray performed in an upright position on an empty stomach with contrast medium

is required to visualize the obstruction and assess intestinal rotation.. This highlights air distension of the stomach, hypo-aeration in the rest of the abdomen, and the absence of colic gas images in the right hemiabdomen in the case of volvulus. Contrast radiography is the examination of choice and reveals the position of the duodenum and small intestine to the right of the spine, with the duodenum appearing imprinted by Ladd's ligament (Figure 11.2). In intestinal volvulus on the common mesentery, the characteristic image is "corkscrew" (Figure 11.3).

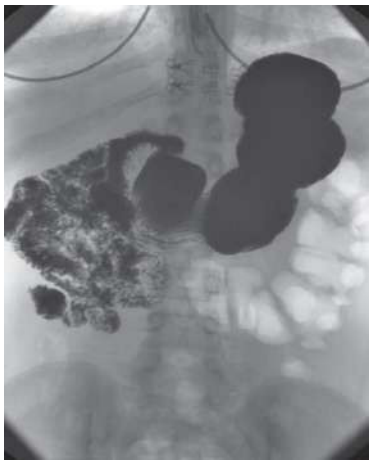


Fig. 11.2. Incomplete intestinal malrotation (180°) with Ladd's ligament

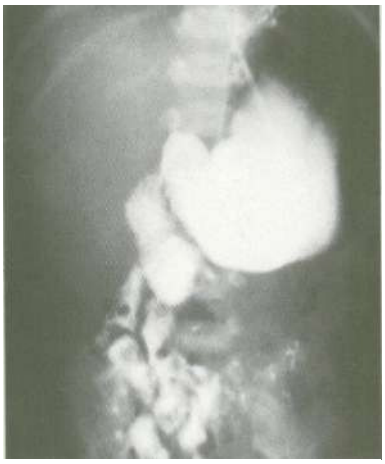


Fig. 11.3. Midgut volvulus, corkscrew image

Treatment. In principle, treatment is primarily surgical in cases of malrotation complications (duodenal stenosis due to Ladd's ligament, volvulus, internal hernia). Intestinal volvulus is a major surgical emergency requiring immediate intervention , therefore preoperative preparation will be shortened as much as possible and limiting ourselves to resuscitation therapy. Median laparotomy will allows visualization of the volvulated intestinal loops, which are then de-rotated, and their viability assessed. A decision will be made regarding the need for resection of the compromised areas. When

extensive resections are necessary or when the viability of the intestinal segments is questionable, resections are not performed; instead, the abdomen is closed and reopened 24-48 hours ("second look"). When we identify volvulated loops with perforations and intraperitoneal intestinal fluid extravasation, intestinal resections will be performed up to healthy segments, followed by either primary anastomosis or creation of diverting enterostomies. If the loops regain normal color and appear viable after devolvement, they are repositioned in the peritoneal cavity, starting with the duodenum and jejunum on the right side and the cecum in the left hypochondrium. It is forbidden to correct malrotation by repositioning the intestinal segments in anatomical position or fixing them to the abdominal wall.

To prevent further volvulation or when malrotation is discovered accidentally, the Ladd procedure is performed. This involves dissecting the peritoneal membrane between the duodenum and the cecum to widen the base of the mesenteric implant. If there is a Ladd's ligament, it must be sectioned. Tactical (Prophylactic) appendectomy may be considered due to the ectopic postoperative position of the cecum and appendix and possible difficulties in diagnosing acute appendicitis that may occur later in life.

Prognosis. The main prognostic factor in intestinal volvulus is early diagnostic and surgical intervention. If there is no massive loss of intestine with secondary short bowel syndrome, the prognosis is favorable.

Chapter 12. CONGENITAL MEGACOLON HIRSCHSPRUNG 'S DISEASE

Definition. Congenital megacolon (Hirschsprung's disease) is a congenital anomaly characterized by functional, partial, or complete colonic obstruction resulting from the absence of ganglion cells in the nervous plexuses (**myenteric (Auerbach) and submucosal (Meissner) plexuses**) within the colon wall.

The **incidence** is approximately 1:5000 live births.

Etiology. During normal prenatal development, cells from the neural crest migrate to the large intestine to form the myenteric plexus (Auerbach's plexus) and the submucosal plexus (Meissner's plexus). In Hirschsprung's disease, the migration that occurs in the first 12 weeks of gestation is incomplete, so that part of the colon lacks the ganglia that regulate colonic activity. The earlier the neural crest cell migration ends, the longer the aganglionic segment of intestine.

Pathological anatomy. The length of the aganglionic colon segment is variable, usually extending from the rectum to the rectosigmoid region, but it can involve the entire colon (total colonic aganglionosis) and even the small intestine. Above the aganglionic area is the transition zone, where ganglion cells are present but insufficient in number for normal function. Proximal to this is the normally innervated intestine (Figure 12.1).

Pathophysiology. The central pathophysiological element is the lack of propagation of the peristaltic wave in the aganglionic segment, so feces cannot pass further, creating a functional obstruction. Due to the functional obstruction downstream, feces stagnate in the normally innervated segment of the intestine, where bacterial activity and fermentation processes are

exacerbated. Furthermore, the bacterian overgrowth and toxins produced at this level may translocate into the bloodstream, resulting to a toxic-septic state.

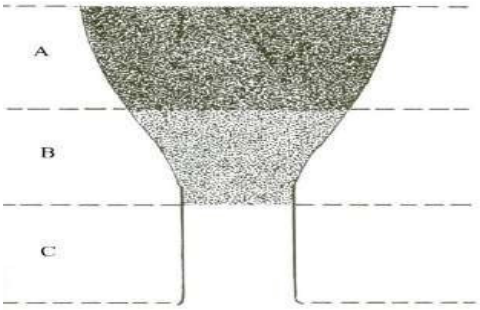


Fig. 12.1. Graphical representation of the three zone in Hirschsprung's disease.
A Normally innervated area
B Transitional zone
C Aganglionic zone

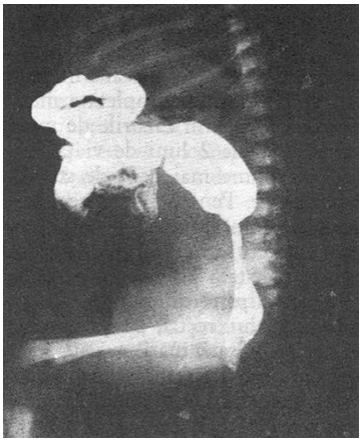


Fig. 12.2. Congenital megacolon, irrigography

Clinical picture. In typical forms of Hirschsprung's disease, newborns with congenital megacolon do not pass meconium in the first 24-48 hours. After a few days, the patient progressively develops abdominal distension, vomiting, initially bilious than fecaloid, and progressive deterioration of their general condition. This may be followed by an onset of explosive diarrhea, characterized by enterocolitis, with partial decompression of the distended abdomen. Enterocolitis is ofte3n particularly aggressive, with rapid progression to severe sepsis and systemic toxemia. It can even be fatal. This sequence of constipation, abdominal distension, and enterocolitis repeats periodically, and the patient's condition progressively deteriorates. There may also be tolerated forms of megacolon in which the symptoms are more attenuated, dominated by constipation (stools every 5-6 days), progressive abdominal bloating, and poor child development. On rectal examination, the rectal ampulla is empty, the abdomen is distended and bloated.

The investigation of choice is the contrast enema. The characteristic image is a funnel-shaped colon. The normally innervated proximal colon is dilated than it progressively narrows (transitional zone) towards the distal, aganglionic segment, which is of smaller or normal caliber (Figure 12.2). On plain abdominal radiography large hydro-air images can be seen.

Positive diagnostic is based on suggestive clinical findings and imagistic investigations. The definitive diagnostis is made by biopsy and histopathological examination. It reveals the absence of ganglion cells in the intestinal wall and the presence of thick unmyelinated nerve fibers. As an alternative to biopsy, acetylcholinesterase activity in the rectal wall can be measured (increased in Hirschsprung's disease). Anorectal manometry is a diagnostic tool that can identify the functional abnormalities of the anorectal complex that controls defecation. In a normal colon, distension of the rectal ampulla causes relaxation of the internal sphincter and a decrease in pressure in the anal canal; failure of this reflux suggests irschsprung’s disease or other motility disorders.

Differential diagnostic. All causes of neonatal mechanical obstruction must be excluded: anorectal malformations, intestinal atresia, meconium ileus, small left colon syndrome, colonic atresia, volvulus, pelvic tumors, hypothyroidism, and other forms of constipation.

Treatment. In the first stage, during the neonatal period or in infants, medical or surgical procedures are necessary to combat constipation and prevent the onset of enterocolitis. Rectal lavages are performed several times a day, initially in the hospital and then by the parents at home. If adequate decompression cannot be achieved through lavages, a temporary colostomy is necessary. If enterocolitis occurs, it requires prompt treatment.

The definitive treatment for congenital megacolon consists of surgical resection of the aganglionic segment and anastomosis of the normally innervated colon to the anal canal. This procedure is usually performed after a minimum stabilization period of 8-12 weeks. There are three common basic surgical procedures: Swenson (anastomosis of the innervated colon 2 cm from the ano-cutaneous line) Soave (excision of the rectal mucosa and lowering of the normally innervated colon through a rectal muscle sleeve and then anastomosis to the anus) and Duhamel (retrorectal anastomosis of the normally innervated colon 2 cm above the ano-cutaneous line with preservation of a rectal segment).

Chapter 13. ANORECTAL MALFORMATIONS

Definition: Anorectal malformations represent a spectrum of congenital anomalies in which the terminal portion of the digestive tract opens abnormally in relation to its anatomical position.

Incidence is approximately 1:3500-1:5000 newborns.

Pathological anatomy. From a pathological anatomy perspective, anorectal malformations include a different spectrum of anomalies in males than in females. In males, there may be anal atresia without fistula, anal atresia with recto-perineal, recto-urethral, recto-prostatic, or recto-vesical fistula. In females, there may be anal atresia without fistula, atresia with recto-perineal, recto-vulvar, recto-vaginal fistula, or cloacal anomaly. Frequently, patients with anorectal malformations will also have other associated anomalies: genital, renal, cryptorchidism, hypospadias, spinal, sacral, or cardiac anomalies. An important aspect in anorectal malformations is the degree of development of the anal sphincter complex. The sphincter is often underdeveloped and poorly innervated, and severity of this deficiency increases with the hight the the malformation.

The **diagnostic** is mainly clinical. The absence of the anus in the perineal region is noted, with or without an obvious fistula. The patient may pass meconium during urination in the case of recto-urethral or recto-vesical fistulas, or in the vagina in the case of anorectal fistulas.

The vital prognosis is good, while the functional prognosis is influenced by the degree of development and innervation of the anal sphincter. A large proportion of patients with anorectal malformations will develop anal incontinence.

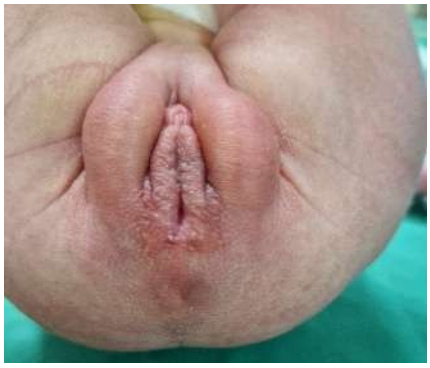


Fig. 13.1. Anal atresia without fistula

Paraclinical diagnostic. The most important imaging investigation is the invertogram (abdominal X-ray with the patient turned upside down), which allows to measure the distance between the end of the digestive tract (rectal pouch) and the perineal skin. This X-ray must be performed at least 24 hours after birth to allow enteric gases to reach the most distal area of the digestive tract. Depending on this distance, anorectal malformations are divided into high and low. Complementarily, for the diagnostic of associated renal-urinary, genital, cardiac, or spinal malformations, abdominal ultrasound, MRI, or CT with contrast medium may be performed.

Treatment. For low anorectal malformations, anorectoplasty (lowering the normal colon to the perineum in the middle of the anal sphincter) can be performed through a perineal approach during the neonatal period. For high anorectal malformations, as well as in patients whose health does not allow for extensive surgery, colostomy is performed during the neonatal period, with anorectoplasty to be performed later, when the patient's health allows (usually around 6 months of age).

Chapter 14. GASTROSCHIZIS

Definition. Gastroschisis (Laparoschisis) is a congenital malformation characterized by a defect in the anterior abdominal wall through which the abdominal viscera herniate freely, uncovered, outside of the abdominal cavity (Figure 14.1).

Etiopathogenesis. The abdominal wall defect occurs in utero because of a thrombotic accident in the right umbilical vein. Occlusion of this vein results in ischemia of corresponding abdominal wall segment, followed by necrosis and resorption. Another possible cause is the antenatal rupture of an omphalocele. Contributing factors include young maternal age (under 20 years), smoking, vasoconstrictive drugs, and toxic drugs.

Incidence is 1:2000-3000 newborns.

Pathological anatomy. In patients with laparoschisis, the defect in the abdo,inal wall is located paraumbilical, usually on the right side, and the exteriorized parenchymal organs are not covered by any membrane. The herniated intestinal loops are edematous, cardboard-like with multiple adhesions (Figure 14.1).

Fig. 14.1. Gastroschisis

The size of the defect and the volume of the viscera herniating through it vary. At the same time, due to the lack of content, the abdominal cavity has a smaller volume, and it is often difficult to completely reintegrate the herniated viscera. The intestine, sometimes the stomach, and exceptionally the liver herniates through the defect. The umbilicus is normally inserted into the abdominal wall. Laparoschisis is frequently associated with small intestine atresia.

Diagnostic. Prenatally, ultrasound in 18-20 weeks of gestation can reveal the defect. Alpha-fetoprotein is elevated. At birth, the diagnosis of gastroschisis is primarily clinical, made by simple inspection. The size of the defect as well as the quality and volume of the herniated intestinal loops are assessed.

Differential diagnostic. Laparoschisis can be confused with omphalocele with ruptured membranes.

Treatment. Patients with laparoschisis require emergency surgical treatment, which aims to reintegrate the intestinal loops into the abdominal cavity and repair the abdominal wall. The exposed intestinal loops cause significant loss of fluid, electrolyte, and heat, and the defect serves as a gateway for infections. At birth, the intestinal loops are immediately covered with sterile compresses soaked in warm saline solution and then protected with a waterproof membrane. A nasogastric tube is inserted, and intravenous fluid and electrolyte replacement and broad-spectrum antibiotic therapy are started. Patients may be placed in the lateral decubitus position, or the viscera will be kept above the abdomen to prevent stretching of the mesentery and volvulus of the loops.

The patient will be taken to the operating room as soon as possible. The surgical intervention consists of reintegrating the herniated intestinal

loops into the abdominal cavity, repairing the edges of the defect, and suturing the abdominal wall. Reintegration of herniated abdominal content is performed under careful intra-abdominal pressure monitoring. If the disproportion between the volume of the reintegrated viscera and the volume of the abdominal cavity is too great, abdominal compartment syndrome may occur. In this situation, the loops will be gradually reintegrated into the abdomen, first being placed in a protective silicone bag (Silobag). The contents of the bag will be gradually reduced through successive ligatures, and the abdominal wall will be closed once the viscera have been completely reduced. The silicone membrane is compressed manually daily (Figure 14.2). If intestinal atresia is associated with this condition, given the suffering of the intestinal loops, it is not recommended that intestinal atresia to be operated at the same time while the defect (gastroschisis) is closed. It is preferable to reintegrate the loops, close the defect, and treat the atresia surgically later (7-14 days) after the inflammation of the loops has subsided and the adhesions have been lysed.

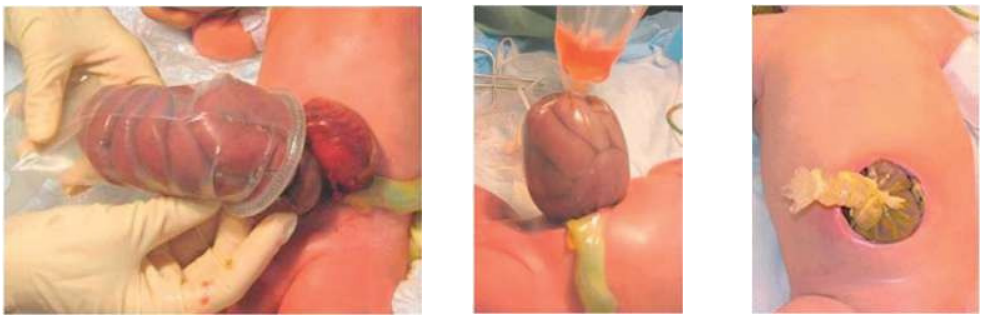


Fig. 12.2. Laparoschisis. Placement of loops in Silobag and staged reduction into the abdomen

Complications. Abdominal compartment syndrome is a serious complication that occurs when the hydrostatic pressure in the abdomen rises above 20 mm H2O. Infectious complications can also be serious.

Prognosis. The postoperative recovery period may be prolonged, requiring 14-21 days for the patient to resume bowel movements. Prognosis is mainly influenced by the timeliness of treatment, with infection being the most significant risk. Long-term outcomes are generally favorable.

Chapter 15. OMPHALOCELE

Definition. Omphalocele is a congenital defect of the anterior abdominal wall through which the abdominal viscera herniate, covered by an avascular, translucent membrane.

Incidence is 1–2:10,000 newborns.

Etiopathogenesis. The abdominal wall is formed by the union of four embryonic fold, one cephalic, one caudal, and two lateral, which converge ventrally and form the anterior abdominal wall. At the junction of the four folds is the umbilical ring, which surrounds the umbilical cord and the yolk sac. Omphalocele occurs when the lateral folds fail to close, and resulting in persistence of the intra-abdominal viscera within umbilical cord, and covered by a membranous sac derived from the yolk sac and peritoneam.

Pathological anatomy. The abdominal wall defect varies in size. The intra-abdominal organs covered by a translucent membrane protrude through the defect (Figure 15.1). The abdominal defect is covered by a transparent, avascular membrane consisting of three overlapping layers: the parietal peritoneum on the inside, Wharton's jelly, and the amniotic membrane on the outside. The umbilical cord inserts into this membrane. The contents of the sac can be seen through the transparency of the membrane. In addition to the intestines, the liver is usually also found in the sac. Depending on the size of the defect, omphalocele is classified as major (diameter > 5 cm) or minor (diameter < 5 cm).

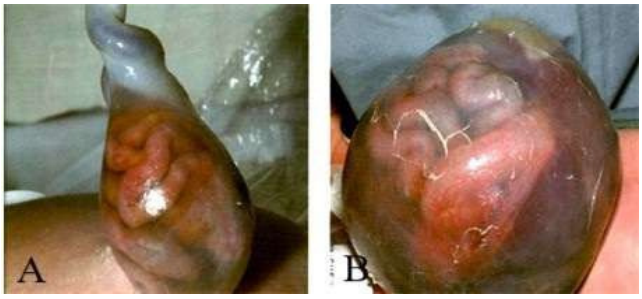


Fig. 15.1. Omphalocele. A Minor form; B Major form

Positive diagnosis. Prenatally, 2D ultrasonography can establish the diagnosis starting at 18 weeks of gestation. Alpha-fetoprotein may be elevated in the maternal blood. Postnatally, the diagnosis is clinically evident. Other congenital malformations (cardiac, gastrointestinal) or chromosomal abnormalities are frequently associated.

Differential diagnosis is made with umbilical hernia in small omphaloceles and with laparoschisis in the case of membrane rupture.

Treatment. If the diagnosis was made prenatally, a comprehensive evaluation for other associated malformations is recommended, as well as amniocentesis with evaluation for chromosomal abnormalities.

Postnatally, the newborn is placed in an incubator, and the membrane is covered with sterile compresses moistened with warm saline solution. The patient is evaluated for associated malformations (cardiac, renal, and central nervous system anomalies).

Surgical treatment of omphalocele is not a surgical emergency. The patient will be taken to the operating room only after being stabilized and fully evaluated. Visceral reintegration and primary closure of the defect is possible in cases of small defects. In large defects, staged closure is preferred to avoid abdominal compartment syndrome. Initially, the omphalocele

membrane is excised and the viscera are placed in a silicone bag (Silobag). Like the technique used in laparoschisis, the reintegration of the herniated viscera into the abdominal cavity will be done gradually, with closure of the abdominal wall undertaken only after the viscera have been completely reduced into the abdominal cavity. Alternatively, the technique of sequential excision of the omphalocele membrane (Fufezan procedure) can be used. The omphalocele membrane is only partially sectioned and then sutured to the edge of the defect. The procedure is repeated every few days until the viscera are successfully reintegrated.

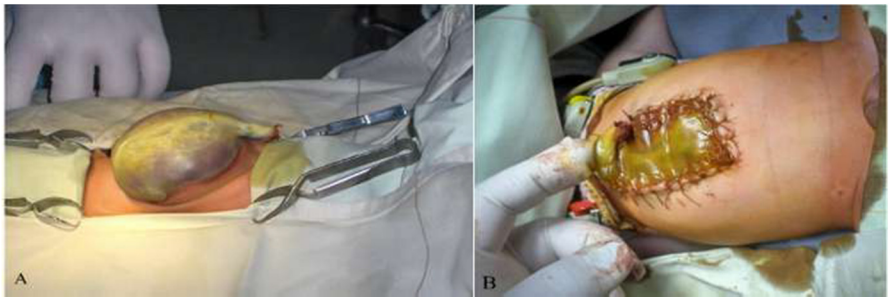


Fig. 15.2. Surgical treatment, Fufezan procedure

Conservative treatment is used when the omphalocele has intact membranes and the patient has other severe malformations that do not allow general anesthesia and surgery. Antiseptic substances are applied to the sac, which cause epithelialization of the omphalocele membrane, transforming the sac into a ventral hernia. The hernia can be repaired later, when the patient's condition allows it.

Possible **complications** include abdominal compartment syndrome, wound dehiscence, and infectious complications.

Prognosis. The vital prognosis is primarily influenced by associated malformations.

Chapter 16. UMBILICAL HERNIA

Definition. It is a defect in the closure of the fascial umbilical ring; where the abdominal consistis only of peritoneum and integument (skin) lacking the normal muscular and fascial layers (Figure 16.1).

Epidemiology. It is a common pathology in the African American pediatric population, among whom the incidence is approximately 50%.

Pathophysiology. With increased intra-abdominal pressure, the hernia becomes evident through the engagement of an intestinal loop or omentum through the defect. Most umbilical hernias close spontaneously during childhood, and the risk of incarceration is extremely low.

Clinical picture. Clinical symptoms are usually absent. A tumor-like formation is observed at the umbilicus, with variable dimensions depending on intra-abdominal pressure. The defect in the fascia is palpable after the swelling has subsided. Umbilical hernias usually close spontaneously by the age of 3-4 years.

The differential diagnosis includes diastasis of the rectus abdominis muscles or supraumbilical hernias.

Treatment is surgical if the hernia persists after the age of 4. If the defect is larger than 1.5 cm at the age of 2, spontaneous closure is unlikely. The pain caused by incarceration of the omentum is a rare indication for early surgery. Sometimes, repair of the primary defect will be accompanied by umbilicoplasty.

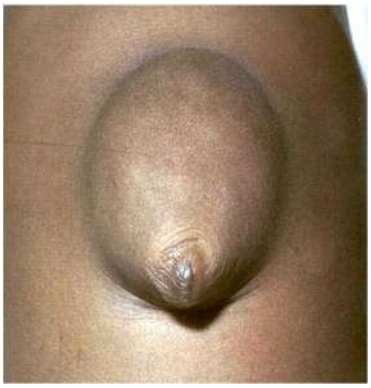


Fig. 16.1. Umbilical hernia

Prognosis is favorable.

Chapter 17. INGUINAL HERNIA and HYDROCELE

17.1. INGUINAL HERNIA

Definition. A hernia is defined as a defect in the abdominal wall through which an intra-abdominal organ (most commonly the omentum, small intestine, or large intestine) protrudes through a preformed opening.

The term inguinal hernia includes indirect inguinal hernia, direct hernia, and femoral hernias. Indirect inguinal hernias are lateral to the inferior epigastric vessels and are by far the most common in children. Even in young adults (16-18 years old), direct inguinal hernias are less common. Femoral hernias (below the inguinal ligament) account for less than 1% of pediatric inguinal hernias.

Incidence. Inguinal hernia is one of the most common pediatric surgical conditions. In preterm newborn children hernia may be evident at birth, with most cases diagnosed within the first month of life. It is more common in males, with male to female ratio of approximately 10:1. Approximately 60% of hernias are on the right and 10% are bilateral side in both males and females.

Etiopathogenesis. At the end of the third fetal month, the parietal peritoneum invaginates in front of the epigastric vessels, forming the peritoneovaginal canal that ends in the scrotum through the vaginal tunic. In males, the testicle and the elements of the spermatic cord adhere to the peritoneovaginal canal as they descend into the scrotum. In females, the vaginal process descends along the round ligament to the labial region (Nück canal). The involution of the peritoneovaginal canal ideally occurs in the last month of pregnancy. In some cases, the involution is much slower, so that in the first year of life the canal is still open or incompletely obliterated. The

absence of partial or total obliteration of the canal allows the intestine to engage, transforming into a hernial sac.

Clinical and paraclinical picture.

Inguinal hernia in males

In newborns and infants, it can occur either with the first cry, especially in premature babies, or due to the effort of coughing, difficult urination (meatal stenosis, tight phimosis), constipation, mechanisms explained by increased abdominal pressure. The canal widens and allows the engagement and progression of an abdominal loop into the inguinal region or scrotum if the inguinal orifice is greatly widened (Figs. 16.1, 16.2).

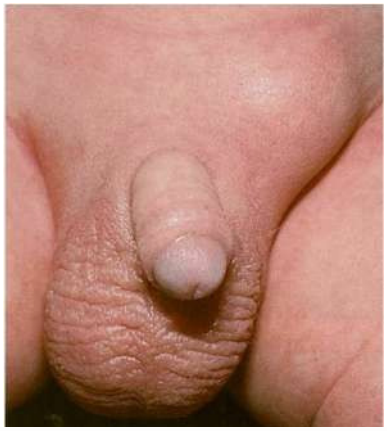


Fig. 17.1. Left inguinal hernia.

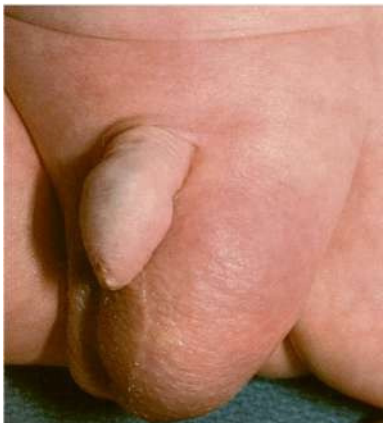


Fig. 17.2. Left inguinal-scrotal hernia.

The inguinal hernia becomes visible as a swelling of varying size, typically located just above the pubis at the external opening of the inguinal canal. In most cases, it reduces spontaneously except during exertion, and when it is large, it remains even during rest and distends the scrotum. If hernia reappears at rest after reduction, it is defined as an “incoercible hernia”. During reduction, the size of the external inguinal opening is assessed by inserting the little finger from the scrotum. In older children, there are fewer

symptoms, as the hernia is small and painless, visible only during exertion and coughing (except in cases of neglected, large, incoercible hernias). Physical examination reveals enlargement of the external opening of the inguinal canal on the side of the lesion. In all cases, the presence of the testicles in the scrotum must be checked.

Inguinal hernia in females

Less common than in males (ratio 10/1), rarely visible in infants, usually becomes visible after few years and is often bilateral. The intestine or ovary and fallopian tube may herniate through the patent peritoneovaginal canal (of Nück). The dimensions are smaller, and strangulation is rare and when it occurs, it typically involves the ovary, without vomiting or cessation of intestinal transit. Difficult reduction signifies involvement of the ovary or a Nück canal cyst like the funicular hydrocele in boys.

Strangulated inguinal hernia

This is a complication of an existing inguinal hernia. In infants, it may be the first sign of manifestation. Clinically, newborns present with pain, manifested by unusual crying and agitation, after short periods of calm that reappear either spontaneously or with movement, vomiting leading to dehydration and cessation of intestinal transit (a late sign, not to be expected). The inguinal tumor is the most important sign. It is round, firm in consistency, with reduced mobility, the upper pole is thicker and continues into the inguinal canal and is painful on palpation. In large, scrotal forms, the skin is stretched, shiny, and often purple. Sometimes it can reduce spontaneously during sleep, but more often it is irreducible.

Paraclinical picture. In the case of inguinal hernias, laboratory tests do not reveal any pathological changes, except in cases that are present late, with a picture of intestinal obstruction due to neglected strangulated inguinal

hernia. In these cases, a chest and abdominal radiography in an upright position is required to highlight hydro-air levels. Abdominal and scrotal ultrasound can confirm the diagnosis.

Differential diagnostic. In males, the differential diagnosis includes vaginal hydrocele and funicular hydrocele, which are common in infants but disappear within the first 6 months in most cases. Strangulated hernia must be differentiated from cord cyst which is irreducible, painless, and lacks signs of occlusion and agitation.

In girls, the differential diagnosis includes Nück's cord cyst (irreducible, elastic, mobile, painless).

Treatment. An inguinal hernia will not heal spontaneously; surgical treatment with ligation of the peritoneal-vaginal canal at its base is always indicated. Surgery can be classical or laparoscopic. Laparoscopy tends to be the preferred technique for more surgeons due to the advantage of contralateral exploration and repair of the defect.

Complications. Hematoma, scrotal edema, iatrogenic undescended testicle, recurrence, damage to the vas deferens, testicular atrophy, and intestinal damage are among the most common complications.

Prognosis. The progression is less severe than in adults; infants frequently experience repeated strangulation (which resolves spontaneously during sleep or transport) through a thinner, more elastic ring, which less often causes necrosis of the loop. Strangulation lasting longer than 24 hours poses a risk of compromising the intestinal loop. The prognosis is generally good.

17.2. HYDROCELE

Definition. Hydrocele is a cystic formation containing clear fluid content, located within a persistent remnant of the peritoneovaginal canal.

Etiopathogenesis. It is common with inguinal hernia.

Pathological anatomy. There are several forms of hydrocele, the most common being vaginal hydrocele, with fluid accumulation in the testicular vagina, the rest of the canal being closed, or communication with the peritoneum being preserved – communicating hydrocele. In many cases, they are associated with an indirect inguinal hernia. They are often bilateral and have a higher incidence on the right side. There are situations when they form between two closed Ramonède rings, resulting in a funicular hydrocele or cord cyst (in rare cases, it occurs in females – Nück's canal cyst).

Clinical and paraclinical picture. The tumor is painless, round-oval, smooth, and stretches the scrotal skin. It is elastic and fluid-filled, and the testicle is located at the lower pole. It is often bilateral, common in newborns after birth, and disappears spontaneously in most cases by the age of 6-12 months. In the case of communicating hydrocele, the size varies depending on physical exertion and is larger in the evening. In general, the communicating form does not heal. The cord cyst presents as a small, ovoid, mobile formation, without changes in volume, is painless and does not reduce. Scrotal ultrasound establishes the diagnosis with certainty.

Positive diagnosis is made based on the clinical picture and ultrasound. Differential diagnosis includes inguinal hernia, idiopathic scrotal edema, Morgagni hydatid torsion, orchiepididymitis, scrotal abscess, and testicular tumors.

Surgical **treatment** is required after the age of 1 year, and the approach is the same as for inguinal hernia. It is necessary to interrupt the continuity of the peritoneovaginal canal with the peritoneal cavity.

Complications. Edema, hematoma, and recurrence are the most common complications.

The **evolution and prognosis** are good.

Chapter 18. CRYPTORCHIDISM

Definition. It represents the delay or cessation of the process of testicular descent, occurring either along the normal migration path or outside it (ectopic). The current term found in the literature is undescended testicle. The variability of the nomenclature regarding the undescended testicle has led to difficulties in classifying testicular migration anomalies.

Pathological anatomy. The clearest classification divides testicles into palpable and non-palpable, with the obvious limitation that a non-palpable testicle may not be cryptorchid (undescended) but absent.

Depending on the position of the testicle, we distinguish the following situations:

- Testicular ectopia: the testicle is in an abnormal position relative to its normal descent path: subcutaneous, prepubic (root of the penis), perineal, in the crural region, in the scrotal bursa on the opposite side (crossed ectopia).
- Cryptorchidism: defines the non-palpable testicle located intra-abdominally and includes testicular agenesis (anorchidia); in the case of unilateral cryptorchidism, the term monorhidia may also be used.
- Testicular retention: defines a palpable testicle, stopped on its normal descent, which cannot be mobilized by traction at the level of the scrotum.
- Retractile testicle: a testicle located at the external opening of the inguinal canal that can be mobilized into the scrotum by palpation or traction but returns to its initial position when the maneuver is stopped.
- Floating testicle: the testicle is seen spontaneously, but intermittently, in the scrotal bursa.

Incidence. Cryptorchidism, or undescended testicles, occurs in approximately 3% of full-term male infants and up to 33–45% of premature infants. Most testicles descend within the first 6–12 months, so that by 1 year of age, the incidence drops to 1%. Descent of the testicle after 1 year is unlikely. In 20% of cases, cryptorchidism is bilateral. It is frequently associated with enuresis, hypospadias, or other genitourinary malformations.

Etiopathogenesis. The testicles form in the lumbar region during the mesonephros phase in the upper portion of the Wolffian duct. Migration to the final position (in the scrotum) occurs in two stages and is a hormone-dependent process in which the integrity of the hypothalamic-pituitary-testicular axis plays a key role. The development and descent of the testicles depend on close interaction between the endocrine and paracrine systems, growth factors, and mechanical factors. Any anomaly that disrupts normal testicular descent leads to cryptorchidism. The complexity of the process suggests that the etiopathogenesis of undescended testicles is multifactorial.

Clinical picture. On examination, the scrotum appears hypoplastic, with blurred folds, asymmetrically or completely retracted, without content (Fig. 18.1). Palpation is used to locate the undescended testicle and assess its volume, consistency, and mobility.

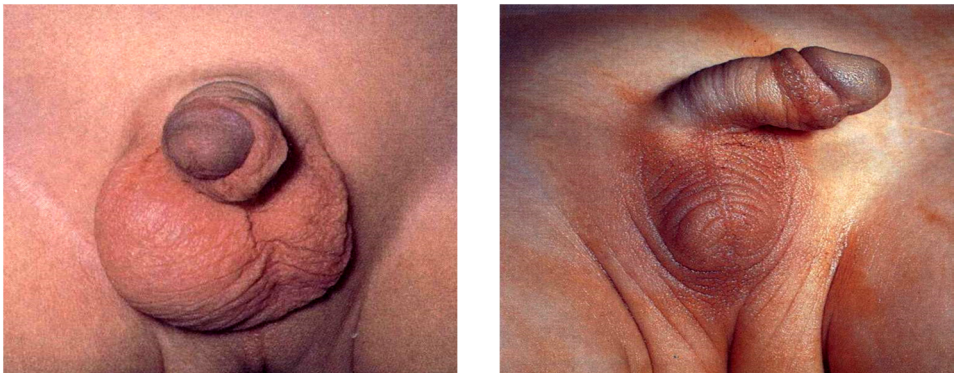


Fig. 18.1. Appearance of the scrotum in unilateral (left) and bilateral (right) undescended testicles.

If the testicle is not found in the inguinal region or in the possible areas of aberrant migration, there are two possibilities: anorchidia or intra-abdominal testicle.

Paraclinical picture. Imaging tests (ultrasound, MRI) are useful when the testicle absent from the scrotum, and it could not be detected by clinical examination. Exploratory laparoscopy is more reliable and can have both diagnostic and therapeutic value. Complementary tests may be performed: karyotype, sex chromatin, hormone dosages (17-ketosteroids).

Positive diagnostic. It is established based on clinical examination and paraclinical investigations.

Treatment. Due to the structural changes that occur early in the undescended testicle, ideally treatment should be instituted so that the testicle is brought into the scrotum by the age of 12-18 months,. Surgical treatment is the main form of therapy for undescended testicles. Primary descent through a combined inguinal and scrotal approach can be achieved if the testicle is palpable along the inguinal canal. If the testicle is not palpable, exploratory laparoscopy is preferred. Depending on the length of the spermatic vessels, the testicle is descended into the scrotum either in one or more stages.

Complications: painful cryptorchidism, torsion or orchitis of the cryptorchid or ectopic testicle, malignancy (10 times more common than in the normal testicle), psychological complexes, and sterility.

Prognosis. If not treated correctly and in a timely manner, cryptorchidism is the most common cause of male infertility. A reserved prognosis is found in patients with primary testicular dysplasia or intrabdominal testicles, or in those whose descent occurs during adolescence.

Chapter 19. ACUTE SCROTUM

Although most conditions encompassed by the term acute scrotum do not require surgical treatment, rapid recognition, prompt differentiation, and appropriate management are imperative due to the possibility of testicular torsion, which is a true surgical emergency with devastating consequences for the young patient in both the short and long term consequences.

Definition. Acute scrotum is found in several conditions with acute onset manifested by violent local pain, neurovegetative reflex phenomena, and local changes (edema, congestion, scrotal effusion). Of surgical interest are testicular torsion, Morgagni hydatid torsion, scrotal and testicular trauma, and idiopathic scrotal edema.

19.1. TESTICULAR TORSION

Definition. Testicular torsion involves twisting of the testis and spermatic cord around their axis, resulting in complete interruption of testicular blood circulation. The torsion is toward the midline of the body, with the usual site of torsion being intravaginal, rarely supravaginal.

Incidence. Although it is often feared, testicular torsion is not the most common cause of acute scrotum in the pediatric population. Testicular torsion has two peaks of increased incidence, in the neonatal period and in adolescents aged 13-16 years.

Etiopathogenesis. Anatomical abnormalities in the fixation of the testis within the scrotum are implicated: absence of the gubernaculum testis, an unfolded epididymis, testis suspended in the scrotum in a “bell-clapper" configuration. The precipitating factor for torsion is tyoically a sudden contraction of the cremasteric muscle.

Clinical and paraclinical picture. The onset of this condition is sudden, in otherwise healthy individuals, often during the night (background of erection and sudden contraction of the cremaster muscle), the dominant symptom being violent and continuous pain, localized in the affected hemiscrotum, radiating to the inguinal region and the root of the thigh, and exacerbated by movement. The pain gradually subsides 24-48 hours after onset, often folllowing necrosis of the affected testis. Associated symptoms include nausea, vomiting, paralytic ileus, pollakiuria-dysuria, and bladder tenesmus.

On local examination, the affected hemiscrotum is enlarged, edematous, reddish-purple. Palpation exacerbates the pain and reveals an enlarged testis that is elevated towards the upper part of the scrotum. The spermatic cord is hardened, thickened, and tender.

When faced with a patient presenting scrotal pain accompanied by the cardinal signs of inflammation, the new guidelines require the TWIST score to be calculated (Table 19.1). This is a clinical score updated in 2019 that predicts the risk of testicular torsion in both adults and children (for a score higher than 3, the guidelines recommend performing a scrotal ultrasound).

Table 19.1. Calculation of the TWIST score.

Prezența edemului testicular	2 pct
Testicul dur la palpare	2 pct
Absența reflexului cremasteric	1 pct
Pozitia aberantă a testiculuuui "high-ride testicle"	1 pct
Greață/Vărsături	1 pct

Scrotal ultrasound is a tool for both positive and differential diagnosis. Associated laboratory investigations are complete blood count and urinalysis with urine culture.

Positive diagnosis is easily made through medical history, clinical presentation, and laboratory tests.

Differential diagnosis. As this is a broad pathology, a differential diagnosis with the following conditions is required: orchitis and epididymitis, strangulated inguinal hernia (swelling extends into the inguinal canal, signs of intestinal obstruction), torsion of the cryptorchid or ectopic testicle (palpation of the scrotal sac on the affected side reveals the absence of the testicle)

Treatment. If testicular torsion is suspected but surgery cannot be performed at that time, manual detorsioning (Nash in 1893) can be attempted—a technique of twisting from medial to lateral like an "open book." A recent study published in 2023 concluded that this maneuver is beneficial in patients with testicular torsion unaccompanied by edema who present within the first 6 hours of symptom onset. Their conclusions also showed that it is beneficial to attempt this maneuver in all patients with torsion before definitive surgical treatment, due to the improvement in blood flow (demonstrated by Doppler ultrasound).

The definitive and accepted treatment is surgical: exploration, detorsion, ipsilateral and contralateral orchidopexy, or orchiectomy (if the testis is nonviable after detorsion) (Figure 19.1).

Complications include scrotal hematoma, surgical wound infection, and suture granuloma.

Prognosis: With prompt treatment within the first 6 hours of onset, the progression and prognosis are good in both the short and long term, with

rapid reintegration into the social environment. Delayed diagnostic and treatment lead to impaired quality of life for men due to loss of the affected testicle.

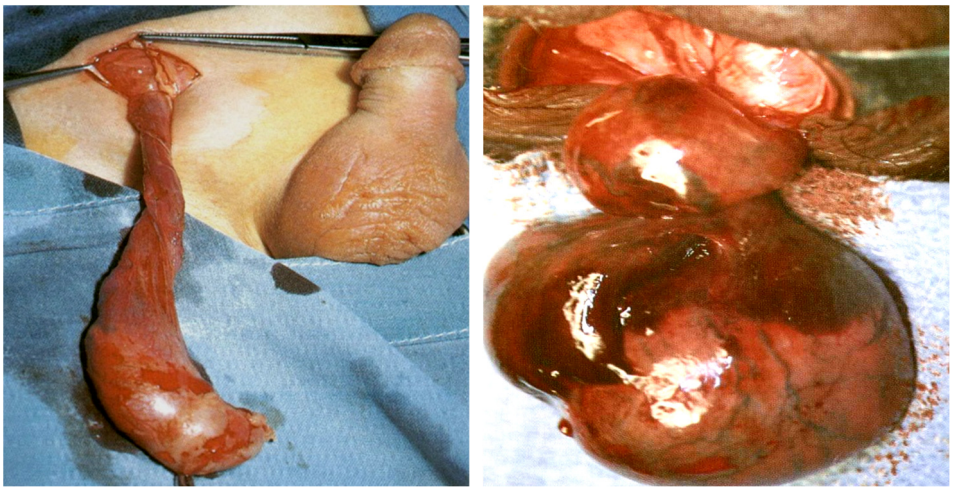


Fig. 19.1. Testicular torsion – intraoperative aspects.

19.2. TORSION of HYDATID OF MORGAGNI

Definition. It represents the torsion around the vascular pedicle of a vestigial appendage located at the upper pole of the testis, causing acute scrotal pain. Torsion of the hydatid of Morgagni can mimic the same clinical picture as testicular torsion.

Incidence. Torsion of the hydatid of Morgagni occurs across the same age group as testicular torsion but is considerably more frequent.

Etiopathogenesis. It is not clear, but long vascular pedicles of hydatids and local trauma are implicated.

Clinical and paraclinical picture. The pain is sharp, located at the upper pole of the scrotal sac, with less pronounced vegetative phenomena, a testicle of normal size that is painful at the upper pole, where the painful

hydatid can be palpated, and a cyanotic tumor formation – "blue dot" – visible through transillumination. The testis and spermatic cord are intact. Scrotal ultrasound identifies the lesion.

Positive diagnosis. It is made based on medical history, clinical presentation, and scrotal ultrasound.

Differential diagnosis includes testicular torsion, whose symptoms mimic those of hydrocele.

Treatment may be conservative, supportive, or surgical in cases of uncertainty or late presentation, consisting of scrotal exploration, pedicle ligation, and hydatid excision.

Complications include scrotal hematoma, surgical wound infection, and suture granuloma.

Prognosis is good, without compromising testicular function.

19.3. SCROTAL AND TESTICULAR TRAUMA

Etiopathogenesis and incidence. These are rare, with an incidence of < 1%. The most common traumas include accidents during playing (cycling, horse riding, water sports, etc.), trauma due to blunt objects, or penetrating trauma.

Clinical and paraclinical picture. Upon presentation, patients show pain, erythema, edema, and traumatic marks (ecchymosis/hematoma/rupture at the scrotal or testicular level, active bleeding). An ultrasound evaluation is required to identify testicular damage.

Treatment. Depending on the clinical and ultrasound findings, treatment may be conservative, supportive, or surgical, consisting of exploration, hematoma evacuation, hemostasis, suturing, or orchiectomy.

19.4. IDIOPATHIC SCROTAL EDEMA

Incidence. Higher frequency in children compared to adults, with a peak at the age of 5-7 years.

Clinical and paraclinical picture. Clinically, patients present with pain/edema/erythema/pruritus, but more specifically with edema and erythema with insidious onset in the inguinal canal/perineum, migrating towards the affected hemiscrotum.

Diagnosis is one of exclusion based on medical history, clinical presentation, and paraclinical investigations.

Treatment is supportive (analgesic/ anti-inflammatory/ antihistamine/ local hydrocortisone applications).

Prognosis. The pathology is self-limiting, resolving in 3-5 days with appropriate treatment.

Chapter 20. VARICOCELE

Definition Varicocele is a phlebopathy, characterized by varicose dilation of the left spermatic venous plexus.

Incidence. The incidence is 4-10% in the male population, usually occurring after the age of 10. If it is left untreated, varicocele is a major cause of male infertility.

Etiopathogenesis. Varicocele occurs due to stasis/reflux of blood in the venous system of the left testis. It is idiopathic in 99% of cases and occurs on the left side. Exceptionally, it occurs on the right side when it is secondary to stasis in the spermatic vein due to large abdominal tumors.

The left spermatic vein drains into the left renal vein, unlike the right one, which empties directly into the inferior vena cava,. There are several factors that can contribute to this stasis:

- Passage of the left renal vein through the aorto-mesenteric clamp (aorto-mesenteric angle < 23°)
- Constitutional predisposition to varicose disease (frequently associated with varicose veins of the lower limbs)
- The great length of the left spermatic vein
- Right-angle junction of the left spermatic vein into the renal vein
- Absence of one-way valves in the left spermatic vein
- Spasm of the distal segment of the spermatic vein due to reflux of adrenaline-containing blood from the adrenal gland
- Decrease in the "vasa vasorum" vascular complex in the structure of the left spermatic vein

Pathological anatomy. Macroscopically, the spermatic veins are dilated, the spermatic cord is thickened, and the left gonad is smaller in size

and consistency. Microscopically, alterations of the seminiferous tubules are detected, decreased seminiferous tubule density, and morpho-pathological changes such as interstitial fibrosis, edema, and hyalinosis. There may also be changes in the seminiferous line, cessation of maturation at various stages of development, an increase in the number of Leydig cells relative to the fibrous tissue in the interstitial spaces, an increase in the number of Sertoli cells, and marked interstitial stasis.

Clinical picture. The usual age of onset is 10-12 years. Varicocele develops slowly and insidiously and becomes evident at puberty. On clinical examination, the left scrotal sac is elongated, with blurred, moist folds, and the local temperature is 1-2° higher than that of the right hemiscrotum (increased moisture occurs through perspiration and chronic stasis). On palpation, thickening of the left spermatic cord ("worm ball") is detected, and on the Valsalva maneuver, the left testicle appears smaller and soft in consistency (in long-term varicocele carriers).

Varicocele has three stages of development

I. Varicocele evident only after the Valsalva maneuver
II. Varicocele palpable without Valsalva
III. Varicocele evident on scrotal inspection

Paraclinical picture. Varicocele can be detected by ultrasound. Abdominal ultrasound is necessary for the differential diagnosis of abdominal tumors. Spermatic phlebography may be useful to highlight the number and caliber of dilated veins. Other tests may also be performed: spermogram, testicular biopsy, or hormone tests.

Treatment is absolutely indicated in any stage of varicocele development. Surgical treatment involves ligation of the dilated spermatic vein(s), which stops venous reflux. This can be done through an open inguinal

approach (Ivanisevich technique) or a laparoscopic approach (Palomo). Alternatively, the spermatic vein can be sclerosed using interventional radiology techniques, or an anastomosis of the spermatic vein to the inferior epigastric vein can be performed (Belgrano).

Prognosis: Postoperatively, transient scrotal edema, testicular atrophy, persistent or recurrent varicocele may occur. Persistence of the varicocele (improperly called recurrence) occurs in 5-10% of cases and is most often due to failure to recognize the anatomical particularities of the cases (multiple spermatic veins). The prognosis is favorable if the surgery is performed in a timely manner.

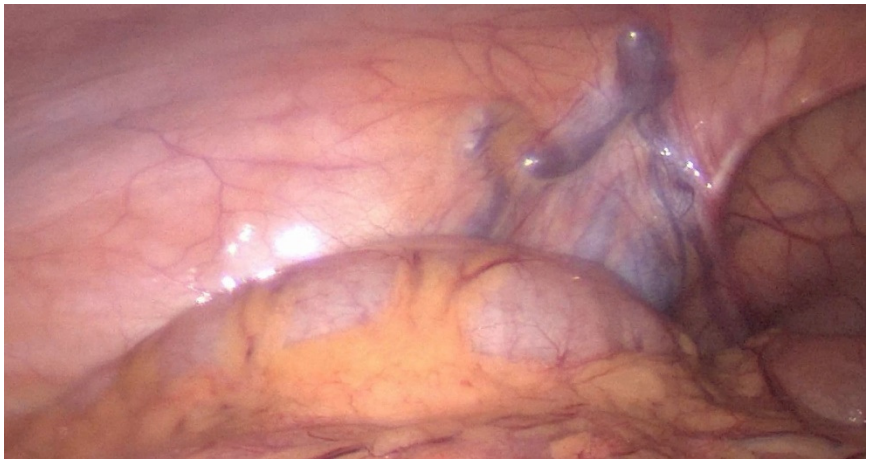


Fig. 20.1. Varicocele, dilated spermatic vein, laparoscopic view

Chapter 21. OVARIAN TORSION

Definition. It represents the twisting of the ovary around the vascular axis, with sudden interruption of blood supply, leading to ischemia and subsequent necrosis. Both the ovaries and fallopian tubes can occasionally twist, constituting a surgical emergency.

Incidence is not fully known. In general, there is a tendency for this pathology to occur in girls during puberty, but it can be found in all age groups.

The **etiopathogenesis** of ovarian torsion is not fully understood. An increase in ovarian volume associated with an ovarian cyst or tumor in the adnexa is a contributing factor. However, torsion can also occur in a normal ovary. Torsion occurs when the ovary twists around its supporting ligaments resulting in obstruction of blood flow. Initially, venous outflow is obstructed, then arterial inflow is interrupted, leading to ovarian necrosis (Figure 21.1), infarction, hemorrhage with hemoperitoneum, and potentially peritonitis.

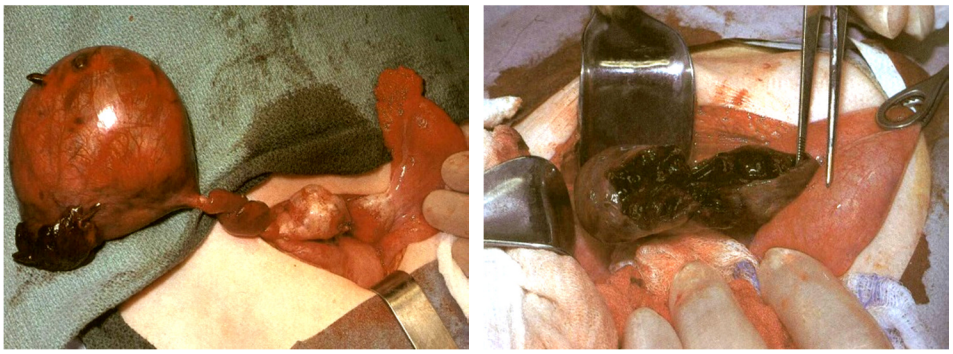


Fig. 21.1. Ovarian torsion

Clinical and paraclinical picture. The cardinal sign in ovarian torsion is pain. The pain is sudden, severe, and may be syncopal in nature. It

is typically located in the small pelvis, hypogastrium, or iliac fossa, and may radiate to the flanks, groin, or abdomen. The patient presents with adistressed facial expression, tachycardia, and adopts antalgic position in lateral decubitus on the affected side. Associated symptoms may include gastro-intestinal manifestations (nausea, vomiting, constipation), and urinary symptoms (pollakiuria-dysuria, bladder tenesmus). On palpation, there is marked tenderness in the lower abdomen, accompanied by reflex muscle guarding. Rectal examination may reveal painful thickening of the pouch of Douglas with palpable adnexal or the tumor formation.

Paraclinical picture. Laboratory investigations have little relevance for diagnosis. Sometimes leukocytosis with neutrophilia can be detected. Abdominal ultrasound is the imaging method used to diagnose adnexal torsion. If an adnexal tumor is identified on ultrasound, ovarian tumor markers CA125, HE 4 (ROMA score), ACE (carcinoembryonic antigen), alpha-fetoprotein, and Beta HCG should be measured. Computed tomography may be indicated if malignancy is suspected.

Positive diagnosis is based on the clinical picture associated with abdominal ultrasound, which detects the absence of ovarian vascularization and/or the presence of a pre-existing pathology (cysts, tumors, etc.). Ultrasound is essential for the diagnosis of torsion.

Differential diagnosis includes acute appendicitis, renal colic, salpingitis, ectopic pregnancy, ruptured follicular cyst, pelvic inflammatory disease.

Treatment. Ovarian or fallopian tube torsion is a surgical emergency and requires prompt surgical intervention, which aims to preserve the ovary. Depending on its viability, the following procedures are performed: detorsion, salpingectomy, oophorectomy, adnexectomy. Some authors also

recommend fixation of the contralateral ovary. Possible complications include intraoperative or postoperative hemorrhage, reactive peritonitis, pelviperitonitis, surgical wound infection, and wall abscess.

Prognosis. The progression varies depending on the underlying cause. For patients whose ovarian torsion was caused by a tumor, the short-term evolution is good, but a series of anatomo-pathological and histological investigations will follow to determine the type of tumor, its aggressiveness, and the required treatment. Patients who experience torsion spontaneously in an otherwise healthy ovary generally have a good outcome, with rapid recovery and return to normal activities.

Chapter 22. INTUSSUCEPTION

Definition. Intussusception is a form of intestinal obstruction in which a segment of intestine telescopes into an adjacent distal segment resulting mechanical blockage.

Epidemiology. The incidence varies, averaging 2 cases per 1,000 infants. Over 70% of intussusception cases occur between 4 and 10 months of age, usually in eutrophic infants. Males are more commonly affected.

Etiopathogenesis. Intussusceptions are divided into two categories:

I. Primary intussusceptions, which have no obvious starting point. This is the most common form and occurs predominantly in infants. The determining cause is increased intestinal peristalsis (hyperperistalsis) caused by the sensitization of the mesenteric ganglions with bacterial toxins, viruses, plant allergens, etc. Contributing causes may include:
   - Abnormal growth of the cecum compared to the small bowel
   - Diversification of diet, favoring increased peristalsis
   - Lack of attachment of the ascending cecum-colon to the posterior parietal peritoneum
   - Seasonal viruses cause enterocolitis with mesenteric adenopathy and increased intestinal peristalsis

II. Secondary intussusceptions that originate from intestinal tumors, polyps, Meckel's diverticulum, intestinal adhesions, a foreign body, or duplications of the digestive tract. It occurs more frequently in older children.

Pathological anatomy. The intussusception tumor is composed of three concentric layers: innermost layer - the invaginated loop (intussuscepted), middle, and outermost layer - the receiving (intussuscipiens) loop. The intussusception has a head and a ring.

Based on the mechanism of production, three types of intussusceptions are described:

1. By prolapse: the head is mobile, and the intussusception ring is fixed, the inner loop progresses through the ring
2. By overturning: the ring is mobile, and the head is fixed
3. Mixed intussusceptions – through both prolapse and inversion

From a topographical point of view, there are several possible variants of intussusception (Figure 22.1), the most common form being ileo-ceco-colic intussusception.

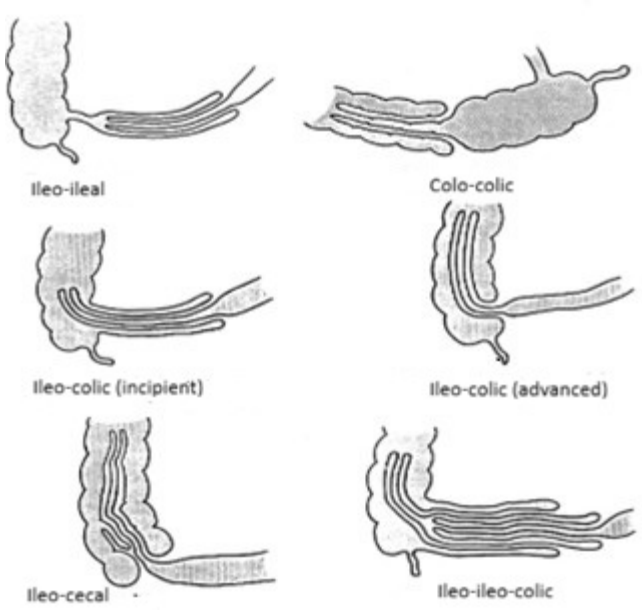


Fig. 22.1. Topographical variants of intestinal intussusception

Pathophysiology. As the intussusception advances into the intestine and mesentery, serious vascular and nerve damage occurs. The elongation of the nerve filaments causes abdominal pain, pallor, agitation, and vomiting. Initially, venous and lymphatic circulation is affected, with stasis in the capillaries of the submucosa of the invaginated loop (turgid, edematous loop) and bleeding into the lumen of the receiving loop. Subsequently, impaired arterial circulation leads to necrosis of the loop due to hypoxia.

Clinical picture. The characteristic triad of symptoms of intussusception consist of abdominal pain, vomiting, and rectal bleeding. Typically, symptoms begin as paroxysmal abdominal colic in a healthy individual, lasting 1- 3 minutes, followed by a period of calm lasting 10-15 minutes. During the painful episode, the patient is agitated, cries, adopts an antalgic position, with the knees flexed on the abdomen, and refuses food. Between painful episodes, the patient initially presents with good general condition but later becomes lethargic. The crises recur as the intussusception progresses. Vomiting occurs from the onset of the symptoms, initially as a neurovegetative reflex to pain, and only after 12-14 hours due to the mechanical occlusion. Initially is non-bilious, later becoming bilious. Stools have a characteristic "red currant jelly" appearance, containing mixture of mucus and blood. Palpation of the abdomen may reveal an empty right iliac fossa (Dance's sign).

Without prompt treatment, the infant's general state deteriorates within a few hours, the abdomen becomes distended, and rectal bleeding occurs. Vomiting becomes more severe, bilious and then fecaloid. Signs of peritonitis may appear due to necrosis and perforation of the invaginated bowel loop.

Paraclinical picture. Abdominal ultrasound (Figure 22.2) may reveal a central hyperechoic area surrounded by a hypoechoic image corresponding to the invaginated segment (donut or target sign). Plain abdominal radiography may reveal air-fluid images. Irrigography shows a sudden stop of the contrast medium at the head of the invagination, with profile images (lateral views) rsembling a "cup" and frontal images (view) showinging concentric rings ("target sign") (Figure 22.3).

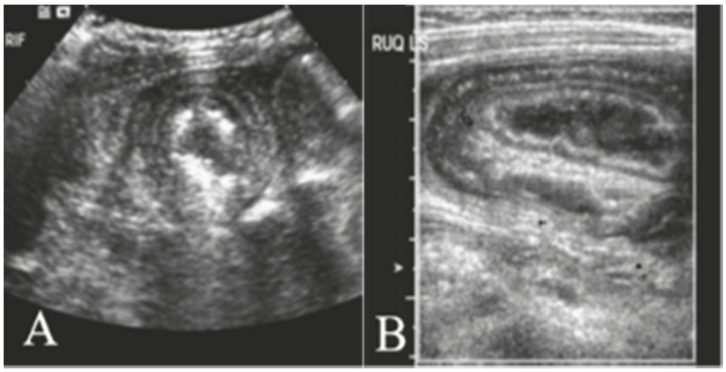


Fig. 22.2. Ultrasound images. A cross-section (target);
B longitudinal section (kidney-like)

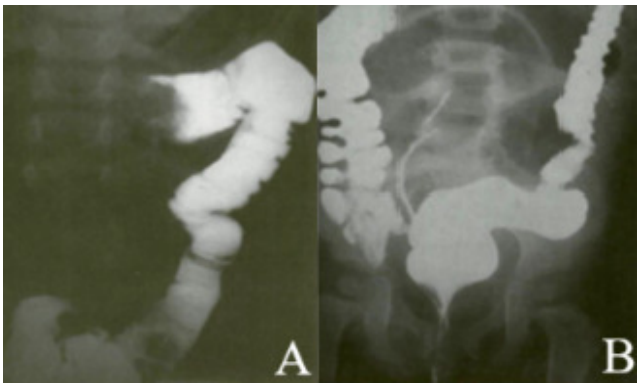


Fig. 22.3. Barium enema. A. Stop of contrast,
B. After the reduction of the invagination

Differential diagnosis. It includes acute enterocolitis, Meckel's diverticulum, acute appendicitis, Henoch-Schonlein purpura, and recto-colonic polyposis.

Treatment can be conservative or surgical.

Conservative treatment involves reducing intussusception by hydrostatic pressure. Under radioscopic control, a contrast medium is introduced into the anus and its progression through the colon is monitored (Fig. 22.3). The intussusception has been reduced when the contour of the cecum is visible, and contrast is visible in terminal ileum (Figure 22.3 B).

The procedure can only be performed under certain conditions:

- Interval from onset less than 24 hours
- Do not apply excessive pressure by raising the irrigator more than 1 m above the radiological table
- Do not insist manually by exerting high pressure on the colon to advance the fluid on the colonic frame
- In case of perforation of the colon wall, surgical intervention must be possible as soon as possible
- The contrast medium can cause dehydration, and fluid loss must be compensated for.
- Ileo-ileal intussusception cannot be reduced conservatively; the fluid instilled into the colon does not reach this level
- If reduction has not been achieved after 3 attempts, surgical treatment is indicated

Surgical treatment is performed in case conservative treatment failure or as a first-line treatment in case of late presentation. It is performed under general anesthesia and consists of manually reducing the intussusception by "squeezing" from downstream to upstream (not by traction). If the involved

bowel loop is devitalized, intestinal resection is performed up to the viable loop and end-to-end anastomosis. When the intussusception is very old, in children with a serious general condition, ileostomy or colostomy is recommended to relieve the patient of the occlusive syndrome as quickly as possible. The approach can be managed surgically via either laparotomy or laparoscopy.

Prognosis depends on the timeliness of diagnosis and treatment. If intervention occurs within the first 24 hours, the cure rate is 75-90%, whereas the mortality rate is up to 70-80% if the diagnosis is made after 3 days. The recurrence rate of intussusception is 5-8%.

Chapter 23. MECONIUM ILEUS

Definition. Meconium ileus is an obstruction of the small intestine due to the presence of abnormal, viscous, adherent meconium in the terminal ileum, and is the earliest manifestation of cystic fibrosis.

Epidemiology. It accounts for 9–33% of neonatal intestinal obstructions and is the third leading cause of neonatal intestinal obstruction after duodenal and intestinal atresia. Sixteen percent of patients with cystic fibrosis have a history of meconium ileus at birth. Complicated forms occur in 40% of patients with meconium ileus.

Etiopathogenesis. In most cases, meconium ileus is the first manifestation of cystic fibrosis (mucoviscidosis). Cystic fibrosis is an autosomal recessive disease in which the mechanism of the transmembrane chloride transporter is disrupted. This causes increased mucus viscosity in various structures of the body (intestine, pancreas, lungs). In cystic fibrosis, viscous meconium adheres to the intestinal wall, causing obstruction. This may lead to ischemia due to compression of the bowel wall, resulring in dilation, and in severe cases, intestinal perforation.

Pathological anatomy. The terminal ileum is filled with gray meconium concretions, arranged like beads on a string. Proximal to this, the intestine contains semi-liquid meconium and is imarkedly dilated with thin walls. Distal to the obstruction, a microcolon is typically observed.

Positive diagnosis can be made prenatally by fetal ultrasound in 15-20 weeks of gestation. A hyperechoic mass, impacted meconium, distended intestinal loops, non-visualization of the gallbladder, and polyhydramnios are observed.

At birth, meconium ileus can be present in two forms: simple and complicated meconium ileus.

- Simple meconium ileus: the newborn appears healthy but will not pass meconium in the first 24-48 hours. The clinical picture begins to take shape. Intestinal obstruction with progressive abdominal distension and bilious vomiting. Rectal examination reveals narrowing of the anal canal and rectum.
- Complicated meconium ileus. The patient shows signs and symptoms of the disease immediately after birth. Complications of meconium ileus include perforation and meconium peritonitis, which can be fibro-adhesive/plastic, meconium pseudocyst, meconium ascites, or infected meconium peritonitis.

Paraclinical picture. Plain abdominal radiography in orthostatic position may reveal large air images (not air fluid levels), "soap bubble" images in the right iliac fossa (Figure 23.1). In complicated ileus, the abdominal X-ray may show ascites, cystic mass, peritoneal calcifications, pneumoperitoneum. Irrigography with water-soluble contrast medium reveals microcolon and contrast medium stop at the ileocecal valve. It can also have a therapeutic role by dislodging meconium from the intestinal wall.

Other necessary investigations include sweat test or genetic testing for cystic fibrosis.

Differential diagnosis is made with intestinal atresia, Hirschsprung's disease, left microcolon syndrome, meconium plug syndrome.

Treatment. In uncomplicated ileus, treatment is conservative and non- surgical. This treatment consists of hydro-electrolytic rebalancing, gastric decompression with a nasogastric tube, and irrigation with a water-soluble contrast medium, usually followed by rapid elimination of meconium in the form of pellets.

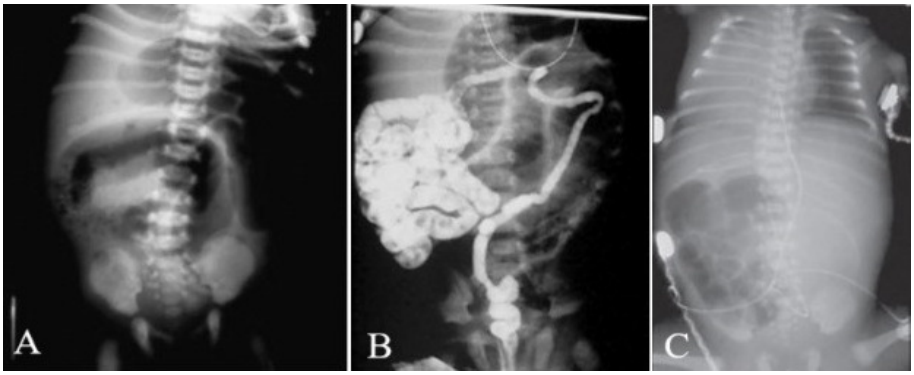


Fig. 23.1. A: A: Plain abdominal X-ray, large air images.
B: Irrigography, microcolon; C: Meconium pseudocyst with calcifications

Surgical treatment is indicated in complicated ileus and if meconium is not eliminated within the first 48 hours or after 2 enemas with contrast medium. Laparotomy is performed to inspect the entire intestinal tract, with several intraoperative techniques available for evacuation of meconium. The surgeon can "milk" the meconium anterograde towards the colon or retrograde towards the stomach or extract the meconium and irrigate the intestine through an enterotomy. If a segment of the intestine is not viable, resection with end- to- end anastomosis or resection with protective ileostomy can be performed.

In addition to surgical treatment, postoperative management is very important. This includes antibiotic therapy, continued enemas to ensure complete evacuation, correction of fluid and electrolyte imbalances, instillation of 2-4% N-acetylcysteine through a nasogastric tube to liquify the meconium, and dietary supplementation with pancreatic enzymes and vitamins. Closure of the stomas is recommended after 4-6 weeks.

Prognosis for patients is determined by the presence of complications of meconium ileus and cystic fibrosis, particularly respiratory complications.

Chapter 24. ACUTE APPENDICITIS

Definition. Acute appendicitis is an acute inflammation of the vermiform appendix and is the most common abdominal surgical emergency in children.

Etiopathogenesis. The appendicular lumen is obstructed by inflammation of the mucosa, coprolites, foreign bodies, scars, or adhesions. This leads to stasis and infection in the lumen of the appendix, distension with stretching and thinning of the wall. Subsequently, circulatory comprise occur in the wall, leading to perforation.

Pathological anatomy. Depending on the nature of pathological injury, acute appendicitis goes through several stages of development:

- Simple acute appendicitis (congestive or catarrhal)
- Phlegmonous appendicitis (suppurative or purulent)
- Gangrenous appendicitis with perforation and secondary peritonitis

Clinical picture. Acute appendicitis is characterized by Dieulafoy's triad of symptoms: point abdominal tenderness, muscle guarding and skin hyperesthesia. Pain is the main complaint, initially localized around the navel or in the epigastric region, and later migrating to the right iliac fossa. Muscle guarding and skin hyperesthesia accompany the pain located in the right iliac fossa. The patient also experiences loss of appetite, nausea, vomiting, and constipation, but these usually begin after the onset of pain. In advanced stages, when appendiceal perforation and secondary peritonitis have occurred, the general condition is affected, fever and abdominal contracture appear. Diarrhea may occur as a sign of peritoneal irritation.

Paraclinical picture. Laboratory tests show leukocytosis with granulocytosis (between 8000-15000 leukocytes/mmc), inflammation tests

(ESR, fibrinogen, CRP) are positive. Abdominal ultrasound may reveal an inflamed appendix (Figure 24.1). In appendicitis complicated by peritonitis, leukocytes usually increase to over 20,000/mmc, signs of dehydration and ion imbalances appear, and ultrasound reveals the presence of fluid in the peritoneal cavity.

Differential diagnosis includes any medical or surgical condition that produces a clinical picture of acute abdomen. The most common conditions are mesenteric lymphadenitis, renal colic, acute gastroenteritis and enterocolitis, Henoch-Schönlein rheumatoid purpura, intestinal parasitosis, acute primary peritonitis, urinary tract infections and pyelonephritis, Meckel's diverticulitis, intussusception, volvulus, and gastro-duodenal ulcer. Differential diagnosis is sometimes difficult, especially in newborns and infants.

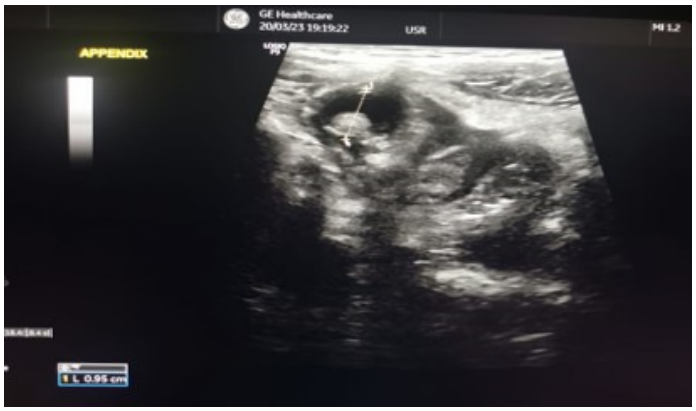


Fig. 24.1. Acute appendicitis. Appendix with dilated lumen and coprolite

Non-surgical treatment can be effective in uncomplicated acute appendicitis detected at onset. It consists of parenteral administration of broad- spectrum antibiotics for a period of 7-10 days with regular monitoring of clinical and paraclinical parameters.

Surgical treatment. Appendectomy can be performed using an open or laparoscopic approach (Figure 24.2). Regardless of the approach, the steps of the procedure are the same: the appendix is identified and ligated at its base, the mesoappendix is ligated, and then the appendix is sectioned and removed. Additionally, a bursa can be created around the base of the appendix, and the appendicular stump can be plugged. In cases of appendicitis complicated by peritonitis, purulent fluid evacuation, lavage, and drainage of the peritoneal cavity are also performed.

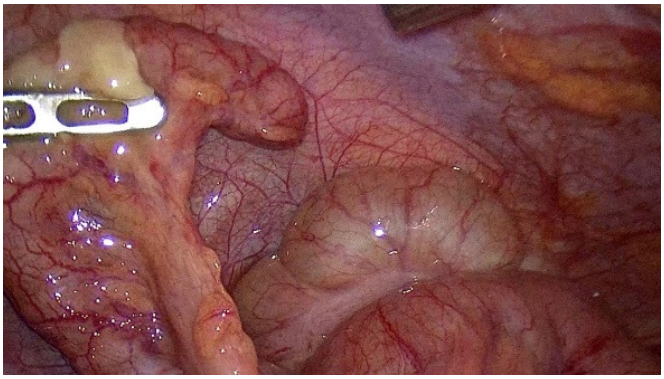


Fig. 24.2. Inflamed appendix with fibrinopurulent exudate on the serosa (laparoscopy)

Postoperative **complications**: hemorrhage due to slippage of the mesoappendix ligature, secondary peritonitis or peritoneal abscess, wall abscess, intestinal obstruction due to adhesions, septic shock.

Prognosis. Untreated acute appendicitis in children progresses rapidly, with perforation often occurring within 24-36 hours. In most cases, postoperative progression is favorable. With correct and timely treatment, the prognosis is favorable, with mortality below 1%.

Chapter 25. MECKEL'S DIVERTICULUM

Definition. Meckel's diverticulum is a malformation of the small bowel caused by the incomplete regression of the omphalomesenteric duct (vitelline duct), the embryonic structure through which the intestine communicates with the yolk sac.

Incidence is approximately 2%, with a male to female ratio of approximately 2:1.

Embryology. In the first weeks of intrauterine life, the midgut communicates with the yolk through (located outside the embryo) the omphalomesenteric duct. The omphalomesenteric duct lies between the elements of the umbilical cord. From the fifth week of intrauterine life, the duct undergoes a process of obliteration and involution starting from the umbilical end towards the intestinal end. The process ends after the seventh week. Incomplete resorption of omphalomesenteric duct led to the persistence of various abnormal structures, including Meckel's diverticulum.

Pathological anatomy. Meckel's diverticulum is finger-glove-shaped structure, typically located at the antimesenteric margin of the ileum, approximately one meter proximal to the ileocecal valve (Figure 25.1). In 50% of cases, the mucosa of Meckel's diverticulum shows various cellular heterotopies like gastric, pancreatic, duodenal, colonic, or endometrial cells. Their presence increases the risk of developing neoplasms originating at this level: carcinoid tumor, adenocarcinoma, gastrointestinal stromal tumor, and lymphoma.

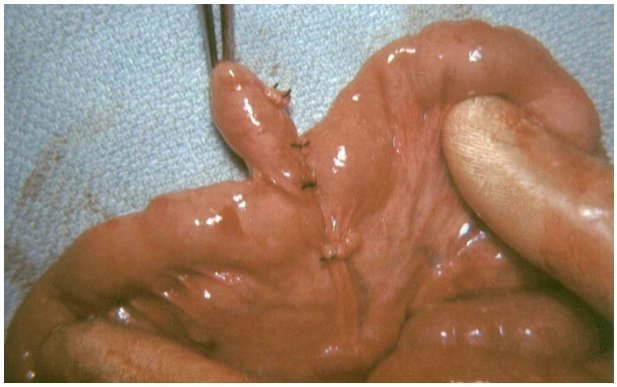


Fig. 25.1. Meckel's diverticulum

Clinical picture. Only 2-4% of people with Meckel's diverticulum develop symptoms during their lifetime, and in approximately 50% of these cases, symptoms appear within the first 2 years of life. In children, clinical manifestations are due to complications: hemorrhage, perforation, intestinal obstruction, or inflammation of the diverticulum (Meckel's diverticulitis). Hemorrhage is the most common complication and occurs following a mucosal ulcer that perforates a blood vessel. This area is usually located at the border between normal mucosa and the mucosa of the diverticula containing heterotopic gastric cells (secreting HCl). It is a type of lower gastrointestinal bleeding and has several characteristics features: the blood is partially digested (hematochezia), the bleeding is typically painless, and it maybe episodic or continuous but slow, allowing significant blood loss before clinical signs become apparent. Perforation, like bleeding, occurs due to ulceration of the diverticulum mucosa. This occurs under the action of hydrochloric acid produced by heterotopic gastric-type cells present in Meckel's diverticulum. The clinical picture is that of secondary peritonitis due to perforation of a hollow organ. Intestinal obstruction occurs either through intussusception starting from the diverticulum or through volvulus around a fibrous cord connecting the diverticulum to the umbilicus, the ileum to the umbilicus

(omphalomesenteric band). In diverticulitis, the symptoms are like appendicitis, but less intense, and the diagnosis is often made intraoperatively. In adults, symptoms may be due to neoplasms starting from Meckel's diverticulum.

Positive diagnosis is often made incidentally during abdominal surgery for other conditions (appendicitis). Laboratory tests may reveal anemia in cases of bleeding or signs of inflammation in cases of diverticulitis or perforation. Abdominal ultrasound may be useful in cases of complications. Tc99 scintigraphy can reveal the presence of a Meckel's diverticulum by fixing Tc99 in the ectopic gastric mucosa. Diagnostic laparoscopy, which can serve as both a diagnostic and therapeutic tool, has higher sensitivity and specificity.

Differential diagnosis includes conditions that cause gastrointestinal bleeding: esophageal varices, gastroduodenal ulcer, solitary rectal polyp, familial intestinal polyposis, Crohn's disease, ulcerative necrotic colitis, or clinical picture of acute surgical abdomen: acute appendicitis, intussusception, intestinal volvulus.

Treatment is surgical. Diagnosed incidentally or based on symptoms, Meckel's diverticulum must be excised. Simple resection of the diverticulum can be performed, or together with the segment of intestine that contains it, followed by restoration of the continuity and integrity of the digestive tract. Exceptions occurs when the diverticulum was discovered in a patient in serious condition or with inflammation of the peritoneal cavity from another cause (e.g. appendicular peritonitis, perforated gastric or duodenal ulcer, etc.). In such cases, if the diverticulum is uncomplicated left in place and excised at a later stage. The approach can be open or laparoscopic.

Prognosis is favorable if the diagnostic and treatment are performed correctly.

TUMORAL PATHOLOGY

Chapter 26. NEFROBLASTOMA (Wilms tumor)

Definition. Nephroblastoma or Wilms tumor is a neoplasm with embryonic origin in the metanephric blastoma cells.

Epidemiology. Wilms tumor has an annual incidence of 0.8-1:100,000 children. It is one of the most common solid tumors in children, accounting for 5% of all pediatric cancers.

Pathology. Wilms tumor is characterized by a triphasic embryonal histologic pattern composed of three types of cells: blastemal, stromal, epithelial. The tumor can have favorable histology in 90% of cases with a good prognosis; however, tumor with anaplastic histology with enlarged, hyperchromatic nuclei, and atypical have a reserved prognosis.

Clinical picture. Most children with Wilms tumor present with an asymptomatic abdominal tumor mass, usually detected by parents or a doctor. The child may reveal hematuria, abdominal pain due to intratumoral or extra-tumoral capsular rupture. Approximately 20% of children with nephroblastoma have hematuria, 10% have coagulopathy, and 25% exhibit hypertension due to activation of the renin-angiotensin system. Fever, anorexia, and weight loss occur in 10% of cases. In rare cases, tumor rupture and bleeding can cause acute abdomen. The tumor extends into the renal vein, inferior vena cava, and right atrium or may descend through the ureter. It metastasizes in the lungs and liver. Vena cava thrombosis can cause mortality when an unidentified thrombus embolizes during nephrectomy. Tumor rupture will determine the stage of the disease and is an emergency. Ruptures may be asymptomatic, confined to the retroperitoneum, or associated with self-limiting bleeding.

Wilms tumor can occur as part of a genetic syndrome such as: WAGR (Wilms tumor, aniridia, genitourinary anomalies, and mental retardation), Denys-Drash syndrome (nephropathy, renal failure, male pseudo-hermaphroditism), Beckwith-Wiedemann syndrome (macroglossia, hyperinsulinemia hypoglycemia of infancy, omphalocele, visceromegaly), asymmetric growth, or a family history of tumors.

Diagnosis. Medical history and examinations often provide clinical diagnostic. Ultrasound, MRI, and CT imaging are used to differentiate the nature of the mass, local invasion and vascular extension along the renal vein or inferior vena cava, and metastatic disease, usually to the lungs (Figure 26.1). **Staging of Wilms tumor** (Table 26.1)

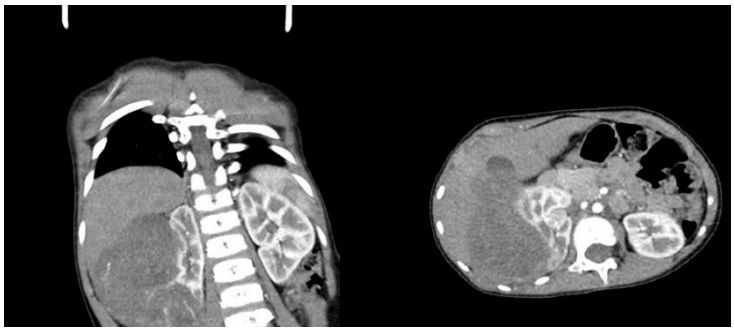


Fig. 26.1. Nephroblastoma, CT images

Table 26.1. Staging of nephroblastoma

Stage	Diagnostic criteria
I	Tumor confined to the kidney and completely resectable Tumor has not ruptured and no diagnostic biopsy was performed prior to excision No penetration of the renal capsule or invasion of renal vessels
II	The tumor extends beyond the renal capsule but has been completely resected There is penetration of the renal capsule and/or renal vessels
III	Micro- or macroscopic residual tumor, including inoperable tumors, positive tumor margins, tumor rupture, locoregional metastases in lymph nodes, positive peritoneal cytology, or tumor biopsy prior to surgery
IV	Hematogenous or lymphatic metastases outside the abdomen
V	Bilateral nephroblastoma. Each tumor will be staged individually.

Differential diagnosis includes benign lesions (multicystic dysplastic kidney) and neoplastic lesions (clear cell sarcoma of the kidney), mesenchymal nephroblastoma, and malignant rhabdoid tumors.

The **treatment** of nephroblastoma is combined and includes surgery, chemotherapy, and, in some cases, radiotherapy. Treatment is based on age, stage, histology, and molecular biomarkers. Appropriate medical therapy is indicated before surgery. The large size of the nephroblastoma makes initial resection of the tumor difficult. Needle biopsy may be necessary in selected cases to confirm the diagnosis. Tumor reduction chemotherapy is initiated before nephrectomy, followed by additional chemotherapy according to stage and histology.

Chemotherapy. Depending on the treatment protocol used, patients may receive adjuvant or neoadjuvant chemotherapy. Either before or after surgical resection, most patients are treated with a combination of Vincristine, Dactinomycin, and Doxorubicin. Cyclophosphamide, Etoposide, and Carboplatin may also be used in selected cases.

Surgical treatment consists of radical nephrectomy and is performed trans-peritoneally, either by open approach (laparotomy) or minimally invasive (laparoscopic, robotic). The entire tumor and surrounding tissue involved are resected "en bloc" to prevent iatrogenic dissemination of the tumor. Partial nephrectomy carries a risk of recurrence and is preferred only in cases of bilateral tumors or tumors on a solitary kidney.

Radiotherapy. The need for radiotherapy depends on the stage of the tumor, histology, and molecular type. Nephroblastoma is very radiosensitive, and radiotherapy should begin 14 days after surgery.

Complications. Intraoperative: damage to adjacent structures (liver, intestine, pancreas) and bleeding. Postoperative: infections, bleeding, ileo-

ileal intussusception. This occurs in 10% of patients; treatment is reduction the intussusception tumor by laparotomy. Late: obstruction due to adhesions, chronic renal failure (<1%), and secondary malignancy (1.6%).

Prognosis. The prognosis is influenced by the stage of the tumor, the histological subtype, and the precocity and accuracy of treatment. The 5- year survival rate reaches 90%. However, in the case of large tumors with unfavorable histology, the survival rate decreases to approximately 50%. Recurrence usually occurs within the first 2 years after diagnosis. The most common sites of recurrence are the abdomen, lungs, and contralateral kidney. Late effects of chemotherapy and radiotherapy include secondary malignancies and cardiac dysfunction, hence the need to reduce therapy for low-risk disease.

Chapter 27. NEUROBLASTOMA

Definition. Neuroblastoma is a malignant tumor originating in the primitive neural crest cells of the sympathetic nervous system. It can occur anywhere in the sympathetic nervous system, but in 65% of cases the origin is in the adrenal gland.

Epidemiology. The incidence is 1 in 100,000 children, with a male to female ratio of 1.2:1. It is the most common extracranial solid malignant tumor in children and the most common malignant tumor in infants. Neuroblastoma is responsible for approximately 15% of all cancer deaths in children.

Pathological anatomy. Macroscopically, it is a firm, white-gray, friable tumor, sometimes with a bleeding surface. Most often, the starting point is one of the adrenal glands, but it can virtually start anywhere where there is sympathetic nerve tissue. Histologically, it is a tumor with small, round, blue, uniformly sized cells with dense hyperchromatic nuclei and minimal cytoplasm. The tumor cells are arranged in rosettes (circular formation) around a neural process, forming structures known as Homer-Wright rosettes.

Morphological classification (Shimada modified INPC) is based on several histopathological prognostic factors such as tumor morphology, mitotic index (MKI), presence or absence of stroma, and age. The prognosis may be either favorable (ganglioneuroma, diffuse ganglioneuroblastoma, low MKI, and age under 18 months) or unfavorable (nodular ganglioneuroblastoma, undifferentiated or poorly differentiated neuroblastoma with intermediate or high MKI, and age over 18 months).

Clinical picture is extremely variable and depends on the location of the primary lesion, the extent of metastatic disease, and the presence of associated paraneoplastic syndromes. It may be diagnosed incidentally, with the patient presenting non-specific symptoms such as pain and malaise. Thoracic tumors may be associated with the Horner's syndrome (ptosis, miosis, and anhidrosis) or symptoms related to spinal cord compression with intrathecal extension. Abdominal tumors cause symptoms due to compression of neighboring organs (constipation or urinary retention).

Children with neuroblastoma sometimes present with characteristic periorbital ecchymosis called "raccoon eyes" (Figure 27.1) and eyelid ptosis. Sometimes, the patient may present diarrhea, flushing, and excessive sweating in tumors that secrete catecholamines or nystagmus/"dancing eyes" with cerebellar syndrome "opsoclonus-myoclonus." Neuroblastoma often infiltrates local structures and surrounds vital nerves or vessels. Tumors usually metastasize to regional lymph nodes and bone marrow. In infants, neuroblastoma metastasizes to the liver. Infants may present with skin metastases that look like "currant berries." Newborns may present with abdominal distension and respiratory compromise due to tumor mass or liver metastases causing massive hepatomegaly. Paraspinal tumors can lead to paralysis due to spinal cord compression, Horner's syndrome (ptosis, miosis, and anhidrosis) or symptoms related to spinal cord compression with intrathecal extension are possible.

Clinical exam is very important. Symptoms such as lethargy, ecchymosis, bleeding, and bone pain are associated with metastatic disease. Important factors to assess in the abdominal examination are tumor location, size, consistency, and mobility, extension across the midline or into the pelvis.

Paraclinical. Imaging investigations assess the presence of the tumor, its origin, and its relationship to surrounding structures and metastases. Chest X-ray can determine the presence of a tumor mass in the posterior mediastinum. Abdominal X-ray is less commonly used. Calcification (speckled appearance) is observed in 90% of tumors and suggests a diagnossis of neuroblastoma. Ultrasound highlights the primary tumor, its origin, and its extent. CT can demonstrate tumor extension and metastases. MRI is the most sensitive imaging method for diagnosis and staging neuroblastoma, being the method of choice, clearly superior to CT for determining local extent and metastases (Figure 27.2). Laboratory tests such as serum lactate dehydrogenase, ferritin, and neuron-specific enolase reflect tumor burden and have prognostic significance. High levels of these are found in advanced stages and indicate a poor prognosis. Collecting urine over 24 hours for catecholamine metabolites: vanillylmandelic acid (VMA) and homovanillic acid (HVA) is valuable in diagnostic evaluation. The VMA/HVA ratio provides information in diagnostic and in monitoring response to therapy and recurrence.

Fig. 27.1. "Raccoon eyes" eyelid ecchymosis

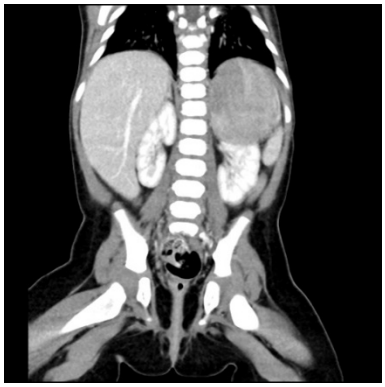


Fig. 27.2. Left adrenal gland neuroblastoma

The **definitive diagnosis** is made by histopathological examination of the primary or metastatic tumor tissue or by demonstrating tumor cells in the bone marrow correlated with elevated urinary catecholamine levels.

Differential diagnosis iincludes tumors (nephroblastoma, lymphoma, Ewing's sarcoma, rhabdomyosarcoma), neurological diseases, and gastrointestinal diseases.

Treatment. Based on clinical and biological variables, infants and children with neuroblastoma are divided into three groups: low, intermediate, and high risk. Treatment is combined. Surgery is followed by chemotherapy and/or radiation therapy. Low-risk patients can be treated by means of surgical excision, with excellent cure rates. Moderate-risk patients are treated with mild chemotherapy or surgery, and the prognosis is good.

High-risk patients are treated with intensive chemotherapy, surgery, radiotherapy, and immunotherapy, but the prognosis remains poor. Surgery is followed by additional high-dose chemotherapy, bone marrow ablation, and stem cell transplantation. Retinoic acid, to encourage cell maturation, immunotherapy with antibodies against GD-2, and radioactive MIBG are used to eliminate minimal residual disease and consolidate remission. After treatment, the patient is monitored by measuring urinary catecholamines and CT and MRI imaging for the primary tumor, as well as MIBG and PET for bone and bone marrow metastases.

Outcome and prognosis. Survival in patients with low-risk neuroblastoma is over 98%. Half of patients with high-risk neuroblastoma will exhibit relapse of the neoplasia within the first 2 years of treatment. The probability of prolonged disease-free survival for patients in each risk group is >95%, >90%, and <30%, respectively.

Chapter 28. TERATOMAS

Definition. A teratoma is a cystic or solid tumor composed of various structures and tissues originating from totipotent embryonic cells. It typically contains a variety of tissues other than those of the area in which it occurs.

Etiopathogenesis. Teratomas are considered germ cell tumors originating in primordial totipotent cells, which develop from yolk sac cells near the origin of the allantois and migrate to the gonadal ridges during weeks 4 and 5 of gestation. Some cells miss their destination and settle anywhere, usually on the midline, from the brain to the sacrococcygeal region, giving rise to a teratoma. Other etiopathogenesis theories support the origin in remnants of the primitive streak and node or incomplete twinning.

Pathological anatomy. Teratomas are composed of various structures and tissues (virtually any type of tissue in the human body, other than those of the anatomical structure in which the teratoma is located), skin, nervous tissue, teeth, fat, cartilage, and intestinal mucosa, etc. In children, the most comm location is in the sacrococcygeal region or in the ovary. Less common locations include the mediastinum, retroperitoneum, testicle, cervix, or CNS. They are usually benign but may contain or develop foci of malignancy. They are classified according to their degree of malignancy as mature or immature.

28.1. SACROCCOCCYGEAL TERATOMA

It accounts for 35-60% of all teratomas and is the most common tumor in newborns, with an estimated incidence of 1 in 35,000-40,000 live births. Females are more frequently affected. The diagnosis can be set prenatally by ultrasound in the second trimester of pregnancy. Small teratomas do not

adversely affect the fetus, but large ones are associated with a significant morbidity and mortality rate, and delivery by caesarean section is recommended. They are frequently associated with other malformations: Curarino triad (ano-rectal malformation, sacral anomalies, presacral tumor), urogenital, vertebral-medullary, or skeletal malformations.

At birth, they are usually clinically evident: sacrococcygeal tumor formation, covered by normal skin or red or ulcerated skin (Figure 28.1). It is usually asymptomatic, but symptoms may occur due to compression of the rectum, bladder, prematurity, or circulatory failure.

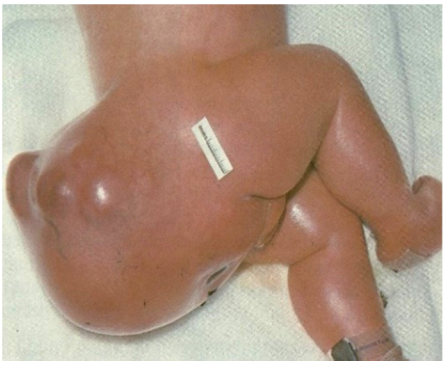


Fig. 28.1. Sacrococcygeal teratoma

The clinical diagnostic is supplemented by imaging using spinal X-ray, ultrasound, CT, or MRI, which specify the extent of the tumor (Figure 28.2) and its structure (cystic or solid). The tumor marker specific to teratomas is alpha- fetoprotein, the measurement of which is useful in diagnostic and postoperative follow-up.

I.	External tumor (45%)
II.	External tumor + pelvic component (34%)
III.	External tumor + pelvic + intra-abdominal (9%)
IV.	Exclusively presacral without external component (10%)

Fig. 28.2. Altman classification is based on the extension of the sacrococcygeal teratoma into the pelvis

Treatment. Surgical resection in the case of benign teratomas is curative. The surgical approach depends on the Altman classification, perineal or perineal and abdominal. In patients with malignant teratoma, surgical resection is combined with chemotherapy.

Prognosis. Local and distant recurrences after surgical excision are quite common. The risk of malignancy is less than 10% at birth but increases to 40-75% after 1 year of age for sacrococcygeal teratoma. The risk of malignancy is high for incomplete excisions.

28.2. TERATOMAS OF THE GENITAL ORGANS

Teratomas of the genital organs are the second most common type of teratoma after sacrococcygeal teratomas. They are usually diagnosed later in life. Ovarian teratomas grow slowly and have few symptoms, often reaching considerable size by the time they are diagnosed. The clinical picture is related to the mass effect of the tumor, palpable tumor mass, torsion of the tumor-bearing ovary, or compression of neighboring organs. Sometimes they are discovered incidentally during an abdominal ultrasound. The imaging diagnosis is based on abdominal ultrasound and may be supplemented by MRI (Figure 28.2) or CT, and laboratory tests usually show elevated alpha-fetoprotein levels. Surgical excision (usually oophorectomy) is the primary method of treatment. If the tumor has been completely excised and the teratoma is mature (benign), no further treatment is necessary. Immature teratomas usually require adjuvant chemotherapy.

Testicular teratoma presents as a palpable tumor mass in one of the testicles. Suspicion of a tumor in the testicles requires preoperative imaging and excision of the testicle through an inguinal incision with high ligation of the spermatic cord. Completely excised tumors are managed solely with

surgical removal and careful observation. More advanced diseases are managed by surgery and chemotherapy.

The **prognosis** for teratomas is generally favorable, with over 90% of the survival rate of 5 years, most being mature and completely excisable teratomas.

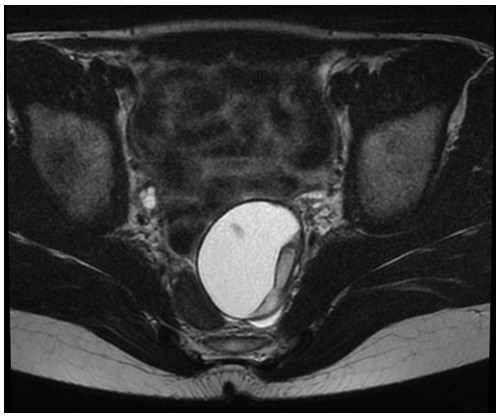


Fig. 28.2. Ovarian teratoma

Chapter 29. RHABDOMYOSARCOMA

Definition. Rhabdomyosarcoma is a malignant mesenchymal tumor that exhibits histological features of aberrant differentiation of skeletal muscle. They usually occur in sites distant from skeletal muscle such as the biliary tract, and urogenital system.

Epidemiology. The global incidence is 6/100,000/year. Rhabdomyosarcomas have a bimodal pattern of occurrence, with two peaks of incidence, one at the age of 2-5 years and a second around the age of 15-19 years.

Pathological anatomy. Rhabdomyosarcomas belong to the family of small, round blue cell tumors. There are three histological subtypes: embryonal, alveolar, and pleomorphic. Approximately 15% of rhabdomyosarcomas have metastases at the time of diagnosis, which is an unfavorable prognostic factor.

Clinical picture. Symptoms tend to be specific to the site of origin, with an obvious tumor mass, due to compression of adjacent structures. Tumors of the bladder and prostate cause urinary symptoms such as hematuria, urinary frequency or retention, and constipation. Vaginal tumors cause bleeding, discharge, or a visible mass, like a "bunch of grapes," botryoid tumors. Paratesticular tumors usually present as a mass in the scrotum or groin area.

Positive diagnosis is made by biopsy with sufficient tissue to perform biological studies. Lymph node sampling, bone marrow aspirates, and bone scans are indicated to evaluate distant metastases. Ultrasound, CT, and MRI are used to evaluate both local disease and distant metastases. PET-CT is used to search for residual and metastatic disease. Tumors are ultimately classified

into low-, intermediate-, and high-risk groups based on stage (TNM classification) and histology.

Treatment is multimodal, involving surgery, chemotherapy, and radiotherapy. Surgery aims to achieve wide local excision of the tumor with preserved safety margins. Chemotherapy regimens are based on Vincristine, Actinomycin-D, and Ifosfamide/Cyclophosphamide, but new agents such as Irenotecan and Topotecan have been introduced for high-risk disease. The effects of pelvic radiotherapy in young children can be very debilitating, which is why European protocols have generally aimed to limit its use.

Prognosis. Overall survival rates for rhabdomyosarcoma treated with current combination therapy regimens are over 60%. The prognosis is related to the risk of the disease; low-risk tumors have a 5-year survival rate of over 90%, intermediate-risk tumors 55-70%, and high-risk tumors less than 50%.

Chapter 30. HEPATIC E TUMORS

Primary liver tumors are rare in childhood, accounting for less than 2% of all pediatric malignancies. Of these, 80% are hepatoblastomas, with an incidence of approximately one in a million children.

30.1. HEPATOBLASTOMA

Hepatoblastoma is a malignant liver tumor originating in fetal hepatocytes.

Incidence. It is the most common primary malignant embryonic liver tumor in childhood and accounts for approximately 1% of childhood cancers. Over 80% of patients are diagnosed before the age of 3.

Premature infants have a significantly increased risk of developing hepatoblastoma, and the increased incidence in recent years has been attributed to improved care and increased survival of these newborns. Hepatoblastoma also has a higher incidence in Beckwith-Weidemann syndrome and familial adenomatous polyposis.

Pathological anatomy. They are generally unifocal lesions and can be histologically differentiated into six subtypes that correlate with clinical progression.

Clinical picture. Presentation is usually with an asymptomatic abdominal tumor mass. Sometimes it may be associated with fever, lethargy, anorexia, and abdominal pain. Jaundice is rare. Occasionally, these tumors may occur in a critically ill child with tumor rupture, requiring resuscitation.

Paraclinical investigations. Ultrasound is useful for evaluating the liver and vascular involvement. CT or MRI evaluation is essential to look for metastatic disease, which most commonly occurs in the lungs. Laboratory

tests should include a complete blood count, as thrombocytosis is frequently associated, and alpha-fetoprotein (elevated in >90%).

Positive diagnosis. Biopsy will make a definitive histological diagnosis. Due to the potential risks of biopsy (bleeding, tumor rupture), selected patients may be treated based solely on age, classic imaging characteristics, and serum AFP alone. In addition to the location of the disease, there are a variety of subclassifications, including lymph node involvement (N), vascular invasion (P and V), multifocal disease (F), extrahepatic spread (E), and distant metastases (M).

Differential diagnosis includes malignancies such as germ cell tumors, sarcoma, hepatocellular carcinoma, and benign lesions such as hemangiomas and mesenchymal hamartomas. Some benign liver tumors, such as mesenchymal hamartomas, may have elevated AFP. Elevated AFP can be difficult to interpret in young infants, where levels are naturally high at birth and progressively decline during the first year of life.

Treatment. Primary surgery is preferred for all tumors where complete resection can be achieved, followed by chemotherapy based on extension and histological subtype. Surgery treatment aims to complete tumor resection with a margin of only a few millimeters considered acceptable. In general, hepatoblastomas occur in non-cirrhotic liver, and resections of up to 85% of the liver tissue can be tolerated. Advanced tumors and those involving the portal vein should be considered for treatment by orthotopic liver transplantation, as complete resection is impossible. Cisplatin-based regimens form the basis of chemotherapy for hepatoblastoma.

Prognosis. Overall five-year survival ranges from 50 to 100%, depending on clinical, biochemical, and histological variables. Outcomes have improved significantly in recent years.

30.2. HEPATOCELLULAR CARCINOMA

It is a relatively rare tumor but it exhibits aggressive malignant behavior. It occurs in older children, with an average age of 11 years. Congenital metabolic and inflammatory liver diseases predispose to hepatocellular carcinoma. The tumor can be multicentric, which limits the possibilities of complete surgical excision.

Clinically, it presents as a palpable abdominal mass and in approximately 10% of cases, it manifests as intraperitoneal rupture of the tumor with hemoperitoneum. Treatment for early stages is complete surgical excision (usually controlled liver resections) followed by chemotherapy. Unfortunately, primary resection is possible in only a few cases (10%). The prognosis is poor, with an overall survival rate of approximately 15%.

30.3. HEPATIC METASTASES

Neuroblastoma is the most common solid tumor in children that metastasizes to the liver (20-30%), followed by Wilms tumor (10-15%). Germinal tumors, gastrointestinal stromal tumors, osteosarcoma, small round desmoplastic tumors, and neuroendocrine tumors can also metastasize in the liver. Resection of liver metastases has demonstrated therapeutic and survival benefits in selected patients with a variety of primary tumors.

PEDIATRIC TRAUMATOLOGY

Chapter 31. ASSESSMENT OF CHILDREN WITH TRAUMA

Introduction. Trauma is the leading cause of death and disability in children. Unintentional trauma predominates in children. The vast majority of cases can be prevented by general measures such as compliance with traffic safety rules, education, supervision, and are influenced by the socio-economic status of the individual and the community.

The anatomical, social, and psychological differences between children and adults have important implications for the initial assessment and management of pediatric trauma victims. In children, more elastic connective tissue and a malleable skeleton protect the abdominal and thoracic structures. The force of an impact is distributed widely throughout a child's body, resulting in multisystem injuries in nearly 50% of children with severe trauma. Fluid and heat loss is greater compared to adults.

Children have a different physiological response to major trauma than adults. Children can maintain near-normal blood pressure even with a 25-30% loss of blood volume. In these situations, subtle changes in heart rate and limb perfusion can signal impending cardiorespiratory failure and should not be overlooked.

Children may not cope well emotionally after an accident. They should be treated in a calm, child-friendly environment. The presence of a parent or guardian in the resuscitation room can help the trauma team by minimizing the fear and anxiety of the injured child. There is evidence that

25% of children suffer from post-traumatic stress disorder after a motor vehicle collision.

Rapid and well-organized assessment of an injured child by a team is essential. The term "golden hour," although not literally defined, refers to an early and critical period in the care of trauma victims, during which proper management can significantly increase patient survival rates.

The main causes of fatal injuries by age group are in children under one year of age, airway obstruction (choking) predominates; in children aged 1-4 years, choking and transport-related injuries predominate; and in children over 5 years of age, transport-related injuries (road accidents) predominate. Most injuries are minor, consisting of soft tissue damage - bruises, abrasions, hematomas, and wounds, but in cases of severe injury or multiple injuries, rapid and correct intervention can be lifesaving.

Primary assessment and initial stabilization. The primary assessment must be carried out sequentially, but usually, the trauma team, under the coordination of the team leader, performs the components of the primary survey simultaneously, so that the entire process takes only a few minutes. The purpose of the primary survey is to find and improve immediate life-threatening conditions. The primary stabilization begins at the scene of the accident and aims to ensure a clear airway, adequate breathing, circulatory support, and to assess major neurological disabilities. The objectives of initial stabilization include providing adequate oxygen and ventilation, fluid resuscitation, and preventing secondary injuries to target organs. The primary stabilization includes frequent reassessments to confirm or rule out injuries that require immediate surgical intervention.

Emergency medicine protocols have been developed to ensure the rapid and systematic assessment and stabilization of polytraumatized patients, following the "ABCDE" sequence:

A. **AIRWAY.** Assessing the airway simply involves determining the ability of air to pass unimpeded into the lungs. The airway can be obstructed anywhere between the lips and the carina by direct trauma, edema, secretions, blood, gastric contents, or foreign bodies. If the level of consciousness (LOC) is low, the child may not be able to maintain a patent airway and/or protect the lungs from aspiration of stomach contents due to loss of the swallowing reflex. The classic sign of partial upper airway obstruction is inspiratory stridor. Breathing effort without airflow indicates complete airway obstruction.

B. **BREATHING.** Respiratory compromise in the polytrauma child after airway clearance generally results from the absence of spontaneous breathing due to brain injury or chest injury (e.g., massive pneumothorax). Breathing should be assessed to determine the child's ability to ventilate and oxygenate. Respiratory failure can be anticipated by monitoring the following signs: increased respiratory rate, especially in the presence of signs of distress (increased respiratory effort, chest retractions, oscillating breath sounds, or grunting), inadequate effort or chest excursion (decreased breath sounds or gasping), especially if the mental status is depressed, cyanosis with abnormal breathing despite supplemental oxygen.

C. **CIRCULATION.** Rapid vascular access is performed immediately. Sometimes it is very difficult to perform vascular access. Often, children with severe multiple traumas have normal vital signs despite significant blood loss due to extraordinary compensation, but cardio-circulatory decompensation can be sudden and difficult to control.

D. **DISABILITY.** A rapid assessment of neurological function is performed at the end of the primary survey: whether the patient is conscious and alert or responds only to verbal stimuli, or only to pain, or is unconscious/unresponsive, followed by an assessment of pupil symmetry and pupillary light reflex. This assessment is repeated during the secondary survey to monitor for any changes in the child's neurological status. Causes of a decreased level of consciousness in injured children include traumatic brain injury (TBI), hypoxemia, and poor cerebral perfusion. The latter two can exacerbate TBI and lead to secondary brain injury.

E. **EXPOSURE.** Full exposing the child is essential to perform a complete head-to-toe examination and to identify life-threatening injuries. It is important to note that this systematic assessment is most effective and significantly reduces mortality if performed correctly and quickly. Hypothermia should be avoided.

After initial assessment and stabilization, the patient will be transferred to a pediatric trauma center. The decision whether to transfer the injured child to a trauma center or the nearest available facility depends on the patient's condition, the extent and severity of the injuries, as well as geographic location and local policies.

Chapter 32. THORACIC TRAUMA

Overview. Thoracic trauma is less common in children than in adults and is more common in boys than in girls. Most thoracic trauma results from non-penetrating agents (traffic accidents) or falls from height, while trauma caused by contact with sharp, penetrating objects is less common. Life-threatening chest injuries, such as airway obstruction, tension pneumothorax, massive hemothorax, and cardiac tamponade, should be identified and treated during the primary survey.

Anatomy and physiology. Children have several anatomical features, such as an elastic and flexible rib cage, no protective muscle mass, and a wider mediastinum. The shape of a child's rib cage allows for significant injuries with few obvious external signs of trauma. This means that energy is transmitted more easily to the intra-thoracic structures, making pulmonary contusions and hematomas relatively more frequent and severe compared with those seen in adult. A child's head is proportionally much larger than that of an adult and predisposes to neck flexion and airway occlusion in the supine position. The larger tongue and soft palate, as well as the more anterior glottis, can make the airway difficult to visualize. A child's trachea is shorter in relation to body size, narrower, and more easily compressed than that of an adult. The subglottic region is the narrowest part of the trachea in children. The transverse diameter of the airway is small, predisposing to obstruction with edema. Because children have high oxygen consumption and low functional residual capacity, pulmonary contusions can lead to severe hypoxemia, which may be refractory to oxygen therapy. These characteristics make rib fractures less common in children, but the more impact forces are transmitted to the intrathoracic organs.

Classification of thoracic lesions according to their location:

- Chest wall injuries: rib fractures, flail chest, open pneumothorax, traumatic asphyxia
- Pleuropulmonary injuries: pneumothorax, hemothorax, pulmonary lesions, diaphragmatic injuries
- Mediastinal injuries: cardiac tamponade, tracheobronchial tree injuries, large vessel injuries, cardiac contusion, esophageal injuries

Diagnosis is based on clinical examination (inspection, palpation, percussion, and auscultation) specific to each type of injury. Imaging diagnostic includes plain chest X-ray in two views (anteroposterior and lateral) or special views (e.g., lateral decubitus) as well as chest CT.

The most common and/or important chest injuries are:

1. **Chest concussion** results from the action of a minor impact. It manifests as local bruise, spontaneous pain and pain on palpation, limited respiratory movement, and antalgic posture. It does not require specific treatment; physical rest and pain management are sufficient.
2. **Costal volet (flail chest)** occurs because of multiple rib fractures resulting in a morphologically and functionally independent segment of chest wall. Paradoxical respiratory movements lead to respiratory failure of varying degrees. The principles of treatment are pain control, positive pressure ventilation (orotracheal intubation), oxygen therapy, and physical therapy aimed at relieving respiratory failure and preventing pneumonia. If non-invasive treatment fails, rib fractures can be stabilized using cerclage, intramedullary nails, or plates with screws.
3. **Traumatic asphyxia** is caused ed by a sudden, severe compression of the chest whith the glottis closed, typically following a deep inhalation by the patient. There sudden increase in intrathoracic pressure leads elevated

pressure in the superior vena cava (which has no valves)- resulting in retrograde blood flow and distension of jugular veins. The patient presents with altered consciousness, hypotension, tachycardia, exophthalmos, subconjunctival and meningeal hemorrhages, mucosal petechiae, epistaxis, hemoptysis, and pulmonary contusion. Treatment consists of oxygen therapy, inotropes, evacuation of a pneumothorax or hemothorax if present, and positive pressure ventilation.

4. **Hemothorax** is defined by the presence of blood in the pleural cavity and results from pleural, pulmonary or vascular injuries (intercostal artery, internal mammary artery). A moderate amount of blood causes minor respiratory changes, while large accumulations compress the lung in the hilum, displace the mediastinum, and sometimes compress the contralateral lung, leading to respiratory failure. Intrapleural blood does not usually coagulate. On percussion, dullness is noted. When this reaches the angle of the scapula, respiratory failure symptoms become evident (cyanosis, dyspnea, retraction, wheezing). Investigation methods include chest X-ray performed in an orthostatic position or lateral decubitus and CT. Treatment consists of evacuation by pleural puncture with a thick trocar in full dullness with evacuation of 100-200 ml initially, 200-300 ml after 24 hours, and 400 ml or more after 5-6 days. Minimal thoracotomy (Bühlau) is performed with continuous active or passive aspiration (the 3-jar method). Rarely, extended thoracotomy is necessary for vessel ligation in cases of massive or persistent hemorrhage with respiratory failure and hemorrhagic shock.
5. **Pneumothorax** is the accumulation of free air in the pleural cavity with compression of the lung toward the hilum. It occurs more frequently in penetrating wounds. In this case, it is called open pneumothorax and may

be associated with subcutaneous emphysema extending to the cervical region. In pulmonary contusions, it arises from alveolar ruptures or even bronchi or terminal bronchioles (simple, closed pneumothorax). Thoracic asymmetry is observed with widening of the intercostal spaces, asymmetric respiratory movements, hyperresonance, and diminution or abolition of vesicular murmur. Usually, a simple chest X-ray confirms the diagnostic. It shows hyper-transparency of one hemithorax and collapse of the lung in the hilum, deviation of the mediastinal shadow due to compression. Valved pneumothorax is an open pneumothorax where air enters the pleura during inspiration but is not evacuated during expiration, leading to the development of a massive pneumothorax that requires prompt drainage.

6. **Chylothorax** is caused by damage to the thoracic duct. Usually, chylothorax becomes evident 3-7 days after injury. The diagnostic is made by obtaining a sample of pleural fluid and identifying the lipid content. Treatment includes drainage and enteral feeding with medium-chain triglycerides or parenteral nutrition. Surgery is indicated only after failure of medical treatment.
7. **Pulmonary contusion** is one of the most common chest injuries in children. It can occur in cases of blunt or penetrating trauma. A child's flexible chest wall allows for lung contusion without rib fractures. The presence of pulmonary contusion contributes to decreased lung compliance and leads to hypoxia and hypoventilation. A chest X-ray performed during the initial assessment can demonstrate pulmonary contusion, but it is difficult to differentiate it from fluid in the pleural space. Chest CT can reveal areas of pulmonary contusion that are not visible on chest X-ray (Figure 32.1). A significant percentage of cases

require ventilatory support. Treatment includes limited fluid resuscitation, supplemental oxygen, pain management, and strategies to prevent atelectasis and pneumonia. A significant percentage of patients may develop pneumonia or acute respiratory distress syndrome after pulmonary contusion.

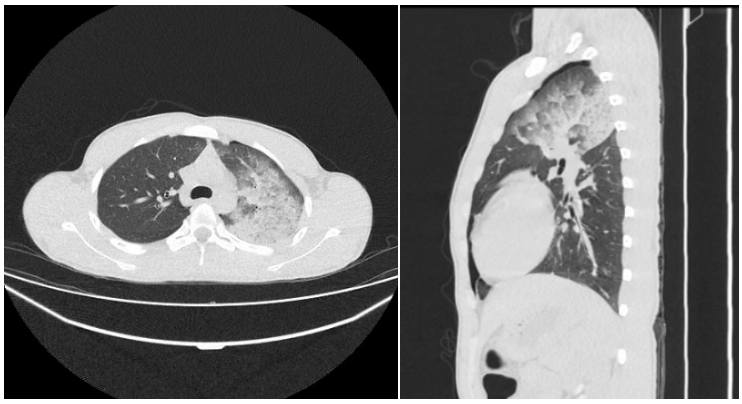


Fig. 32.1. Pulmonary contusion and right traumatic pneumothorax

8. **Diaphragmatic rupture** caused by blunt chest trauma is an uncommon event. The left diaphragm is more often involved, due to the protective effect of the right hepatic lobe. Bilateral diaphragmatic rupture is extremely rare. Clinical manifestations include chest pain radiating to the shoulder, dyspnea, and/or abdominal pain. Breath sounds may be diminished, and bowel sounds may be heard over the ipsilateral chest. On imaging studies, an abnormal diaphragmatic contour, a high diaphragm, or questionable overlap of abdominal visceral shadows may suggest a diaphragmatic injury. Greater awareness of this injury is necessary to avoid late complications of visceral herniation or intestinal injury. When penetrating trauma is below the nipple line, a diaphragmatic injury should be considered. Treatment consists of surgical repair of the diaphragm

using a classic or minimally invasive thoracic, abdominal, or combined approach.

9. **Tracheobronchial injuries** are rare in children. They can occur in cases of penetrating or blunt trauma, such as acceleration or deceleration. Almost half occur in the first 2 cm of the right main bronchus. Most patients with tracheal injuries have cervical subcutaneous emphysema and mediastinal air on chest X- ray. More distal injuries produce tension pneumothorax. Chest CT or bronchoscopy may sometimes be necessary to establish the diagnosis. Once diagnosed, the injury requires prompt treatment such as chest drainage, mechanical ventilation. Delayed diagnosis is not uncommon in children, as injuries may be incomplete and symptoms minimal. Distal bronchial injuries are generally well managed by lung resection, while more proximal airway trauma is best treated by direct repair. Non-operative management of a tracheobronchial injury may lead to airway stenosis.
10. **Lesions of large vessels** are rare in young children. Most children who suffer a rupture of the thoracic aorta will have significant associated injuries to the lungs, heart, skeletal system, abdominal organs, or central nervous system. Diagnosing an aortic injury in a child can be difficult. Aortic angiography provides excellent anatomical detail. The most common finding in an aortic injury is a pseudoaneurysm located in the proximal descending aorta. All blunt aortic injuries should initially be treated with control of heart rate and blood pressure. While lacerations and intimal tears can be managed nonoperatively and followed with repeated imaging, pseudoaneurysms require semi-urgent repair.
11. **Cardiac injuries.** The manifestation of these injuries can vary from completely asymptomatic to rapid exsanguination and death at the scene

of the accident. Commotio cordis is a disorder described in the pediatric population that results from a sudden impact on the anterior chest wall, causing cessation of normal cardiac function. The child may present with immediate arrhythmia or ventricular fibrillation refractory to resuscitation efforts.

12. **Esophageal injuries** are less common in pediatric patients. Penetrating trauma accounts for majority of esophageal injuries, with the cervical esophagus being the most commonly affected segemnt. Clinical signs such as hypersalivation from the oral cavity are useful for guiding the diagnosis. Symptoms such as dysphonia and dysphagia and chest or epigastric pain are nonspecific. Intrathoracic esophageal injuries often produce hemothorax, pneumothorax, or pneumomediastinum. Esophagoscopy is often used to set diagnosis. Although iatrogenic injuries can be treated non-operatively with antibiotics, surgery is the standard approach for esophageal injuries caused by external penetrating or blunt force. Delay in surgical exposure and drainage often result in sepsis and death. Late-discovered injuries are often associated with sepsis and are best treated with broad-spectrum antibiotics, debridement, extensive drainage, and digestive surgical bypasses.

Chapter 33. ABDOMINAL TRAUMA

Overview. Abdominal trauma is the most common cause of unrecognized fatal injuries in children. Due to its severity, abdominal trauma ranks first in importance. It is more common among boys. Bicycle handlebars are a common cause of blunt abdominal trauma. The most affected organs in abdominal trauma are the spleen and liver, followed by the kidneys and pancreas.

Anatomy and physiology. Children have proportionally larger solid organs, less subcutaneous fat, and less protective abdominal musculature than adults. They suffer relatively more solid organ injuries due to both blunt and penetrating mechanisms. Approximately one-third of children with major trauma will have significant intraperitoneal injuries.

Classification of abdominal injuries. Based on clinical exam, abdominal trauma can be included in one of the following syndromes:

- Internal bleeding syndrome - clinical signs of hemorrhagic shock (pale skin, psychomotor agitation, intense thirst, tachycardia, hypotension) dominate, along with signs of intraperitoneal effusion. It is caused by injury to the intraperitoneal parenchymal organs (liver, spleen) or by mesenteric rupture. Peritoneal puncture, and if necessary peritoneal lavage, confirms the diagnostic, along with ultrasound examination.
- Peritoneal irritation syndrome result rupture of hallow abdominal organs and is characterized by muscular rigidity or contracture, which is a pathognomonic sign.
- Mixed syndrome is characterized by simultaneous injury to both a parenchymal and a hallow (cavitary) organ. There may be other forms of abdominal trauma manifestation (two-stage intraperitoneal hemorrhage, pancreatic contusions, duodenal contusions), each with specific symptoms.

Clinical diagnosis. Physical examination of the child's abdomen begins during the secondary investigation. The stomach and bladder must be decompressed, and abrasions and contusions of the chest must be looked for.

The examination may reveal traumatic marks and parietal bruising. The participation of the abdomen in respiratory movements and the presence of any asymmetries are assessed. Persistent and gentle palpation of the abdomen may reveal significant tenderness if the level of consciousness is not affected and there are no concomitant injuries. Palpation of the abdomen is first performed superficially (tenderness, wall hematomas, Morel-Lavalle seroma), then deeply (muscular guarding, abdominal contracture, acute urinary retention). Percussion may reveal tympany with loss of hepatic dullness, indicating pneumoperitoneum from rupture of a hallow (cavitary) organ or displaced dullness on the flanks, with proximal concavity, associated with the wave sign in the case of peritoneal effusion. A rectal examination should be performed in both boys and girls. It detects any lesions of the rectum and provides information about the condition of the peritoneum, suggested by the contents and sensitivity of the Douglas pouch.

Paraclinical exam. Focused Assessment with Sonography for Trauma (FAST) has become a useful part of the initial assessment. It can be performed in 3 minutes, is noninvasive, portable, can be performed during resuscitation, and does not irradiate. FAST assesses free intraperitoneal fluid, which often coincides with intra-abdominal injuries. Computed tomography (CT) remains the gold standard for diagnosing abdominal injuries. Paracentesis can quickly determine or rule out the presence of intraperitoneal hemorrhage. This diagnostic tool is used less frequently with the increasing use of FAST and abdominal CT in many traumatology centers.

33.1. LIVER TRAUMA

The clinical picture may include pain in the right hypochondrium radiating to the right shoulder, abdominal distension, abdominal dullness, signs of hemorrhagic shock, absence of bowel sounds, hemobilia – manifested as jaundice, pain in the right hypochondrium, and hypovolemic disturbance of intestine (HDI). Ultrasound (conventional, FAST, CEUS) reveals the lesion and hemoperitoneum. Contrast CT is the imaging method of choice, providing detailed images of the lesion in hemodynamically stable patients (Figure 33.1). Transaminases are elevated.

Classification of liver trauma:

I. Subcapsular hematoma <10% of surface area, intraparenchymal rupture <1 cm
II. Subcapsular hematoma 10-50% of the surface, rupture 1-3 cm of the parenchyma without involvement of the trabecular vessels
III. Subcapsular hematoma >50% of the surface, parenchymal hematoma >5 cm, rupture >3 cm in the parenchyma with involvement of the trabecular vessels
IV. Deep rupture involving the hilar vessels or segmentation with devascularization of 25%–75% of the organ, or 1–3 segments of a single lobe
V. Parenchymal laceration >75% of a single hepatic lobe, >3 juxtahepatic vessels, hepatic avulsion

Approximately 90-95% of liver injuries can be treated conservatively. The patient is monitored in the ICU (BP, pulse, RR, SpO2, urine output). A 20 mL/kg crystalloid infusion is administered, along with broad-spectrum antibiotic therapy and blood transfusion in case of bleeding. These patients must remain in bed rest, and are followed up by frequent abdominal

examinations, serial blood counts, and may require repeated imaging studies, such as CT or ultrasound. Surgical treatment is instituted in cases of hemodynamic instability and signs of biliary peritonitis. The surgical options include median laparotomy, suture of the hepatic parenchyma with thick absorbable sutures, lobectomies, controlled resections, selective embolization of the hepatic artery, or atriocaval shunt. Postoperatively, antibiotic treatment, rebalancing infusions, total parenteral nutrition until transit resumes, and relative physical rest are instituted.

33.2. SPLENIC TRAUMA

The spleen is the abdominal organ most frequently affected by trauma. Treatment may be surgical or conservative, depending on the severity of the injury. Diagnosis is based on clinical and paraclinical examination (ultrasound, abdominal X-ray in standing position, CT scan).

Classification of spleen injuries

I. Subcapsular hematoma <10% of the surface, intraparenchymal rupture <1 cm
II. Subcapsular hematoma 10-50% of the surface, rupture 1-3 cm of the parenchyma without involvement of the trabecular vessels
III. Subcapsular hematoma >50% of the surface, parenchymal hematoma >5 cm, rupture >3 cm in the parenchyma with involvement of the trabecular vessels
IV. Deep rupture involving hilar or segmental vessels with devascularization of 25%-75% of the organ, or 1-3 segments of a single lobe
V. Parenchymal laceration >75% of a single splenic lobe, >3 juxtasplenic vessels, splenic avulsion (Fig. 33.2)

Non-surgical (conservative) treatment is possible in approximately 90% of cases (when the patient is hemodynamically stable). The patient is monitored in the ICU (BP, pulse, RR, SpO2, diuresis). A 20 mL/kg crystalloid infusion, broad- spectrum antibiotic therapy and blood transfusion in case of hemorrhage are instituted. Surgical treatment is performed in cases of hemodynamic instability (BP <90 mmHg, weak pulse, heart rate >90 bpm, transfusion >40 mL/kgc). It consists of median laparotomy, hemostasis using thick absorbable sutures, splenectomy, or upper or lower polar splenectomy. In case of splenectomy, auto-transplant of splenic tissue is indicated. Postoperatively, high-dose antibiotic therapy will be instituted and immunization for Pneumococcus species, Hemophilus, and Meningococcus species will be performed.

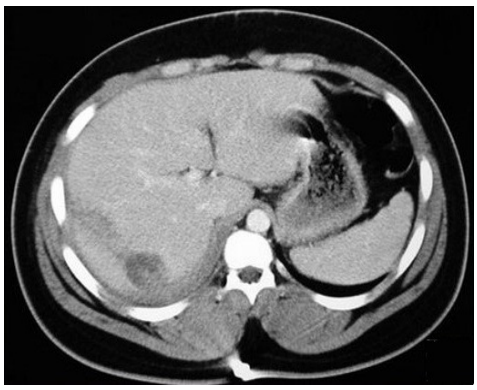


Fig. 33.1. Liver rupture with subcapsular hematoma segments VI-VII

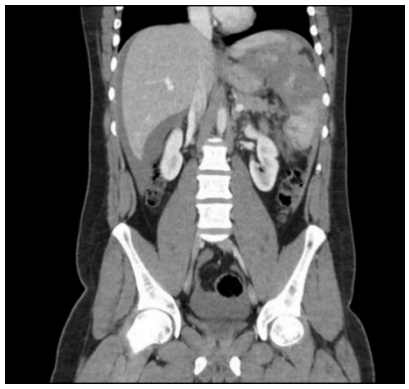


Fig. 33.2. Grade V splenic rupture

33.3. PANCREATIC TRAUMA

Pancreatic trauma is rare in children. Often, other abdominal organs (duodenum, liver, spleen) are also involved. The initial clinical picture has few symptoms, followed by pain of varying intensity, abdominal distension,

and ileus. Lipase and amylase enzymes are elevated, and imaging investigations (CT, ERCP, ultrasound) reveal the lesion.

Classification of pancreatic trauma

I. Small hematoma without duct involvement or superficial laceration without duct involvement
II. Large hematoma without duct involvement or tissue loss, or large laceration without duct involvement or tissue loss
III. Distal laceration or parenchymal laceration with duct involvement
IV. Proximal laceration or parenchymal trauma with involvement of the ampulla
V. Massive laceration of the head of the pancreas

Treatment is conservative for all patients who are hemodynamically stable and show no signs of peritonitis. Surgical treatment is performed on hemodynamically stable patients if they have fever, pain, prolonged ileus, and elevated amylase and lipase levels. The surgical intervention consists of median laparotomy and exploration of the pancreas by detaching the duodenum and opening the gastrocolic ligament; excision of devitalized tissue and drainage, caudal pancreatectomy +/- splenectomy, pancreatojejunostomy, cephalic duodeno-pancreatectomy. Pancreatic pseudocyst is a complication of pancreatic trauma. It occurs more frequently after non- surgical treatment. The clinical picture is represented by recurrent epigastric pain, vomiting, and palpable tumors in the epigastrium. Amylases may increase and the cyst can be detected by ultrasound. It can resolve spontaneously. Those that do not decrease in size but grow or become infected require surgical treatment (ultrasound-guided puncture, cysto-digestive anastomosis, external drainage).

33.4. TRAUMATIC PERFORATION OF CAVITARY ORGANS

The most common causes are penetrating wounds, shearing between mobile and fixed segments, compression of full viscera, or crushing of the spine. The clinical picture is that of peritonitis due to rapid contamination of the peritoneal cavity. Paraclinical assessment may reveal increased inflammatory markers (CRP, ESR), serum amylase (intestinal lesion), pneumoperitoneum (plain abdominal X-ray), peritoneal effusion (abdominal ultrasound), CT scan reveals the lesion. Traumatic perforation of abdominal cavitary organs is an absolute indication for surgical treatment, except for duodenal hematomas without perforation. Supraumbilical laparotomy (for gastric trauma), subumbilical laparotomy (for intestinal trauma), and supra-subumbilical laparotomy (for uncertain or multiple injuries) are performed surgical interventions.

Gastric injuries typically occur through the greater curvature of the stomach, with non-linear lacerations being more common. Treatment involves debridement, hemostasis, debridement of devitalized tissue, and two-layers suturing.

Duodenal lesions. Hematoma without perforation is treated conservatively. In the case of linear ruptures, double-layer suturing is performed. In the case of transverse ruptures, duodeno-duodeno anastomosis, latero-lateral gastro- jejuno anastomosis, and pyloric ligation, cephalic duodeno-pancreatectomy are performed.

Small intestine injuries are managed by double-layer suture (for simple injuries), resection with end-to-end anastomosis (for complex injuries), and stoma formation (for old injuries and those with peritonitis).

Colon lesions require surgical treatment, which consists of double-layer suture with protective superimposed ileostomy/colostomy or stoma at the wound site.

33.5. VASCULAR INJURIES IN THE MESENTERY ROOT

Intraoperatively, extensive hematoma between the mesenteric layers is suggestive. Ligation of the vessel may be sufficient. In case of injury to the main mesenteric axis, suture of the vessel is performed. It is possible to ligate the superior mesenteric artery in children, as enteral ischemia occurs exceptionally due to rich anastomoses and the absence of atherosclerosis. Reoperation at 48 hours allows for assessment of intestinal viability. This avoids extensive bowel resection and the onset of short bowel syndrome.

Chapter 34. RENO-URINARY TRAUMA

34.1. RENAL LESIONS

Incidence. It ranks third in frequency, after liver and spleen trauma. It accounts for approximately 25% of abdominal trauma and is more common in males.

Mechanism of occurrence.

- Contusions (80-90%) due to traffic accidents, falls from height, crushing
- Penetrating wounds, much less common in children
- Iatrogenic, during various medical procedures.

Children have several anatomical features that favor the occurrence of renal trauma:

- Less perirenal fat
- More elastic ribs, energy is transmitted more easily to deeper planes
- The kidney occupies proportionally more space than in adults
- Fetal lobulation in small children favors parenchyma ruptures
- Presence of renal shape/position abnormalities in (1–5%)

Pathological anatomy. According to the AAST (American Association for Surgery of Trauma), renal trauma is classified (Fig. 32.1):

I. Renal contusion (50–85%)
II. Laceration/subcapsular hematoma/ perirenal hematoma ≤1 cm (8–11%)
III. Parenchymal lacerations/peripheral hematoma >1 cm deep but <5 cm
IV. Rupture of segmental or hilar vessels, devascularization of >25%
V. Vascular lesions of the renal pedicle (2–4%)

Clinical picture. In the context of a history of trauma, the patient may present with low back pain, palpable mass in the flank or lower back, hematuria, or signs of hemorrhagic shock (hypotension, tachycardia, altered consciousness). Hematuria is the cardinal clinical sign in cases of renal-urinary trauma. Renal trauma is often associated with injuries to other organs and systems in cases of polytrauma. In this case, the clinical picture is complemented by symptoms or signs due to associated injuries.

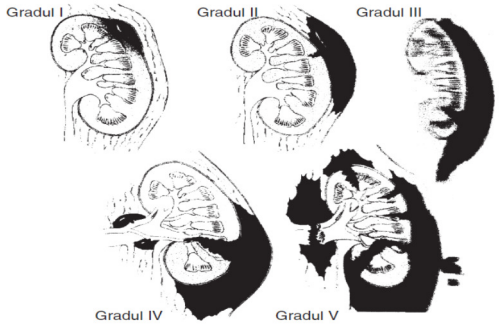


Fig. 34.1. AAST (American Association for Surgery of Trauma) classification of renal trauma

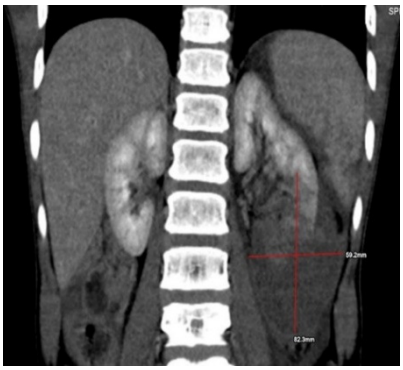


Fig. 34.2. Lower renal pole laceration pole of the kidney

Paraclinical picture

- Laboratory tests reveal posthemorrhagic anemia and hematuria.
- Imaging:

- Abdominal X-ray: blurring of the psoas shadow, possible rib fractures.
- FAST ultrasound: renal lesions, presence of perirenal effusion
- Contrast CT is the imaging investigation of choice in the evaluation of abdominal trauma in hemodynamically stable patients (Fig. 32.2)
- Angiography, less commonly used (allows selective percutaneous embolization)

Positive diagnosis is based on medical history, clinical examination, and laboratory tests.

Differential diagnosis. Kidney ruptures should be considered as a differential diagnosis in any case of road accident or trauma due to a fall from height.

Treatment. Several factors are considered when determining the therapeutic approach: the severity of the injury, the presence of injuries to other organs (multiple trauma), and hemodynamic stability. Non-surgical treatment is preferred for grade I, II, and III injuries. It consists of hydro-electrolytic rebalancing, antibiotic therapy, urinary drainage (Foley catheter), dynamic monitoring of pulse, blood pressure, hematocrit, and hemoglobin, and physical rest for 2–4 weeks. Treatment is surgical in grade III injuries if the initial conservative treatment is unsuccessful (hemodynamic instability with urine leakage into the perirenal space, renal parenchymal infarction or secondary hydronephrosis). In grade IV and V injuries, treatment is emergency surgery from the beginning. Surgical options are laceration suturing, partial or total nephrectomy.

Complications include renal hemorrhage due to two-stage rupture of a renal hematoma, renal/perirenal abscess, formation of a perirenal hematoma, urinary tract obstruction, secondary renovascular hypertension (1–3%).

34.2. URETERAL TRAUMA

These are extremely rare in children and most often occur because of stab wounds or are associated with renal trauma. Another cause is iatrogenic injury involving accidental ligation or incision.

Clinically, the patient exhibit pain, hematuria, urine extravasation into the peritoneal cavity or retroperitoneum, or hydronephrosis. The diagnostic is confirmed by imaging: urography, CT, or MRI with contrast.

Treatment is surgical, involving suturing of the ureter or, in cases with a long segment of devitalized ureter, reconstruction of the ureter using a segment of the digestive tract. As an alternative to surgical treatment, endourological treatment can be attempted by placing a JJ stent.

34.3. URETHRAL TRAUMA

They are rare, accounting for approximately 1.5% of all pediatric injuries, and occur almost exclusively in males. The mechanisms of injury may include penetrating scrotal or perineal wounds, pelvic fractures, or iatrogenic causes (bladder catheterization, cystoscopy).

There are two forms: complete ruptures (25%) and incomplete ruptures (65–75%). The injury can be located at any level: prostatic urethra, membranous urethra (through shearing at the uro-genital diaphragm), bulbar urethra, or penile urethra.

Clinical picture. Patients present with perineal pain, hematuria, acute urinary retention, perineal ecchymosis in a butterfly pattern, pelvic fractures, and rectal examination reveals prostate enlargement and pain on palpation.

Voiding cystography is useful for detecting contrast extravasation, while ultrasound valuable for evaluating perineal hematomas (Figure 34.3).

Treatment is non-surgical in most cases. A Foley catheter is inserted to bypass the injury for 10-14 days until the injury heals. If possible, a cystoscopy is performed to assess the severity of the injury before inserting the catheter. In cases of urethral rupture with urinary retention, an emergency cystostomy is performed. Surgical treatment is indicated in cases of complete rupture, cases with concomitant rectal injuries, massive perineal hematoma, or bladder neck injuries. Complications include urethral stricture, impotence or incontinence, urinary tract infections, lithiasis.

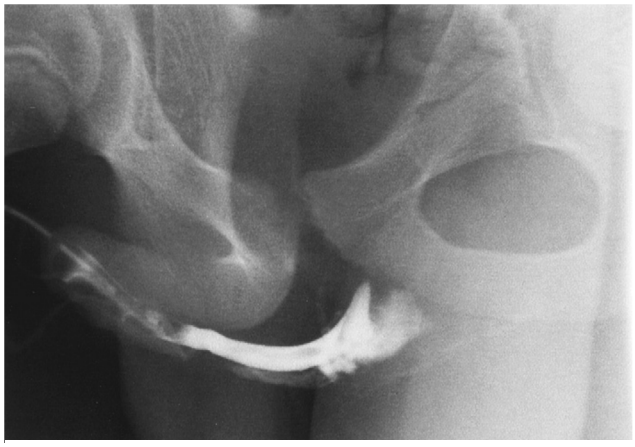


Fig. 34.3. Urethrography, side view. Urethral rupture

Chapter 35. BURNS

Definition. A burn is a traumatic injury to the skin or mucous membranes caused by external physical or chemical agents, leading to protein coagulation, accompanied by systemic nervous, vascular, metabolic, and humoral reactions. It is one of the most common causes of traumatic injury in children, with extremely serious medical, surgical, psychological, and social consequences, with the potential for disability and risk to life.

Pathophysiology. A burn wound is a dynamic injury that cannot be viewed separately from its systemic consequences. Regardless of how it is produced, the essential injury factors are the temperature to which the skin is exposed and the duration of exposure. A burn injury has three zones (Jackson):

- Coagulation zone (central)
- Stasis around the coagulation zone, cells are partially damaged, initially viable but the necrosis may progress
- Hyperemia at the periphery. Minimal cell damage, marked vasodilation, and increased blood flow

Burns are not confined to the skin; they represent a systemic condition that can have serious, potentially life-threatening consequences. The hormonal and metabolic response to burns, depending on severity, can cause local and/or systemic manifestations. Vasoactive mediators, catecholamines, and inflammatory markers lead to a local and systemic phenomenon of capillary leak with protein loss and interstitial edema and the onset of systemic inflammatory response syndrome (SIRS).

A hypermetabolic state occurs due to increased energy consumption and catabolism and decreased anabolic hormones. All these leads to loss of

muscle protein, bone mineral density, and bone mineral content. Thermo-regulation is disrupted and cannot be achieved. When the burned area exceeds 40% of the total body surface, myocardial depression and arterial hypotension can occur. If the respiratory tract is involved, acute respiratory distress syndrome (ARDS) may occur. Gastrointestinal system dysfunction may occur and intestinal bacterial translocation is the major cause of septic shock in patients with severe burns. As the condition progresses, renal and hepatic dysfunction occur due to decreased perfusion, secondary to hypotension caused by volume loss. Predisposition to infection is due to the loss of the skin's barrier function and the immunosuppressive effect of the burn.

Classification of burns

- By mechanism of production

- thermal burns through contact with hot fluids, flames/explosions, hot objects
- electrical burns (electrocution)
- chemical burns
- radiation injuries

- By depth (degree of burn):

I. Superficial, affecting the epidermis

II. Partial thickness

II.A. Partial superficial, the epidermis and superficial dermis are affected

II.B. Partial deep, epidermis and deep dermis affected

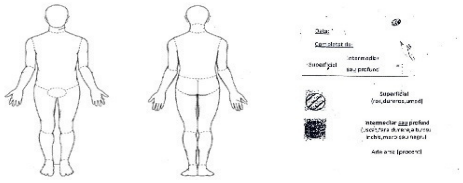
III. Full thickness, all layers of the skin are affected: the epidermis, dermis, and hypodermis

IV. Subcutaneous, complete destruction of the integument and extension to the subcutaneous cellular tissue, muscle, and bone

Assessment of the burned area. The burned area is the essential element in assessing the severity, prognosis, and treatment plan for burns. There is a direct correlation between the burned area and the risk of death. The patient's age is an important prognostic factor for the burns of the same depth and extent; burns are more serious in individuals at extremes of age - children under 3 years old, and adults over 60 years. A burn should always be considered serious in infants and the elderly. The burned area is calculated based on the following formulas:

Wallace formula, used for adults and adolescents over 15 years of age, calculates the burned area based on the "rule of 9."

Lund and Browder formula is used for children, because the ratio between different body segments changes with age (Figure 35.1).



ZONA	1 AN	1-4 ANI	5-9 ANI	10-14 ANI	15 ANI	Adult	Superficial	Intermediar ea adancime
CAP	19	17	13	11	9	7		
GAT	2	2	2	2	2	2		
TRUNCHI ANTERIOR	13	13	13	13	13	13		
TRUNCHI POSTERIOR	13	13	13	13	13	13		
FESA DREAPTA	2.5	2.5	2.5	2.5	2.5	2.5		
FESA STANGA	2.5	2.5	2.5	2.5	2.5	2.5		
ZONA GENITALA	1	1	1	1	1	1		
BRAT DREPT	4	4	4	4	4	4		
BRAT STANG	4	4	4	4	4	4		
ANTEBRAT DREPT	3	3	3	3	3	3		
ANTEBRAT STANG	3	3	3	3	3	3		
MANA DREAPTA	2.5	2.5	2.5	2.5	2.5	2.5		
MANA STANGA	2.5	2.5	2.5	2.5	2.5	2.5		
COAPSA DREAPTA	5.5	6.5	8	8.5	9	9.5		
COAPSA STANGA	5.5	6.5	8	8.5	9	9.5		
GAMBA DREAPTA	5	5	5.5	6	6.5	7		
GAMBA STANGA	5	5	5.5	6	6.5	7		
PICIOR DREPT	3.5	3.5	3.5	3.5	3.5	3.5		
PICIOR STANG	3.5	3.5	3.5	3.5	3.5	3.5		
TOTAL								

Fig. 35.1. Lund şi Browder chart

In burns associated with smoke inhalation and/or burns to the respiratory tract, the burned area is estimated by a plus of 10-15%.

In addition to the surface area, the following parameters should be also considered when assessing the severity of the burn:

- Location. Burns located on the face, hands, feet, perineum, airways, and those with circular distribution are very serious. Circular burns on the extremities carry a risk of peripheral ischemia. Full thickness burns with circular distribution in the cervical region and/or trunk carry a risk of edema and laryngeal obstruction and respiratory failure.
- Depth of the burn. The depth must be assessed dynamically (2-5 days) by trained personnel. A burn affecting the entire thickness of the dermis should be considered serious, with surgical indication, even in the case of relatively small areas. Certain etiological agents generate predominantly deep lesions such as electric current, flame, explosion, chemicals, hot oil, viscous substances, contact with hot surfaces. Full-thickness burns and deep partial-thickness burns are considered serious in terms of the potential risk of permanent scarring, with functional and cosmetic implications.
- Associated injuries. Fractures, head injuries, internal injuries caused by falling from a height (high-voltage electrocution) or by projection (explosion, domestic voltage electrocution), carbon monoxide poisoning (fire in an enclosed space) must be assessed during the initial examination. Associated conditions must be considered and treated concurrently from the beginning.

The prognostic index has predictive value for the evolution of the burn and is calculated using the formula: PI = Burned area (%) x Degree of burn.

- PI = 50, burn without complications
- PI = 50-100, uncomplicated/possible complications
- PI = 100-150, possible recovery/death
- PI = 150-200, majority of deaths
- PI > 200, exceptional recovery

Management of burn patients is complex.

First aid involves freeing the patient, interrupting contact with the harmful agent, removing burnt clothing and jewelry, and cooling the burnt area for 20 minutes with water at room temperature (no less than 8°C, no ice). The patient can be covered with clean sheets. Essential initial measures include supplemental oxygen for open flame burns, orotracheal intubation (OTI) for suspected airway injury, establishment of venous access (can also be inserted through burned skin), urinary catheterization, analgesia, ATPA (Tetanus prophylaxis with tetanus toxoid) for those who have not been immunized in the last 5 years, tetanus immunoglobulin in those without primary immunization, emergency wound management consists of cleaning, debridement, and application of topical agents and dressing.

Table 35.1 shows the treatment of burns according to the severity of the burn.

Prognosis is closely related to the severity of the burn. Local complications such as hypertrophic scars (32-72%), contractures that impede motor functions, chronic or general pain, including death, may occur.

Table 35.1. Treatment strategy for burns in children according to severity

Degree	Layers affected	Time to healing	Treatment	
			Local	General
I. Superficial	Epidermis	Re-epithelialization 5 days	Room temperature water 20 min Cleaning with saline solution Local topical	Oral rehydration Antihistamine Mild analgesia
IIa. Partial superficial	Epidermis Superficial dermis	Re-epithelialization 1-2 weeks	Room temperature water for 20 minutes Cleaning with saline solution and disinfectant solution Debridement and removal of blisters Application of compress antimicrobial ointment Sterile dressing	High-protein, high-calo diet Oral rehydration ATPA/ VTA Antibiotic antihistamine, gastric protector, analgesic, vitamin C
IIb. Deep parti	Epidermis Deep dermis (reticular)	Re-epithelialization 2-5 weeks Skin graft sometimes necessa	Room temperature water 20 min Cleaning with saline solution and disinfectant solution Debridemer and removal of blisters Application of compress, antimicrobial ointment Grafting if spontaneous epithelialization does not occur Sterile dressing	High-protein, high-c a l o r y diet Oral rehydration ATPA or VTA Antibiotic, antihistamine, gastric protector, analgesic, vitamin C
III. Total	Epidermis, Dermis, Hypodermis	Prolonged requires surgical interventio for closure	Excisional debridement and early grafting OR Conservative treatment until granulation of the area, approximately 21 days, followe by skin grafting	High-protein, high-calorie diet Oral rehydration ATPA or VTA Antibiotic, antihistamine, gastric protector, analgesic, vitamin C
IV. Subcutaneous	Epidermis Dermis Hypodermis Muscle Bone	Does not heal spontaneously Surgical interventi reconstruction	Complex Surgical debridement, grafting, amputations, etc.	Admission to intensive care

Chapter 36. SPECIFIC FRACTURES IN CHILDREN

The growing skeleton has certain particularities in children:

- Growth plates are a weak area in the structure of long bones that are prone to specific fractures such as Salter-Harris fractures.
- The periosteum is much thicker and more resistant. It plays an important role in fracture healing.
- Mechanisms of bone growth and repair. The greater osteogenic potential of children influences callus formation and the fracture healing process. The capacity for remodeling and the time required for fracture healing are inversely proportional to the child's age (a younger child will heal faster and compensate for a greater degree of angulation, shortening, or gap in the fracture site through growth).
- The distinct physical properties of immature bone; compared to adult bones, children's bones are more porous, and less mineralized, making them less resistant, but they have higher water content, which make sthem more elastic. The elasticity of bones means that the risk of fractures is much lower than in adults, despite trauma is more common in children.
- Due to these morphological and functional characteristics, children may suffer specific types of traumatic osteoarticular injuries.

36.1. OBSTETRICAL FRACTURES

Obstetrical fractures are injuries sustained during childbirth. Currently, the incidence of obstetric fractures is declining. The causes are maternal-fetal dystocia, forceps delivery, fetal macrosomia, associated pathologies (myelomeningocele, omphalocele, osteogenesis imperfecta), complicated delivery, prolonged labor. The most common obstetric fractures involve the clavicle, humerus, and femur.

- Obstetrical fractures of the clavicle account for 90% of obstetrical fractures. The diagnosis is most often made when the callus has already formed. Clinically, the newborn is restless and cries when the involved upper limb is mobilized. Healing occurs spontaneously in all cases.
- Obstetrical fracture of the humerus. The most common fracture is a separation fracture at the upper epiphysis. Another fracture type is in the middle third of the humerus. It may be associated with brachial plexus palsy. Treatment consists of immobilizing the affected limb on the chest.
- Obstetrical fracture of the femur. The most common fractures are those of femoral diaphysis, while detachment of proximal epiphysis is less common. Clinically, it can be mistaken with hip dislocation, as the affected pelvic limb is shorter and externally rotated. Treatment consists of orthopedic reduction and plaster cast immobilization.

36.2. GREENSTICK FRACTURE

- is an incomplete type of fracture in which bone bends and cracks, resembing a young, flexible tree branch that is breaks partially but not completely. The bone fractures, at the maximum level of convexity only along one side but doesn't snap all the way through (Figure 36.1 A). Treatment consists of orthopedic reduction, breaking of both sides and casting. Radiological follow-up is important. If displacement of the fracture occurs, surgical correction shall be performed.

36.3. PLASTIC DEFORMITY

- is a particular type of fracture in which the diaphysis of long bones, especially in the forearm, becomes angulated without interruption of bone continuity (Figure 36.1.B).

SUBPERIOSTAL FRACTURE is a complete fracture in which the bone fragments remain end-to-end due to the integrity of the periosteum (Figure 36.1. C). Treatment consists of plaster cast immobilization.

36.4. COMPRESSION FRACTURE

- is also a subperiosteal fracture, affecting the metaphyseal area of long bones, in which compression of the bone trabeculae occurs, resulting in bone compaction with bulging of one portion of the cortex and collapse of another. Treatment is orthopedic and consists of immobilization with a plaster cast (Figure 36.1. D).

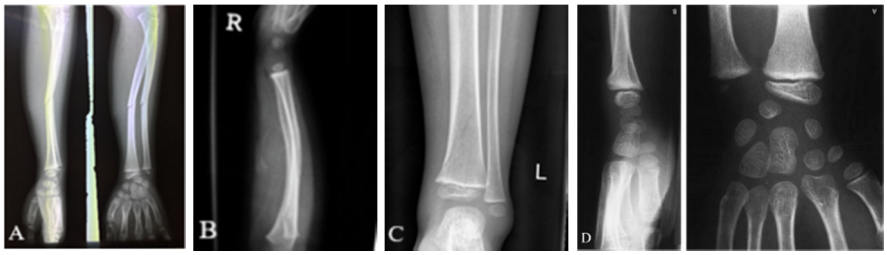


Fig. 36.1. A Greenstick fracture; **B** Plastic deformation; **C** Subperiosteal fracture; **D** Compression fracture

36.5. GROWTH PLATE FRACTURES

There are 5 types according to the Salter-Harris classification (Figure 36.2)

Type I. Simple epiphyseal separation

Type II. Epiphyseal separation with fracture line at the metaphysis

Type III. Epiphyseal separation with fracture line at the epiphysis

Type IV. Epiphyseal separation with fracture line at the epiphysis and metaphysis

Type V. Growth cartilage is crushed between the epiphysis and metaphysis, with irreversible destruction of the cartilage

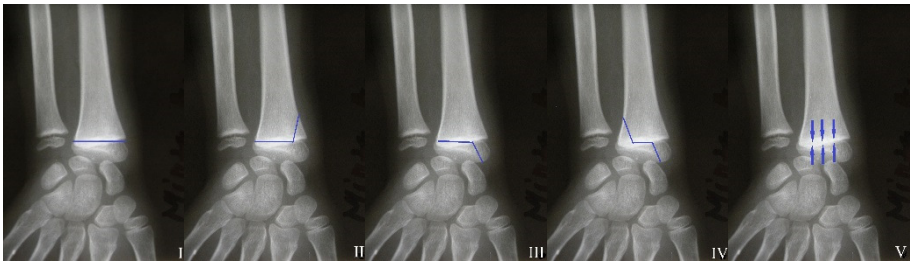


Fig. 36.2. Salter -Harris classification

Chapter 37. SUPRACONDYLAR FRACTURE OF THE HUMERUS

Definition. A supracondylar fracture is a break in the bone at the distal metaphysis of the humerus in the supracondylar portion. The mechanism is most often a fall on the upper limb with an extended elbow.

Epidemiology. It is the most common fracture of the elbow and one of the most common fractures in childhood. Most supracondylar fractures are extension fractures (95-98%). This type of fracture occurs most frequently in the 5-7 years old age group and affects both sexes in relatively equal proportions.

Etiopathogenesis. Supracondylar fractures are most often caused by an indirect mechanism. Risk factors include activities that involve an increased risk of falls, such as contact sports or activities with a risk of falling from a height. Osteoporosis can also increase susceptibility to fractures in general.

Associated injuries: Neurapraxia (the median and anterior interosseous nerves, radial and ulnar nerves may be affected), vascular damage (brachial artery), compartment syndrome, Volkmann's contracture, ipsilateral forearm fractures, open fractures.

Classification. Based on the mechanism of injury, fractures can be classified as extension fractures (95-98%) and flexion fractures (<5%). Rogers' line on the lateral radiograph is also important for assessment. Normally, it intersects the humeral head at the transition from the middle third to the posterior third (Figure 37.1).

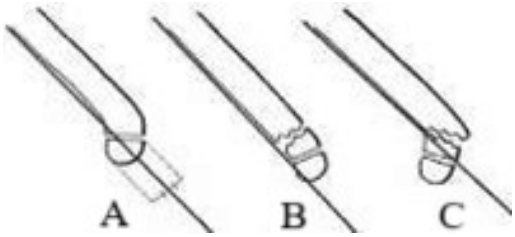


Fig. 37.1. Rogers' line: **A.** Normal, **B.** Extension fracture, **C.** Flexion fracture

Gartland classification (Figure 37.2)

I. Fracture without displacement – usually treated by immobilization in a cast for 3-4 weeks

II. Displacement in a single plane, usually in the sagittal plane – usually treated with orthopedic reduction and percutaneous broaching with Kirschner wires, followed by immobilization in a cast for 3-4 weeks.

III. Displacement in 2 or even 3 planes – usually treated with orthopedic/ open reduction and K-wire pinning, followed by casting for 3-4 weeks

IV. Major displacement of the fractured fragments with marked instability in extension and flexion is usually treated by orthopedic/ open reduction and percutaneous and K-wire pinning, followed by casting for 3-4 weeks.

Clinical diagnosis. Probability and definite signs of fracture are present. It is very important to assess the neurological and vascular status, as well as peripheral motor function.

Radiological diagnostic. Usually by X-rays in 2 views (front and side).

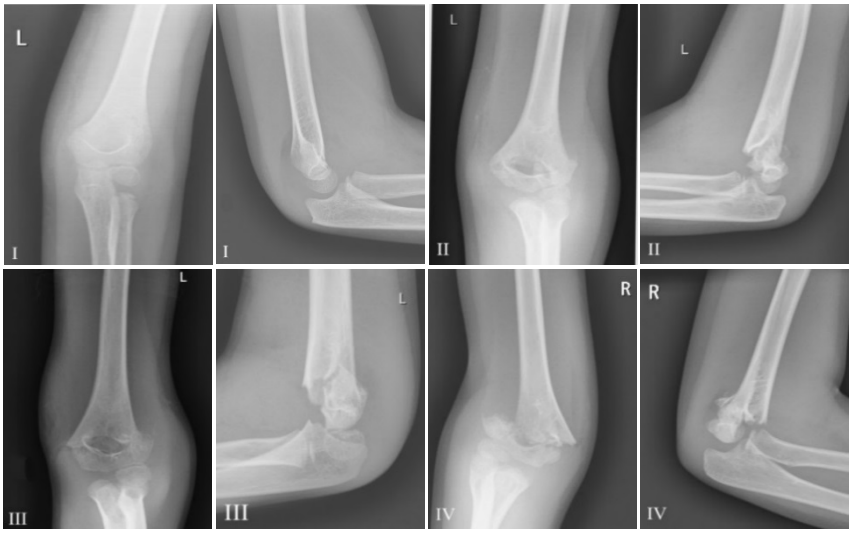


Fig. 37.2. Gartland classification

Conservative treatment. Immobilization in a plaster cast or Blount bandage in type I and possibly type II fractures after adequate orthopedic reduction for a period of 3-4 weeks. Never apply a circular cast or cast in hyperflexion (angle greater than 90°) as a first-line treatment; the cast is completed after remission of edema/healing of surgical wounds.

Surgical treatment. Orthopedic or open reduction and osteosynthesis using one of the following methods: - K-wires in an "X" shape, K-wires only on the radial pillar (to reduce the risk of ulnar nerve damage), external fixator, intramedullary elastic nails inserted anterogradely. For open fractures, choice of treatment is related to the specifics of each type of fracture according to the Gustilo-Anderson classification.

Complications. Ulnar nerve injury (during percutaneous broaching from the ulnar pillar), secondary displacements or rotations, bleeding, infections, osteomyelitis, nerve or vascular damage, muscle and tendon damage, decreased range of motion in the elbow, especially flexion/extension.

PEDIATRIC ORTHOPEDICS

Chapter 38. DEVELOPMENTAL HIP DYSPLASIA

Definitions:

Hip dysplasia or coxofemoral dysplasia includes a spectrum of developmental disorders of the coxofemoral joint that manifest as an abnormal relationship between the femoral head and the acetabulum. If undetected and untreated, it progresses to subluxation and ultimately hip dislocation.

Subluxation involves partial loss of normal contact between the femoral head and the acetabular cavity.

Dislocation involves the complete loss of contact between the femoral head and the acetabular cavity.

Teratological hip dislocation is a different type of congenital hip dislocation, has a different embryogenesis from dysplasia, and occurs when associated with neuromuscular syndromes such as neuromuscular paralysis or arthrogryposis. It is also not reducible by orthopedic maneuvers.

Incidence. Approximately 1.4 out of 1,000 newborns have congenital hip dysplasia, with females being 4-6 times more frequently affected.

Etiology. A clear etiology has not been established, but due to familial aggregation, it is believed to be a multifactorial condition with a genetic predisposition. There are several factors that may be responsible for the development of hip dysplasia: capsuloligamentous hyperlaxity, intrauterine malposition, mechanical factors, or postnatal environmental factors. Maternal hormones that increase the capsuloligamentous laxity necessary for birth

cross the placental barrier may influence the fetus. The more pronounce response if female infants to maternal hormones explains why congenital hip dysplasia is more common in females. Congenital hip dysplasia is related to fetal position in utero, being more common in babies delivered in breech position. When the child stands with one or both knees extended during pelvic presentation, there are changes suggestive of congenital coxofemoral dysplasia. Other risk factors include oligohydramnios, macrosomes, and first-born children. In addition, there is a link between congenital coxofemoral dysplasia and postnatal positioning (parents holding their newborns in full lower limb extension). This tradition is becoming less common, which has also to a slight decrease in the number of cases of congenital coxofemoral dysplasia. Congenital coxofemoral dysplasia is often associated with other conditions, the most common being torticollis and congenital clubfoot.

Clinical picture. The age and severity of the condition determine the clinical presentation of patients with congenital hip dysplasia. Clinical examination is essential in the neonatal period, as radiological examinations at this age do not provide reliable data for diagnostic (ultrasound examination is more useful). The Ortolani and Barlow maneuvers are used for routine clinical examination of the hips. In the Ortolani maneuver, the child is placed in the supine position with the thigh flexed at 90 degrees, and the hand holding the knee in the palm. The middle and index fingers rest on the lateral side of the thigh upon the greater trochanter, and the thumb is on the medial side of the thigh in proximal 1/3. A gentle abduction movement of the thigh is performed with the fingers pressing lightly on the greater trochanter forward and medially. When the femoral head passes over the edge of the acetabulum and reduces into the acetabulum, a click is felt, a proprioceptive and sometimes auditory sensation.

The Ortolani maneuver highlights a dislocated hip. When muscle contractions begin to appear after 7 days of life and increase slowly with age, the Ortolani maneuver becomes increasingly difficult to perform and loses its clinical value.

The Barlow test assesses the occurrence of dislocation. Fixing the patient and grasping the thigh with the contralateral hand are the same as in the Ortolani maneuver. A click is felt as the femoral head comes out of the acetabulum. This occurs when the thigh is in adduction, pushing the proximal femur outward with the thumb and pressing the palm on the knee to push the femoral head posteriorly. The second stage of the Barlow test, which is identical to the Ortolani maneuver, reduces the dislocation caused in the first stage (Figure 38.1).

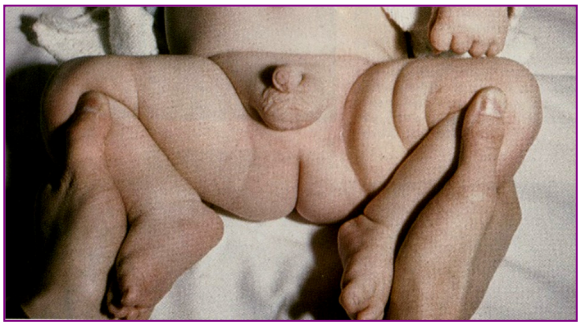


Fig. 38.1. Positioning the infant for the Ortolani maneuver and the Barlow test.

Shortening of the limb, asymmetry of the inguinal folds, gluteal folds, or asymmetry of the vulvar cleft are some of the signs that can be observed in infants with dislocation. Children over one year of age (after they start standing and walking) with hip dislocation exhibit delayed walking, limping, and unilateral or bilateral Trendelenburg gait (the pelvis drops on the unsupported side when standing on one leg, swaying or waddling gait) when the dislocation is bilateral.

Imaging investigations. Hip ultrasound. The hip of a newborn is difficult to evaluate radiologically due to its cartilaginous structure (radiolucent). Graf introduced hip ultrasound for congenital coxofemoral dysplasia in 1980. It allows for an accurate assessment of the relationship between the femoral head and the acetabular cavity. For hip ultrasound, ultrasound machines have specific software settings. Currently, the interpretation of hip ultrasound results is well standardized. The recommended age for performing this investigation is 4-6 weeks (Figure 38.2). All newborns with risk factors for dysplasia, such as a family history of congenital coxofemoral dysplasia, the first newborn, female, birth weight greater than 4000 grams, oligohydramnios, breech presentation, and congenital clubfoot should undergo a hip ultrasound.

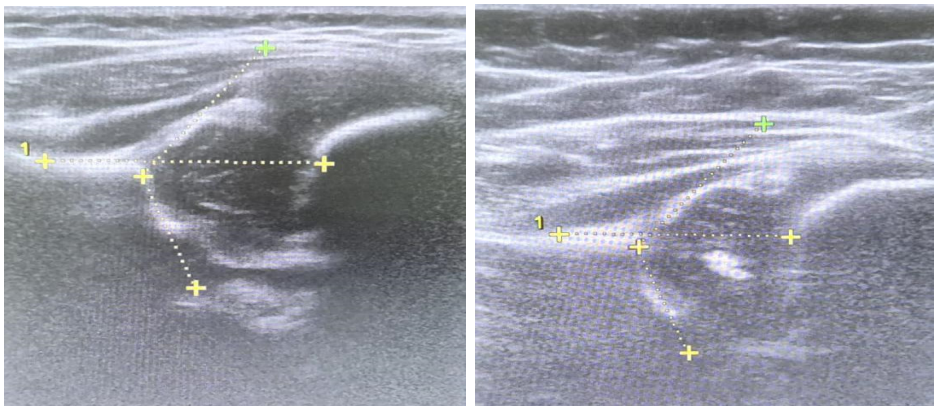


Fig. 38.2. Hip ultrasound at 5 weeks and 5 months of age.

X-ray should be performed with the lower limbs in full extension, with the patella towards zenith, so that the frontal plane of the pelvis is completely parallel to the plane of the radiological table. The cephalic ossification center of the femur becomes visible on X-ray in 66% of children between 4 and 7 months of age, is the one providing accurate information regarding the femoral head position. The radiological signs of hip dysplasia and dislocation:

Interruption of the Shenton's line (imaginary curved line drawn along the inferior border of the superior pubic ramus and along the inferomedial border of the femoral neck) is observed in cases where the nucleus is hypoplastic and appears late.

Ombredanne's quadrants are used to determine the position of the cephalic nucleus (Figure 38.3). By drawing a horizontal line passing through the upper edge of the triradiate cartilages (Hilgenreiner's line) and a vertical line perpendicular to Hilgenreiner's line descending from the most external ossified part of the acetabular roof, four quadrants are obtained (Figure 38.4). The normal position of the femoral head is in the infero-internal quadrant. It is in the infero-external quadrant in subluxation and in the supero-external quadrant in dislocation.

The acetabular angle, also known as Hilgenreiner's index, is angle between the horizontal line and a line tangent to the acetabular roof. It must be less than 30o (40o according to some authors up to one year of age, Figure 38.5).

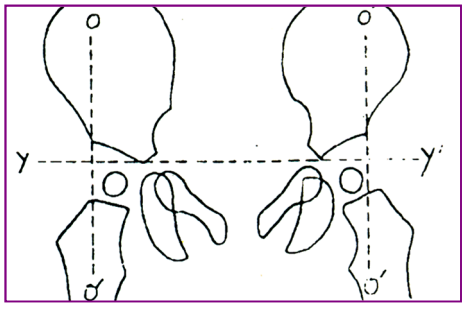


Fig. 38.3. Ombredanne's quadrants

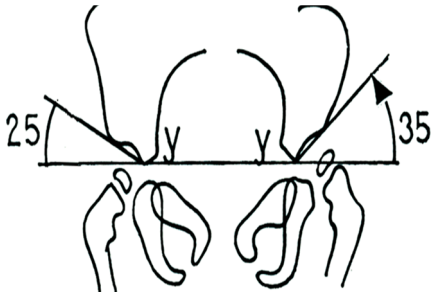


Fig. 38.4. Hilgenreiner's angle

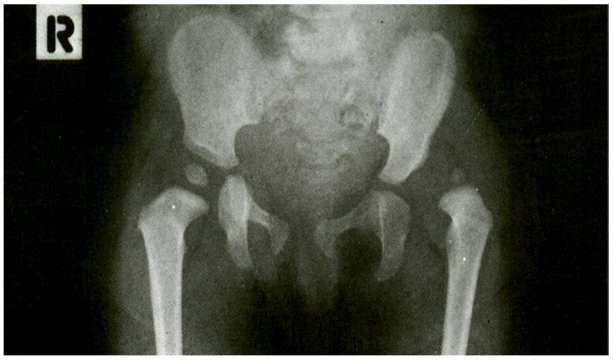


Fig. 38.5. Left coxofemoral dislocation, X-ray

MRI and CT scans are additional imaging tools for investigating congenital coxofemoral dysplasia. They are used mainly for complicated cases in older children, usually in preparation for surgery.

Treatment. Even though this pathology is complicated, early diagnostic allows for appropriate treatment. Treatment of congenital coxofemoral dysplasia in newborns and young infants is relatively simple and involves immobilizing the hip in abduction over the pelvis by using dedicated orthosis like the Pavlik harness. The abduction position is maintained for six weeks allowing the acetabular cavity to deepen and secure a better contain mechanism for the femoral head. The treatment has exceptional results. As the patient's age and the severity of the disease increases, the treatment becomes more complex, requiring surgical treatment over the containment mechanism, even pelvic osteotomies in older children, in combination with orthopedic immobilization.

Progression. This condition can progress to severe locomotor disability if left untreated. This has serious repercussions on quality of life: limping, deformities and ankylosis, chronic pain, scoliosis, early coxarthrosis, and the acquisition of a visible disability that impairs the ability to work, drive a vehicle, or play sports. All of this can be avoided if the condition is correctly recognized and treated.

Chapter 39. LEGG-CALVÉ-PERTHES DISEASE

Definition. Legg-Calvé-Perthes disease is a unilateral or bilateral avascular necrosis of the femoral head with serious consequences over hip mobility.

Epidemiology. The incidence varies from 0.4/100,000 to 29/ 100,000 children under 15 years of age. It occurs usually between the ages of 3 and 12, with the highest incidence between 5 and 7 years of age. The male to female ratio is 3-5:1. Occurs bilaterally in 10-24% of cases and is hereditary in approximately 8-12% of patients. The incidence is higher among the lower socioeconomic class, and the geographical distribution is uneven. The incidence increases with the latitude (low incidence around the equator), and Caucasians are more affected than Asians and Africans.

Etiopathogenesis. Osteonecrosis occurs secondary to interruption of normal blood supply to the femoral head, followed by revascularization with subsequent resorption and remodeling. The etiology is controversial. There are several theories that attempt to explain the disease:

- Vascular theory. The association with abnormal coagulation factors (protein S and protein C deficiencies) has been demonstrated. Thrombophilia has been reported in 50% of patients.
- Inflammatory theory. Repeated acute synovitis can lead to deformities of the femoral head.
- Traumatic theory. Repeated subclinical trauma and mechanical overload lead to bone collapse and repair. The lesions are consecutive to epiphyseal bone resorption, collapse, and subsequent repair.

Evolution. The disease has 4 stages of progression:

I. Initial stage: The blood supply to the femoral head is interrupted, resulting in bone necrosis. This stage lasts several months.

II. Fragmentation stage: The dead bone is resorbed and replaced by softer bone tissue. This stage lasts for 1-2 years. Bone loss causes collapse/deformation of the femoral head and structural deformities associated with the hip joint.

III. Repair stage (re-ossification): Continued re-ossification of the ischemic epiphysis generates the formation of new, stronger bone. This process takes years.

IV. Healing stage (remodeling): In this phase, bone growth is complete. The femoral head has reached its final shape/configuration. The degree of deformity correlates with the prognosis, along with a lasting change in biomechanical function.

Classification. The Catterall classification is based on the radiographic appearance of the femoral epiphysis. There are four grades according to the extent of the epiphyseal lesion:

- Grade I: 0–25% (bone absorption visible on the anterior side of the epiphysis, no sclerosis)
- Grade II: 25–50% (bone absorption on the anterior and central side, sclerosis present)
- Grade III >50% (more than half of the epiphysis involved)
- Grade IV—100% (entire epiphysis)

Clinical diagnostic. The onset is insidious, the first symptom being usually painless limping. Later, they complain of intermittent pain in the hip, knee, or thigh. Physical examination reveals gait disturbances, antalgic limping, Trendelenburg gait, hip stiffness, loss of internal rotation, and abduction. Unequal limb length is a late finding.

Paraclinical diagnostic. Pelvic X-ray shall be performed in both anteroposterior and "frog leg" (Leuvenstein) views. The earliest visible sign is widening of the medial joint space. Subsequently, irregular ossification of the femoral head with sclerotic appearance can be noticed. Later, the crescent sign the Gage sign becomes visible (V-shaped rarefaction on the lateral side of the epiphysis and underlying metaphysis with the tip pointing toward the center of the head), (Figure 39.1). CT can confirm a suspected case and provide information about the degree of involvement of the femoral head. MRI is more sensitive than radiography and highlights change in the femoral capital epiphysis sooner than X-ray (Figure 39.2).

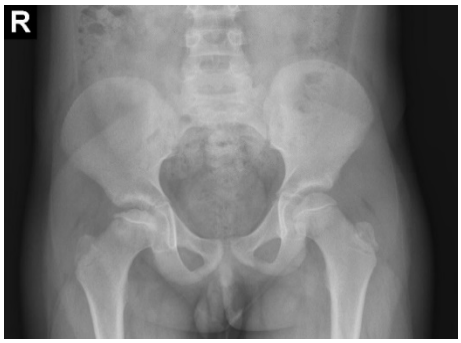


Fig. 39.1. Perthes disease, right hip

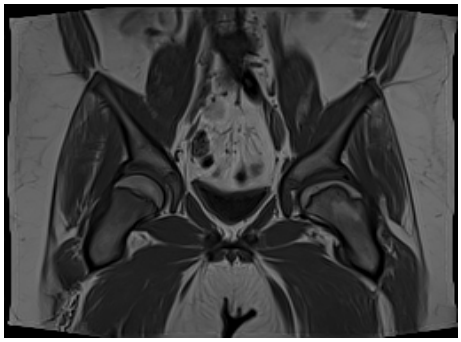


Fig. 39.2. Perthes disease, MRI

Differential diagnosis includes septic arthritis, osteomyelitis, transient synovitis, multiple epiphyseal dysplasia, Meyers dysplasia, proximal femoral epiphysiolysis, and femoral neck fractures.

Treatment. The main objectives of treatment are to maintain the femoral head inside the acetabular cavity, maintain good movement, and reduce further degenerative changes. All patients require periodic clinical and radiographic follow-up until the disease process is complete. Good results

correlate with a spherical femoral head. In 60% of cases surgery is not required. Good results are associated with Catterall group I. Treatment is individualized according to age, extent of injury, and stage of progression.

Conservative treatment involves restriction of physical activity. The hip can be placed in an unloading orthosis to reduce the pressure over the femoral head. Physical therapy (light exercise) is beneficial, especially for children under 8 years of age and adolescents. Nonsteroidal anti-inflammatory drugs are administered in painful forms.

Surgery is recommended for children over 8 years of age with difficult abduction and significant radiological changes. There are several surgical techniques: varus (varisation) osteotomies, valgus (valgisation) osteotomies, pelvic osteotomies. All surgical techniques aim to reposition the femoral head in the acetabulum.

Hip arthroscopy is useful for emergency release of mechanical impairment of the hip in Legg-Calvé-Perthes patients.

Possible complications include deformation of the femoral head: coxa magna, coxa plana; lateral subluxation of the hip (extrusion); premature closure of the growth plate; shortening of the femoral neck; limb inequality; acetabular dysplasia; osteochondritis dissecans; degenerative arthritis.

Prognosis. Children under 6 years of age in incipient state, with normal weight, have the best prognosis towards complete recovery. The recovery period can last 2-5 years. Poor prognosis is associated with female gender, obese patients, decreased hip abduction (adduction contracture). Long-term studies suggest that most patients do well until their fifth or sixth decade of life. Approximately half of patients develop premature coxarthrosis secondary to an aspherical femoral head.

Chapter 40. SLIPPED CAPITAL FEMORAL EPIPHYSIS

Definition. Slipped Capital Femoral Epiphysis is condition where the upper epiphysis of the femur is the displaced (slipped). The femoral head is displaced inferiorly and posteriorly relative to the femoral neck through the growth plate (physis).

Epidemiology. The prevalence is 10.8/100,000 cases. It is more common in boys than in girls, with a ratio of 2-3:1. It affects children in the 10-14 age group, with the average age at diagnosis being 13.5 years for boys and 12 years for girls. The left hip is more affected than the right hip, and in 18% of cases it is bilateral. The incidence increases during the cold periods of the year, being correlated with vitamin D deficiency. It mainly affects obese children or those with skeletal and sexual immaturity.

The **etiopathogenesis** is unknown. It is believed to be caused by an abnormality in the growth of the proximal growth plate of the femur. Mechanical factors and endocrine disturbances are incriminated. Risk factors include obesity, hypothyroidism, hypogonadism, or patients with recurrent transient synovitis.

Diagnosis. Clinically, the patient experiences pain in the affected hip with radiation towards the knee. The patient develops an antalgic gait (Trendelenburg gait). Any patient presenting with knee pain without a history of traumat should be investigated radiologically for femoral head epiphysiolysis. Sometimes it is impossible to bear weight on the affected pelvic limb. On examination, limb inequality is evident, with a characteristic external rotation. Functional impotence occurs, with limitation of flexion, abduction, and internal rotation of the hip. External rotation occurs when the thigh is flexed on the trunk.

Paraclinical: Hip radiography is performed in two views, anteroposterior and Lauvenstein ("frog leg") (Figure 38.1). CT is useful in certain situations for accurate diagnostic and classification. MRI can diagnose "pre-slip" and avascular necrosis of the femoral head. Technetium scintigraphy is used to assess the avascular necrosis.

Classification.

Temporal:

- Acute (15%) - symptoms present for less than 3 weeks
- Chronic (85%) - symptoms lasting for several months
- Acute-on-chronic - acute exacerbation of symptoms lasting for more than 3 weeks

Functional (Loeder) - based on hip stability and load-bearing capacity

- Stable - less severe with good prognosis, no avascular necrosis
- Unstable - severe, poor prognosis, avascular necrosis in 50% of cases.

The objectives of **treatment** are:

- Stabilization of the proximal epiphysis to prevent further displacement
- Closure of the growth plate
- Reducing displacement and stabilizing

Conservative treatment is indicated as a temporary measure until surgery or in cases where epiphysiolysis is related to endocrine conditions like hypothyroidism. Continuous traction for 12 weeks followed by plaster cast immobilization for 8-16 weeks (increased risk of chondrolysis) may be an alternative to surgical treatment.

Surgery is the main therapeutic method. Surgical options are:

- Percutaneous fixation in situ with Kirschner wires or screws
- Reduction and fixation – unstable epiphysiolysis and with high degree of displacement

- Bilateral in situ fixation (prophylactic) indicated in - endocrine disorders in children under 10 years of age
- Proximal femoral osteotomy – extremely painful, functional limitations associated with severe chronic displacement

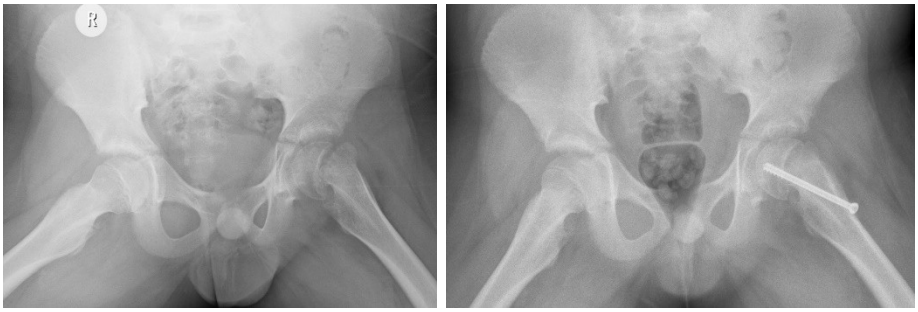


Fig. 40.1. Left femoral head epiphysiolysis, Lauvenstein (frog leg) radiograph, pre- and post-fixation with a canulated screw

Complications: Aseptic necrosis of the femoral head, contralateral femoral head epiphysiolysis, chondrolysis, residual deformity ± limb length discrepancy, progression of proximal femoral epiphysis displacement, secondary coxarthrosis, subtrochanteric fracture associated with inadequate fixation, limited hip movement.

Chapter 41. OSGOOD-SCHLATTER DISEASE

Definition. Osgood-Schlatter disease is avascular necrosis and traction- induced inflammation in the tibial tuberosity in adolescents.

Epidemiology. Boys are more commonly affected than girls. It occurs predominantly in children aged 11-14 who play sports. Bilateral involvement has been observed in approximately 25% of cases.

Etiopathogenesis. It is avascular necrosis and aseptic inflammation, triggered by repetitive stress because of traction exerted by the patellar tendon on the immature, cartilaginous apophysis. This occurs during the sudden growth phase of puberty. Mechanical traction ultimately causes microtrauma in the hormonally weakened growth plate. Current studies have shown a link between "patella alta" (a high riding patella) and Osgood-Schlatter disease.

Clinical diagnosis: the patient complains of pain in the tibial tuberosity area, usually after sporting activity.

Clinical examination reveals tenderness and swelling on palpation of the tibial tuberosity. Pain may also be provoked by asking the patient to lift the lower limb in extension against resistance. The diagnostic is established based on medical history and clinical examination.

Paraclinical diagnostic. Lateral radiography sometimes shows fragmentation of the tibial tuberosity because of the repair process following microtrauma. (Figure 41.1). Fragmentation of this apophysis is occasionally observed in completely asymptomatic patients. No further imaging investigations are necessary.

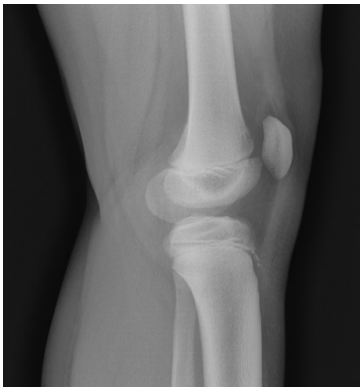


Fig. 41.1. X-ray of the left knee,
Osgood- Schlatter disease

Treatment. Osgood-Schlatter disease is treated conservatively. Parents and children should understand that the healing period can take 1-2 years. In the acute stage, treatment consists of local application of ice, physical therapy, and massage with anti-inflammatory ointments. It is also recommended to stop sports activities during painful periods. Oral anti-inflammatory drugs are not recommended because they have little effect on the course of the disease and must be taken for a very long time. Instead, applying a plaster cast with the knee in extension for 6 weeks may be helpful in cases of very persistent pain. Its effectiveness is since adolescents, who are usually very athletic, are prevented from practicing sports for an extended period. Surgical treatment is only indicated if a loose and irritating fragment in the area where the patellar tendon attaches is still prominent in adult patients. This fragment is removed during surgery.

Chapter 42. SCOLIOSIS

Definition. Scoliosis is a lateral deviation of the spine from the normal vertical line. The term "scoliosis" comes from the Greek "scoliosis," meaning curvature, lateral curvature of the spine.

Pathological anatomy. In the sagittal plane, the spine has a series of physiological curves:

- Cervical lordosis (anterior convexity)
- Thoracic kyphosis (posterior convexity)
- Lumbar lordosis (anterior convexity)
- Sacrococcygeal curvature (posterior convexity)

In the frontal plane, the spine is straight. Scoliosis is a progressive disease characterized by one or more lateral curves of the spine in the frontal plane, associated with rotation of the vertebrae. The deformation of the spine in scoliosis comprises the primary and secondary, compensatory curves. The primary curve is where the lateral deviation of the spine is associated with a rotation of the vertebrae, while the secondary, compensatory, supra- or sub-curves are oriented in the opposite direction. Vertebral rotation is the one producing thoracic cage deformity (rib hump).

Incidence and etiopathogenesis. Scoliosis predominantly affects females (75- 80% of cases) and usually occurs during puberty or prepuberty. Approximately 2-3% of scoliosis cases are congenital and are due to vertebral or costal malformations, while 6-7% of cases are due to other causes (neuromuscular diseases, neurofibromatosis). The remaining 91-92% of scoliosis are idiopathic. The onset is usually at puberty and have an their etiopathogenesis is insufficiently deciphered. There are two categories of factors: intrinsic and extrinsic.

Intrinsic factors:

- Genetic - in 25-30% of patients there is a family history of scoliosis
- Anomalies in the growth of the rachis, the length of the vertebrae is determined by enchondral ossification, and it has recently been demonstrated that there is a difference between the growth of the concave part in relation to the convex part.
- The role of the intervertebral disc. The nucleus pulposus is poorer in proteoglycans and richer in collagen. This creates local conditions that favor spinal deformity.

Extrinsic factors:

- Melatonin - low melatonin level is a marker for the progression of scoliosis
- Blood platelets - platelet abnormalities are associated with the onset of scoliosis
- The nervous system - numerous central or peripheral neurological disorders are implicated in the onset of scoliosis
- Paravertebral muscles play an important role in spinal stability. Muscular imbalance is a possible extrinsic factor in the development of idiopathic scoliosis.
- Osteoporosis - decreased bone density worsens the progression of scoliosis.

Classification

***Etiologically*, scoliosis is divided into structural and non-structural.**

- **Non-structural scoliosis** (functional scoliosis or scoliotic posture) is characterized by a simple lateral deviation of the spine, which is reducible and without vertebral rotation. The causes of deformity can be inequality of the lower limbs, muscle contractures (trauma, fractures), torticollis

(retraction of the sternocleidomastoid muscle), unilateral atrophy of the upper limbs, upper limb amputation, poor posture, scoliosis without apparent causes, scoliosis in young girls.

- **Structural scoliosis** is characterized by persistent curvature, vertebral rotation, vertebral and rib deformities regardless of the patient posture: anterior flexion of the spine, supine position or suspension. They can be:
  - Idiopathic
    - Infantile (0-3 years)
    - Juvenile
    - Adolescent
  - Neuromuscular
    - Neurological: central etiology, paresis, peripheral neuropathies, spinal amyotrophia, arthrogryposis
    - Muscular: myopathies (Duchenne, Becker), hypotonia
  - Congenital malformations
    - Bony malformations: vertebral block, rib fusions, vertebral
    - Neurological: syringomyelia, diastematomyelia
  - Constitutional diseases
    - With vertebral resonance (dwarfism, spondyloepiphyseal dysplasia, mucopolysaccharidosis)
    - Of ectodermal or mesodermal origin (neurofibromatosis, Marfan syndrome, Ehlers-Danlos syndrome)
  - Scoliosis Secondary: post-radiation, post-infectious, post-traumatic, post-operative

Evolutive classification (Ponseti-Cotrel):

- Infantile/congenital scoliosis (0-1 years)
- Infantile scoliosis (1-3 years)
- Juvenile scoliosis (3 years-early puberty)
- Adolescent scoliosis

Topographical classification based on frontal radiography using the "apex" vertebra as a reference point

I. Scoliosis with a single main curve (approximately 70%)

- Cervicothoracic (1%) - apex vertebra C7-T1
- Thoracic (25%) - apex vertebra T2-T11
- Thoracolumbar (20%) - apex vertebra T12-L1
- Lumbar (25%) - L2-L4 tip vertebra

II. Scoliosis with two main curves (double, major) 30%

- Double thoracic scoliosis
- Double thoracolumbar scoliosis

Other classification principles:

- Curvature (convexity) orientation
  - Right thoracic
  - Left lumbar
- Lead wire test
  - Balanced (the thread falls into the interspinal groove)
  - Unbalanced
- Reducibility
  - Reducible
  - Non-reducible

- Number of curves
    - Single scoliosis
    - Double scoliosis
    - Triple scoliosis
- Progression
    - Progressive > 30°- 80-90°, sometimes even 140°
    - Non-progressive at 15-16 years < 30°

Positive diagnosis. A thorough medical history, date of detection, circumstances, family, genetic, or neurological history, previous treatments, and the presence of paravertebral muscle pain are essential.

The clinical exam is performed with the patient undressed and barefoot.

- In orthostasis, assessment should include the condition of the muscles and skin, the shape of the legs and knees, pelvic balance, lower limb length, hip joint movement, and secondary sexual characteristics. Measurement of lateral and sagittal curves, gibbosity height, shoulder, and shoulder blade.
- Examination of the patient in a supine and then in prone position with their knees slightly bent, allows for the assessment of the reducibility of the deformity under load and the persistence of an oblique pelvis.
- Visual inspection from posterior, straight up and then bending forward reveals posterior deformation of the thorax and rib hump.
- Inspection from the front can reveal the deformation of the rib cage or asymmetry of the shoulders and pelvis.
- Balance of the shoulders (normally at the same level).
- Balance of the pelvis: intergluteal cleft and gluteal folds

Paraclinical diagnostic is based on radiography:

- Full spine X-ray in orthostatism, from 2 incidences
- X-ray of the iliac crest and hand for bone age assessment

The most important radiological landmarks are the determination of the upper and lower limits of the curvature. These are the vertebrae with maximum inclinations in relation to the horizontal line.

The magnitude of scoliosis is determined by measuring the Cobb angle, formed by the intersection of the tangent to the upper plateau of the upper limit vertebra with the tangent to the lower plateau of the lower limit vertebra (Figure 42.1.). Vertebral rotation (Nash and Moe method) is made by assessing (classified in 5 degrees) the symmetry and equidistance of the vertebral pedicles against the lateral edges of the vertebral bodies that are most rotated.

The choice of **treatment** depends on the magnitude of the scoliosis:

- 0-30° - physical therapy
- 30° -50°- physical therapy and orthotics (brace)
- > 50° - surgery

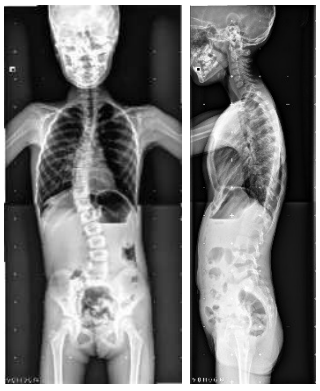


Fig. 42.1. Front and side X-ray of the spine. Idiopathic thoracolumbar scoliosis; Cobb angle of 31^0

Physical therapy is a complex treatment, specific to each type of scoliosis, and personalized. Physical therapy does not correct spinal deformities, but will stop the progression of scoliosis, reduce pain, correct posture, and improve respiratory function. Depending on age the patient and type of scoliosis, stretching maneuvers, guided active mobilization, massage and skin stretching, posture education, corrective dynamic and static exercises to maintain correct posture are alternated.

Orthopedic treatment (orthotics) is indicated for idiopathic scoliosis with curves greater than 30°. The corset prevents the worsening of scoliosis and, in some cases, reduces the curvature of the spine.

Surgical treatment (spinal instrumentation) is indicated for idiopathic scoliosis with curvatures exceeding 45-50°, as well as for neurological or congenital scoliosis. Various types of instrumentation (Harrington, Lugue) are used to fuse the vertebral bodies and correct pathological curvatures of the spine.

Prognosis. The prognosis depends on the type of scoliosis, the severity of the curvature, and the precocity and promptitude diagnostic and appropriate treatment. In idiopathic scoliosis, the progression of spine curvatures is in relation to child's growth. It will worsen as the patient grows. This also means that in most cases scoliosis stabilizes once growth has stopped. Scoliosis in neurological diseases or malformations has poorer functional prognosis and often requires surgical treatment.

Chapter 43. FLAT FOOT

Definition. It is a condition in which one or both feet have a partially or completely flattened arch, causing the entire sole to touch into the contract with the ground (Figure 43.1).

Incidence. It is a common condition, especially in children, but it is not considered pathological under the age of 8. In these children, the arch collapses due to age-related bone and ligament laxity, adipose tissue growth, and neuromuscular immaturity.

Clinical picture. In young children, it is usually asymptomatic. Symptoms usually appear later, in older children and adolescents, or more frequently in adults. Pain occurs in the feet after standing or walking for long periods of time. Clinical examination reveals:

- Absence of the longitudinal arch of the foot
- Calcaneal valgus
- Abduction of the forefoot
- Supination of the forefoot

Classification. There are three types of flat valgus foot:

- Flat foot with shortening of the Achilles tendon
- Flexible flat foot with valgus
- Rigid flat feet

Severity is assessed in three grades:

I. Slight separation of the inner edge from the ground
II. The inner edge is straight and in contact with the ground
III. Convex inner edge due to bulging of the talus head and scaphoid tubercle

Treatment is conservative in most cases. In young children with flexible, asymptomatic flat feet, treatment is not necessary. In most cases, the abnormal position will gradually correct itself by the age of 8-10. For rigid flat feet, older children with or without Achilles tendon shortening are treated with orthopedic boots, heel lifts, or physical therapy. If severe pain occurs during walking, poor school performance, or significant aesthetic impairment, surgery will be necessary. Calcaneotalar arthrodesis is a minimally invasive procedure that stabilizes the medial arch of the foot by inserting an implant (screw) into the "sinus tarsi" through an approximately 1 cm incision.

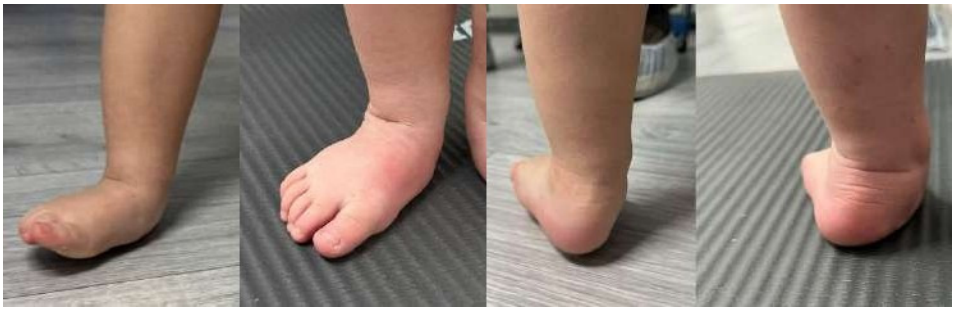


Fig. 43.1. Flat foot

Chapter 44. CLUBFOOT (TALIPES EQUINOVARUS)

Definition. Clubfoot or talipes equinovarus is a congenital deformity in which the foot in an abnormal, permanent position with tip of the foot pointing inward and downward (Figure 44.1).

Epidemiology. It is the most common congenital malformation of the osteoarticular system. Its incidence is 1/1000 newborns, being 2-3 times more common in males than in females. It is bilateral in 50-60% of cases.

Etiology. Most cases of congenital clubfoot are idiopathic. Genetic factors, maternal smoking or diabetes, neurological or mechanical factors are implicated. During the embryonic period, the foot develops normally, but in the second trimester of pregnancy, the foot deviates into its characteristic position. Clubfoot may be associated with neurological diseases (neurological clubfoot) or genetic syndromes (syndromic clubfoot).

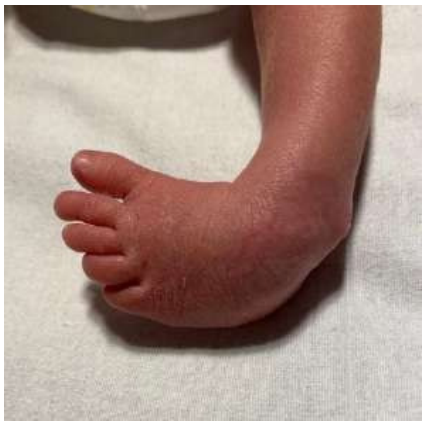


Fig. 44.1. Clubfoot

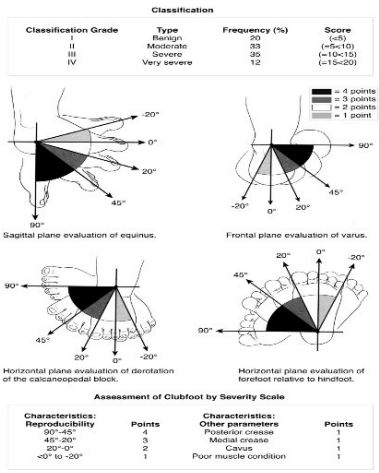


Fig. 44.2. Dimeglio classification

Pathological anatomy. Congenital clubfoot is a complex anomaly characterized by structural changes involve the bones, tendons, ligaments,

and muscles of the foot and lower leg. There is three- dimensional foot deformity which has four components:

- Adduction of the foot on the calf
- Adduction of the rear foot to the forefoot (cavus)
- Internal rotation of the foot relative to the lower leg (varus)
- Plantar flexion of the foot (equinus)

The foot is rigidly fixed in this position, the calf is thinner, and the sole of the foot will be narrower and shorter compared to the foot on the opposite side. The severity of the deformity can be assessed using the Dimeglio classification, with the foot being classified into four degrees (Figure 44.2).

Diagnosis Prenatal diagnosis can be made by ultrasound from the 16th week of gestation (Figure 44.3). Postnatal, the visual inspection reveals the malformation (Figure 44.1). It is necessary to assess the severity using the Dimeglio score or the presence of other congenital anomalies.

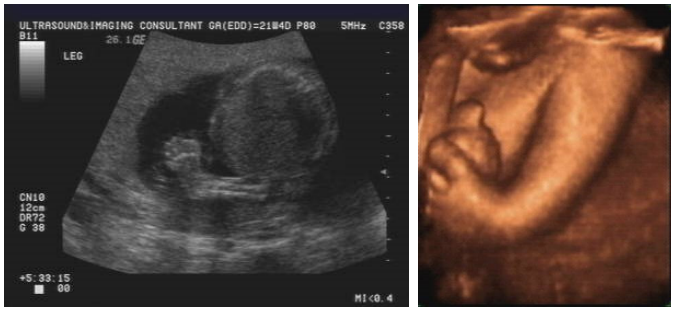


Fig .44.3. Clubfoot in a fetus, 16th week of gestation

Differential diagnosis refers to idiopathic, neurological, or syndromic clubfoot. It can also be made with arthrogryposis, amniotic disease, or posttraumatic clubfoot.

Fig. 44.4. Ponseti method for treating club foot. Sequential cast immobilizations followed by tenotomy of Achile's tendon.

Treatment should be started as soon as possible after birth, during the phase of absolute reducibility. The Ponseti method for treating congenital clubfoot is the first-line treatment and has very good results. This consists of serial plaster casts that will put tension on the structures of the foot and progressively correct the deformity. Plaster casts are applied at 7-day intervals to sequentially correct the deformity, in following order: cavus, adduction, varus, and equinus. After 4-5 successive casts, Achilles tendon tenotomy is performed to achieve definitive correction of the equinus (Figure 44.4). After tenotomy, the feet must be immobilized in a Denis-Brown brace until the age of 4 years, 24 hours a day for the first 3 months and then only during nighttime sleep.

If the treatment is delayed, the foot enters the phase of absolute rigidity and surgery is needed to correct the deformity. Surgical interventions are performed on the soft tissues and bones. Postero-internal release (Codivilla procedure) is the most used surgical procedure.

Prognosis. The natural course of untreated congenital clubfoot is characterised by progressive and rapid worsening of the deformity. If left untreated, congenital clubfoot goes through several stages of progression:

- Absolute reducibility, 14-30 days after birth

- Relative rigidity phase, soft tissue retractions, up to 2-4 years
- Absolute rigidity, with bone deformities and loss of joint mobility

When left untreated, the onset of walking will aggravate this condition. The patient will develop crippling clubfoot. Delaying or failing to initiate appropriate treatment will lead to the permanent distortion of the foot due to the retraction of certain muscle groups and changes in the capsules, ligaments, and bones, which will subsequently prevent correction through external maneuvers (inveterate clubfoot).

Chapter 45. TALUS VALGUS

Definition. It is a pathologic foot position present from birth, characterize by marked eversion and dorsiflexion, with the dorsal surface of the foot in direct contact with the anterior aspect of lower leg.

Epidemiology. The deformity is found in 30-50% of newborns and is more common among girls.

Etiopathogenesis. It occurs due to space restriction in the uterus. Consequently, one or both legs are forced to remain in an exaggerated dorsiflexion position during the last weeks of pregnancy. Risk factors include oligohydramnios, high birth weight, twin pregnancy, or breech presentation. It is frequently associated with developmental hip dysplasia.

Clinical diagnostic. The condition is clinically evident at birth (Figure 45.1). The foot is positioned in exaggerated dorsiflexion and external rotation (eversion). The position of the foot can be easily corrected by passive movements.

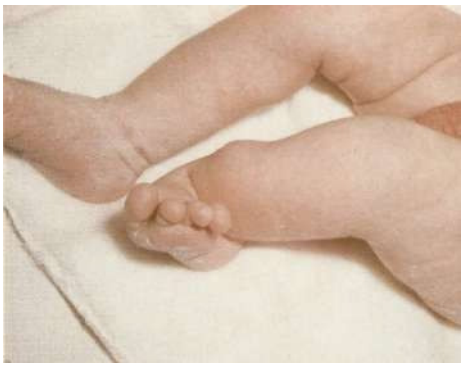


Fig. 45.1. Congenital talus valgus

The **differential diagnosis** includes other forms of congenital foot deformities, especially with vertical talus.

Treatment begins immediately after birth. Mild forms require physical therapy, passive mobilization, and massage. The position of the foot is usually corrected within the first 3-6 months. In cases that do not correct spontaneously or with physical therapy and massage by around 4-6 months of age, immobilization in a cast in the corrected position for 1-2 months is recommended.

Prognosis. In most cases (90-95%), the abnormal position corrects itself spontaneously or through physical therapy by around 6-8 months of age. If left uncorrected, talus valgus can lead to flat feet in older children, which are much more difficult to treat orthopedically and surgically.

Chapter 46. COMPARTMENT SYNDROME

Definition. Compartment syndrome is a surgical emergency and occurs following trauma that causes an acute increase in pressure within a closed muscle compartment, resulting in compromised tissue perfusion, ischemia, and, if untreated, necrosis of the affected tissues.

The **incidence** depends on the population studied and the etiology of compartment syndrome. For example, for leg fractures, the incidence in various studies is between 1 and 9%. The frequency of compartment syndrome increases when there are associated vascular injuries in the affected limb. Compartment syndrome most often affects, in descending order of frequency: the forearm, hand, lower leg, thigh, and foot, however, it can involve any closed fascial compartment of the body, including the abdomen.

Etiopathogenesis. The causes that can lead to the onset of the syndrome can be intrinsic (fractures, crushing, hematomas, animal and insect bites, infections) or extrinsic (external compression by tight circular plaster casts, bandages, tourniquets, circular burns, or the umbilical cord in fetuses).

Compartment syndrome can be classified as acute or chronic. Chronic compartment syndrome occurs predominantly in athletes who engage in high-intensity physical exercise.

Pathophysiology. Compartment refers to a group of muscles, nerves, and blood vessels, all covered by a fascia. Depending on the intrinsic or extrinsic cause, pressure within a closed compartment increases either through the accumulation of edema fluid or through a decrease in compartment volume due to external compression. Normal pressure inside a compartment is less than 10 mmHg. If the pressure reaches 30 mmHg or more, the blood vessels collapse, and tissue ischemia occurs.

Clinical picture. In the context of a predisposing clinical context (fractures, muscle crushing, circular burns of a limb), there is an exacerbation of pain that does not respond to analgesics, increases in intensity, and is exacerbated by muscle mobilization in the affected area. The overlying skin is pale, the patient exhibit paresthesia, weak or absent pulse, paralysis of the affected area (late sign and poor prognosis). The pressure in the compartment can be measured using a dedicated manometry system. This is an objective diagnostic method and a value greater than 30 mmHg is an indication for surgery.

Treatment. To avoid devastating and irreparable complications, early diagnosis and prompt therapeutic measures are necessary. The external etiological factor must be removed if necessary (plaster casts or compression bandages). To relieve elevated pressure within the compartment, decompressive incisions (fasciotomies) are performed. These are oriented longitudinally on the affected segment and include the skin, subcutaneous cellular tissue, and fascia covering the affected muscle compartment. If necessary, non-viable tissue is excised. The incisions are left open and will be closed after the compartment syndrome has been resolved.

Prognosis. Early and correct treatment results in complete functional recovery in approximately 6 months. Delaying treatment for more than 6 hours results in permanent sequelae.

Complications include chronic pain, infection, neurological impairment (paresis), functional limitation, amputations, and even death. The most common and formidable sequela is permanent Volkmann's contracture, which is the result of prolonged ischemia and muscle necrosis in the forearm.

Chapter 47. ACUTE HEMATOGENOUS OSTEOMYELITIS

Definition. Acute osteomyelitis is a bacterial infection of the bone associated with inflammation and bone destruction, characterized by symptoms lasting less than 2 weeks.

Epidemiology. Acute hematogenous osteomyelitis results from hematogenous spread of bacteria to the metaphyseal region of the bone. The incidence is estimated at approximately 8 per 100,000 children each year in developed countries. It is more common in children younger than 5 years of age.

Etiopathogenesis. Microorganisms enter the bone through the nutrient artery and are deposited in the metaphyseal capillaries where they subsequently proliferate. Factors favoring the development of infection are mechanical (local trauma) and infectious agents. Initial bacteremia may occur due to skin injury, infection or even trauma from tooth brushing. The most common infectious agent in children over 1 year of age is Staphylococcus aureus. Group B Streptococcus is the most common organism in newborns. Infants under 1 year of age may have the infection spread across the growth plate, causing osteomyelitis localized in the epiphysis and septic arthritis. The spread within the bone occurs through the Haversian and Volkmann canal systems. It usually affects the metaphysis of long bones (femur, tibia, or humerus).

Clinical diagnosi. Clinical picture depends on age and type of disease. Often, the onset is insidious, with fever, pain, and localized edema. Subsequently, there is limited movement in the affected joint, limping, and refusal to step on or use a limb (pseudo paralysis). Systemic symptoms and signs are more commonly seen in children with methicillin-resistant

Staphylococcus aureus osteomyelitis than in those with methicillin-sensitive Staphylococcus aureus osteomyelitis.

Paraclinical diagnostic. Blood tests performed include complete blood cell count, C-reactive protein (CRP), ESR, procalcitonin, blood culture, bone marrow culture, and PCR. C-reactive protein is useful in diagnosing and monitoring disease progression. CRP values normalize after 7-10 days of treatment, compared to ESR, which normalizes after 1 month or more. The role of procalcitonin as a diagnostic aid in bone and joint infections is unclear. The white blood cell count varies. Blood culture should be collected during a febrile episode to increasethe likelihood of detecting the causative pathogens. Bone/joint sample cultures are far superior to blood culture in identifying the infectious agent. Other newer diagnostic methods include PCR/16S ribosomal RNA or DNA, which can detect bacterial sequences more efficiently and quickly.

Plain radiography is useful after two weeks of evolution when poorly defined lytic lesions can be seen, simulating an aggressive lesion (Figure 47.1). A lamellar periosteal reaction may also occur. Plain radiography performed at onset can rule out a fracture or malignancy (e.g., Ewing's sarcoma). New bone formation may be evident on plain radiographs 3 weeks after symptom onset.

Ultrasound is used to detect lytic and periosteal lesions. CT is not useful in diagnosing acute infection. MRI is the imaging modality of choice. It has significant sensitivity (97%-100%) and specificity (92%). Early changes in osteomyelitis can be detected within the first 2-5 days after the onset of the disease. It detects extraosseous manifestations or complications (pyomyositis, joint effusion, or subperiosteal abscess). It is useful for

planning the surgical approach. Disadvantages include high cost, availability, long scan time, and the need for sedation or anesthesia.

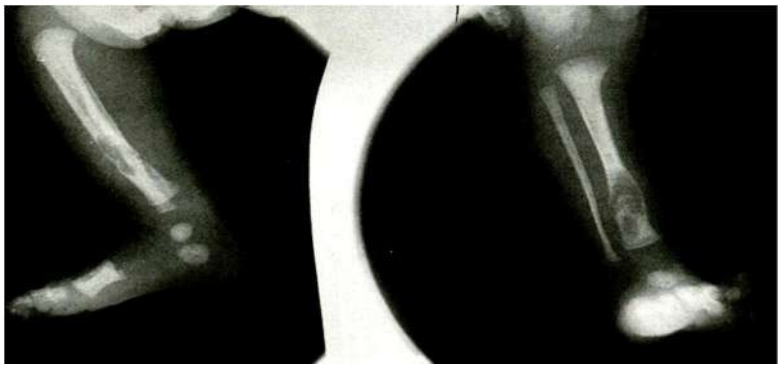


Fig. 47.1. Acute osteomyelitis of the distal tibial metaphysis, X-ray

The **differential diagnosis** includes septic arthritis, cellulitis, osteoid osteoma, acute lymphoblastic leukemia, Ewing's sarcoma, osteosarcoma, bone infarction, sickle cell anemia, Gaucher disease, vitamin A deficiency, avascular necrosis, chronic recurrent multifocal osteomyelitis.

The **treatment** of acute osteomyelitis in children involves pediatricians, infectious disease specialists, orthopedic surgeons, and radiologists.

Antibiotics. β-lactam antibiotics have satisfactory bone penetration and proven efficacy for osteomyelitis. First-generation penicillin or cephalosporins are the antibiotics used as first-line therapy for treating methicillin-sensitive S aureus (MSSA) infection. Beta-lactam agents (ampicillin, ampicillin sulbactam, and cephalosporins) are the drugs of choice for treating acute osteomyelitis caused by K. kingae or Streptococcus pneumoniae. Other alternative agents are daptomycin and linezolid in patients who do not respond to vancomycin. Oxazolidinones and fifth- generation cephalosporins may also be used. In children who are unvaccinated or

incompletely vaccinated in the first 5 years of life, Cefuroxime or a combination of nafcillin or oxacillin or clindamycin with ceftriaxone may be used. In neonatal osteomyelitis and septic arthritis, treatment includes a penicillin stable to penicillinase (Nafcillin or Oxacillin) or Vancomycin in combination with Gentamicin or a third-generation Cephalosporin (cefotaxime); Clindamycin is a good long-term choice.

For uncomplicated acute hematogenous osteomyelitis, a short course of parenteral antibiotics followed by oral therapy for a minimum total duration of 3- 4 weeks is appropriate. Acute hematogenous osteomyelitis complicated by MRSA may warrant prolonged drug therapy.

Surgery. In 90% of cases of acute hematogenous osteomyelitis, appropriate antibiotic therapy is sufficient for cure. Indications for surgical treatment are persistent symptoms (fever, local inflammation) that do not respond to drug treatment. In some complicated cases, surgical incision and drainage (including multiple procedures) may be indicated.

Complications: subperiosteal and intraosseous abscesses, deep vein thrombosis, septic pulmonary emboli, disseminated infection, multiple organ failure, pathological fractures, longer average hospital stay, disruption of bone growth.

Chapter 48. SEPTIC ARTHRITIS

Definition. Septic arthritis is the invasion of a joint by a pathogenic microoganism and can lead to serious consequences if not promptly diagnosed and treated.

Epidemiology. The incidence of bacterial arthritis in children is 5 to 37 cases per 100,000. Children under the age of 3 are more commonly affected.

Etiopathogenesis. The joints most involved are those in which the metaphysis is embedded in the joint capsule: hip, elbow, and shoulder. When bacteria invade a synovial joint, the inflammatory process can cause rapid, severe destruction of articular cartilage. In children, septic arthritis occurs as a complication of hematogenous osteomyelitis. With the metaphysis enclosed in the joint capsule, the initial osteomyelitic infectious process crosses the cortex, involving the joint in the septic process. Thus, in children, the organism that causes septic arthritis is the same as that involved in osteomyelitis.

The infection is usually monoarticular. The bacteria most isolated in children with bacterial arthritis vary according to age, vaccination status, and geographical region. Empirical therapy in children older than three months should be directed toward *Staphylococcus aureus*, the most common germ in this age group, followed by other Gram-positive germs (group A streptococci, *Streptococcus pneumoniae*). Regarding the Gram-negative group, we must consider the fact that, in the last two decades, systemic vaccination against *Hemophilus influenzae* has changed the pattern of bone infection. Currently, *Kingella kingae* is the most often implicated Gram-negative germ.

The **diagnosis** is sometimes difficult to establish. Clinically, the affected children present with altered general condition, appearing lethargic and febrile, along with joint pain and restricted range of motion. The affected joint is tender to palpation. In newborns, a severely altered general condition combined with limited movement and pain during joint mobilization, should raise suspicion for septic arthritis.

Paraclinical diagnostic. The patient shows signs of inflammation: leukocytosis with neutrophilia, increased inflammatory markers. Early X-rays are uncharacteristic.

Since delaying treatment has serious consequences, clinical suspicion of septic arthritis is sufficient to justify emergency aspiration of joint fluid for culture and antibiogram. Until the results are obtained, the patient should be treated with antibiotics.

Treatment. Early treatment saves the joint. Delayed therapy has been associated with long-term sequelae. To prevent rapid degradation of articular cartilage by pyogenic toxins, treatment of septic arthritis is an emergency. In most cases, surgical treatment combined with intravenous antibiotic administration is necessary. Surgical incision of the joint capsule (arthrotomy), drainage, debridement of infected or necrotic tissue, and abundant lavage are performed.

Empirical antibiotic treatment is initiated until the results of the antibiogram are obtained. Cefuroxime or amoxicillin-clavulanic acid is administered to newborns and infants up to 3 months of age. Cefuroxime is administered to infants over 3 months and up to 2 years of age. Cefadroxil is administered for children over 2 years of age.

Complications: arthrosis, epiphyseal necrosis, pathological joint dislocation, growth disorders, limb inequality.

PEDIATRIC UROLOGY

Chapter 49. URETEROPELVIC JUNCTION STENOSIS

Definition. Hydronephrosis is defined as distension of the renal pelvis and calyces, with accumulation of fluid (urine) in these structures. Ureteropelvic junction stenosis is the most common cause of congenital hydronephrosis and is defined as an abnormality of the junction between the renal pelvis and ureter that causes a mechanical and/or functional obstruction in the urine flow (Figure 49.1).

The **incidence** is 1:5000 newborns. It is more common in males 2:1, and 2/3 occur on the left side. Bilateral stenosis is a very rare entity.

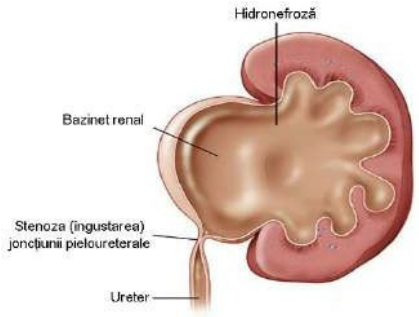


Fig. 49.1. Hydronephrosis due to ureteropelvic junction stenosis

Etiology. During embryogenesis, the kidney and ureter have different origins, and their union occurs at the ureteropelvic junction. At the same time, the ureter goes through a period in which it has no lumen, subsequently recanalizing. If this process is disrupted or delayed, an area of stenosis may occur at the junction between the ureter and the renal pelvis. Another possibility is the presence of an artery that branches off from the renal artery

toward the lower pole of the kidney and crosses the ureter anteriorly on its way to the lower pole of the kidney (polar vessel).

Pathophysiology. The obstruction will lead to increased urine pressure in the kidney, which will result in progressive atrophy of the renal parenchyma. The degree of renal impairment is directly proportional to the severity of the obstruction and its duration. In extreme cases, the affected renal parenchyma may be completely atrophied (dysplastic kidney). In addition, urine stagnation upstream of the obstruction predisposes to urinary tract infection, which has an additional detrimental effect on the renal parenchyma.

Clinical picture. Currently, most cases of hydronephrosis are diagnosed prenatally, so patients are monitored and treated before symptoms are present. The condition may be asymptomatic, and when symptoms are present, they are mild and usually occur late in the course of the disease. Low back pain, a palpable mass in the lumbar region or flank ("phantom tumor"), and recurrent urinary tract infections may occur.

Paraclinical evaluations. Laboratory tests may show impaired renal function or signs of urinary tract infection. Ultrasound is the main imaging method and reveals dilation of the renal pelvis and calyces as well as thinning of the renal parenchyma. In most cases, prenatal ultrasound is used to detect hydronephrosis in the fetus. After birth, ultrasound remains the basic imaging investigation for the evaluation and monitoring of hydronephrosis (Figure 49.1). To assess the severity of obstruction and renal function, renal scintigraphy with Tc-99m DMSA (Dimercaptosuccinic acid) or MAG3 (Mercaptoacetylglycine) is necessary. Computed tomography with contrast medium (URO-CT) or magnetic resonance imaging (URO-MRI) provides more detailed information but they are generally reserved for specific or

unclear situations. A positive diagnostic is established in most cases prenatally based on fetal ultrasound. Postnatally, renal ultrasound and scintigraphy are routinely used.

The **differential diagnosis** includes polycystic kidney, hydronephrosis of other causes (vesicoureteral reflux, ureterovesical stenosis, urethral valves, etc.), solitary renal cyst, renal lithiasis, acquired ureteral stenosis, and renal tumors.

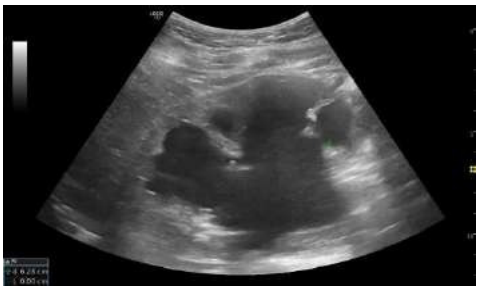


Fig. 49.2. Ureteropelvic junction stenosis (ultrasound)

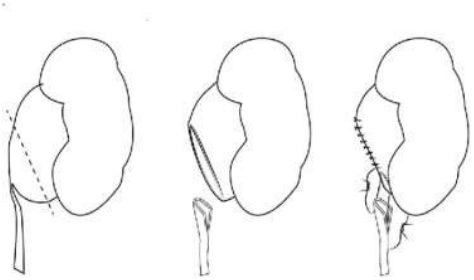


Fig. 49.3. Anderson-Hynes pyeloplasty

Treatment. Treatment plans are based on the severity of the obstruction and of the hydronephrosis. In approximately 75% of prenatally diagnosed cases, the obstruction of the ureteropelvic junction is insignificant and the hydronephrosis does not progress or sometimes resolve spontaneously. These cases are monitored clinically and by ultrasound, and treatment is not necessary.

Treatment of ureteropelvic junction stenosis is surgical and consists of removal of the stenotic portion of the ureter followed by ureteropelvic anastomosis. The gold standard is Anderson-Hynes pyeloplasty (Figure 49.2). The surgical procedure can be performed by open approach or by minimally invasive approach (laparoscopic, retroperitoneoscopic, robotic).

Chapter 50. CONGENITAL MEGAURETER

Definition. Megaureter is defined as the partial or complete dilation of the ureter. A ureter diameter greater than 7 mm in a child is considered megaureter.

Classification

- Obstructive megaureter – ureterovesical junction stenosis
- Refluxing megaureter – vesicoureteral reflux
- Nonobstructive, non-refluxing megaureter

50.1. URETERO-VESICAL JUNCTION STENOSIS

Definition. It represents congenital dilation of the ureter due to obstruction located at the ureterovesical junction.

Incidence. It is the second most common cause of congenital hydronephrosis, with a prevalence of approximately 1:10,000 newborns. It is four times more common in males.

Etiopathogenesis. It occurs due to a disorder in the formation of the distal portion of the ureter, leading to the formation of an adynamic, non-functional portion of vesical segment of the ureter. This area of stenosis has an obstructive effect on the passage of urine from the ureter to the bladder.

Clinical picture. Most cases are diagnosed prenatally and treated before symptoms appear. Postnatally, most of the cases manifests as urinary tract infections with fever, polyuria, low back pain, pyuria, and hematuria.

Paraclinical picture. Laboratory tests show changes when urinary tract infections occur leukocytosis with neutrophilia, increased inflammatory markers, abnormal ur

ine test, positive urine cultures.

Ultrasound is the basic imagistic exam used pre- and postnatally. Most cases are diagnosed prenatally by ultrasound. Postnatal ultrasound reveals hydronephrosis with a dilated (>7 mm) and sinuous ureter (Figure 50.1). Renal scintigraphy is useful for assessing the severity of obstruction and the condition of the renal parenchyma. Uro-CT or uro-MRI provide more detailed images but are reserved for unclear situations. Cystography is useful for differentiating between obstructive and reflux megaureter.

Positive diagnosis. Most cases are diagnosed prenatally by ultrasound (80%). Postnatally, the diagnostic is usually made by ultrasound in a clinically suggestive context. The diagnostic is completed by renal scintigraphy with MAG3 or DTPA, which highlights the obstruction.

The **differential diagnosis** includes refluxing megaureter or other types of low ureteral obstructions: ureterocele, ectopic ureter, ureteropelvic duplication. It is also important to differentiate from secondary obstructive megaureter in urethral valves or neurogenic bladder.

Treatment. In approximately 50% of prenatally diagnosed cases, spontaneous resolution occurs within the first year of life. Ureteral dilatation and the degree of hydronephrosis are monitored by ultrasound immediately after birth, and prophylactic antibiotic treatment may be administered in minimal doses over a prolonged period ("a la long") to prevent urinary infection. Surgical treatment is indicated in patients with significant obstruction on renal scintigraphy and/or in whom no improvement in ureteropelvic dilatation is observed on dynamic ultrasound examination.

Classic surgical treatment involves removal of the stenotic portion and antireflux reimplantation of the ureter into the bladder by open or laparoscopic approach. Alternatively, endoscopic dilation and stenting of the

ureter can be performed. Recent studies have demonstrated the efficacy of endoscopic treatment, which is currently preferred as a first-line treatment before surgical treatment.

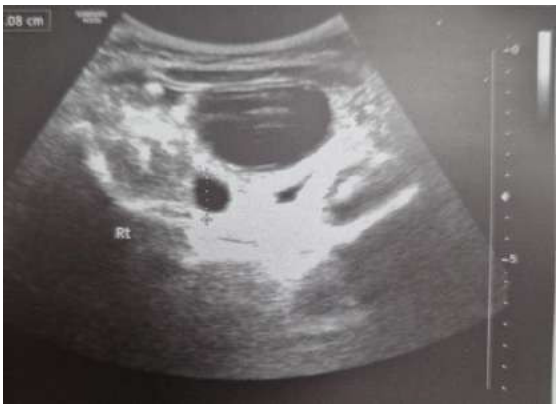


Fig. 50.1. Obstructive megaureter (ultrasound)

50.2. VESICOURETERAL REFLUX

Definition It is the reflux of urine from the bladder into the ureter and then upward to the pelvis and pyelocaliceal system.

The **incidence** varies between 0.4–2%. Between 25–50% of children who develop urinary tract infections have vesicoureteral reflux.

Etiopathogenesis. The antireflux mechanism of the ureterovesical junction is normally maintained by an oblique trajectory of the ureter through the bladder wall, the circular arrangement of muscle fibers around the ureteral orifice, and the slit-like shape of the ureteral orifice.

Vesicoureteral reflux can be:

- Primary reflux caused by a primary deficiency of the antireflux mechanism, not associated with other renal-urinary anomalies or malformations

- Secondary reflux occurs consecutively to other congenital anomalies or malformations:
    - Abnormalities in the implantation of the ureter in the bladder, ectopic ureter, ureterocele
    - Ureteral duplications, bladder diverticula
    - Malformations that cause increased pressure in the bladder: neurogenic bladder, bladder neck hypertrophy, urethral valves, urethral strictures, urethral meatus strictures.

Urinary reflux into the ureter has two consequences:

- Stagnation of urine in the urinary system, which predisposes to urinary tract infection
- Increased pressure in the collecting system during urination

Both urinary tract infection and increased pressure in the upper urinary tract have a detrimental effect on the renal parenchyma, causing damage to the renal parenchyma.

Pathological anatomy. Vesicoureteral reflux can be unilateral or bilateral and is classified as 5 degrees (Figure 50.1)

❖ Grade I – reflux in the pelvic portion of the ureter, without dilation

❖ Grade II – reflux in the ureter and pyelocaliceal system, without dilation.

❖ Grade III – reflux in the ureter and pelvicalyceal system, with mild or moderate sinusoidal dilatation of the ureter.

❖ Grade IV – reflux in the ureter and pelvicalyceal system, with dilatation of the renal pelvis and calyces.

❖ Grade V – reflux into the ureter and pyelocaliceal system with tortuous ureter, appearing as megaureter. The papillary structure in most calyces is not visible.

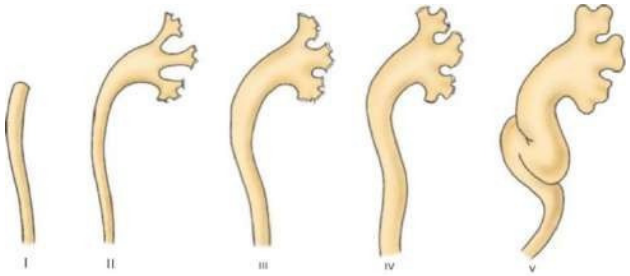


Fig. 50.1. Staging of vesicoureteral reflux

Clinical picture. The main characteristic of vesicoureteral reflux is its association with urinary tract infection. Urinary infections might be severe (urosepsis) and, although treated correctly, recur frequently. Patients have fever, polyuria, oliguria, and nitrogen retention.

Paraclinical investigations. Laboratory tests performed in the acute phase indicate signs of urinary tract infection: leukocytosis with neutrophilia, elevated urea and creatinine levels. Urinalysis indicates the presence of leukocytes, bacteria, red blood cells, and positive nitrites. Urine culture is necessary to identify the germ responsible for the urinary tract infection and for antibiogram.

Abdominal ultrasound may reveal, depending on the severity of the reflux: dilatation of the ureter +/- renal pelvis +/- calyces. In low-grade I or II reflux, the ultrasound may be normal. Fetal ultrasound shows dilation of the urinary tract but cannot determine the etiology of hydronephrosis. The gold standard for the diagnostic of vesicoureteral reflux is cystography (Figure 50.2). Cystography reveals reflux and determines its degree. Recently, cystography has been replaced by contrast-enhanced ultrasound (Sonoview). Renal scintigraphy is also useful because it shows the degree of renal scarring.

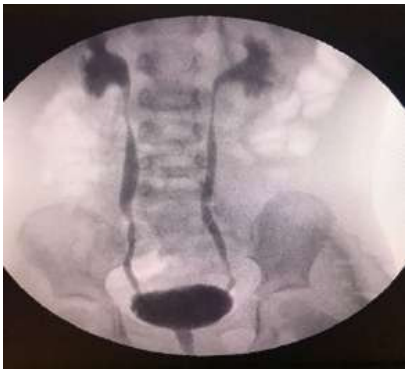


Fig. 50.2. Bilateral vesicoureteral reflux (cystography)

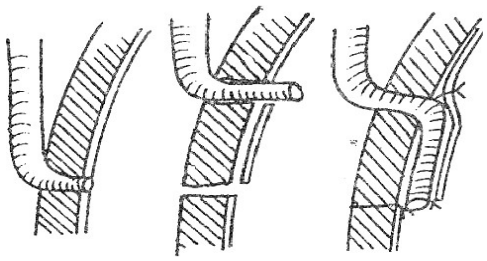


Fig. 50.3. Ureterovesical reimplantation

Positive diagnosis. Vesicoureteral reflux should be suspected whenever a child has urinary tract infections that, despite being treated correctly, recur more than 2-3 times a year. The diagnostic is confirmed by cystography.

The **differential diagnosis** includes obstructive megaureter, ureterocele, ectopic ureter, ureteropelvic duplication. It is also important to differentiate between primary and secondary reflux.

Treatment. The therapeutic approach is dictated by the etiology of the reflux (primary or secondary), the degree of reflux, the age and sex of the patient, and the presence of symptoms (urinary tract infection).

Cases with mild or moderate primary reflux that is non-progressive and asymptomatic do not require treatment. Patients are followed up by regular clinical exams, urine analyses, and ultrasound. The “wait and see strategy” is preferred because it has been observed that over time the ureterovesical junction matures and the reflux ameliorate and heals.

Medical treatment aims to treat and prevent urinary tract infections. It consists of specific antibiotic therapy according to the antibiogram,

antipyretic, analgesic, anti-inflammatory, hydro-electrolytic and acid-base rebalancing in severe infections.

In cases of mild to moderate primary reflux (I, II, or III) with symptoms, endourological treatment can be performed using by cystoscopic approach. The ureteral ostium is recalibrated by injecting a silicone-based substance (bulging agent) into the submucosa of the ureteral orifice.

Surgical treatment is indicated in cases of severe reflux, symptomatic reflux (urinary tract infection) and reflux that cannot be controlled by medical means. Reimplantation of the ureter into the bladder is performed, creating an antireflux mechanism by connecting the ureter to the bladder through a submucosal tunnel (Cohen, Leadbetter- Politano, Lich-Gregoire)., (Figure 50.3).

In secondary vesicoureteral reflux, treatment addresses the underlying condition causing the reflux. Depending on the severity and consequences of the reflux, medical, endourological, or even surgical treatment may be necessary.

Chapter 51. DUPLEX COLLECTING SYSTEM

Definition. This is a congenital anomaly consisting of the presence of two urinary collecting systems (ureter, renal system) on the same side.

Incidence. It is the most common anomaly affecting the ureter, present in 0.7% of the population and in 2-4% of the patient getting medical attention for symptoms related to the reno-urinary system. Both sides are affected equally and are bilateral with 15% of cases. It occurs more frequently in females at a ratio of 2:1.

Etiopathogenesis. The development of the ureter begins in the fourth week of gestation when the ureteric bud arises from the Wolffian duct. The bud elongates in the cranial and caudal directions, giving rise to the ureter, renal pelvis, calyces, and collecting duct system. In the case of the double collecting system, two such buds appear and develop from the Wolffian duct.

Anatomical pathology. Duplications can be complete or incomplete, unilateral or bilateral. In incomplete cases, the common ureter can be distal or proximal, i.e., Y-shaped or inverted Y-shaped. In complete duplications, there are essentially two separate collecting systems: two renal pelvises, two ureters that open separately into the bladder. In this case, according to Weigert-Mayer's law, the ureter of the lower moiety opens more cranially and laterally than the lower one in the urinary bladder (Figure 51.1).

Clinical picture. Duplication may be completely asymptomatic or may have structural and functional consequences on the reno-urinary system: vesicoureteral reflux, ureterovesical strictures, ureterocele, or ectopic opening of the ureter into the bladder, urethra, vagina, or prostate. The clinical picture is variable, depending on the type of anomaly, and may include: hydronephrosis, obstruction by stones, urinary tract infections, or progressive impairment of renal function.

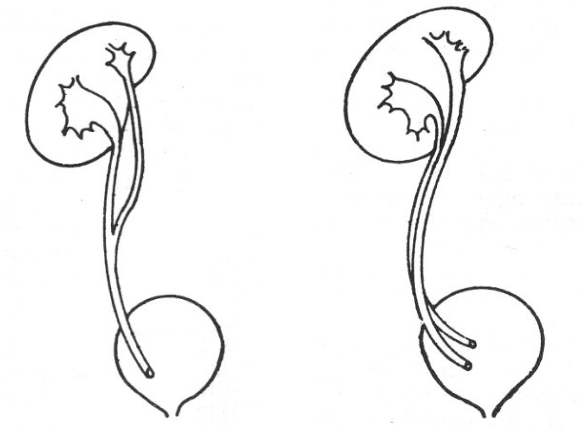


Fig. 51.1. Left: incomplete ureteral duplication;
Right: complete ureteral duplication

Paraclinical picture. Laboratory tests are abnormal in cases of urinary tract infections or impaired renal function.

Prenatal ultrasound can diagnose hydronephrosis but does not usually provide additional information. Ultrasound is the basic imaging test that can reveal the degree of hydronephrosis, clinical form, condition of the renal parenchyma, presence of stones, etc. To accurately determine the anatomical-pathological form of duplication and its consequences, additional imaging investigations are necessary: cystography, renal scintigraphy with DMSA (dimercaptosuccinic acid). Uro-MRI or uro-CT provides more detailed information about the anomaly.

D**ifferential diagnosis** includes renal-urinary malformations that result in the symptoms mentioned above: obstructive megaureter, ureterocele, vesicoureteral reflux, ureteropelvic junction stenosis, etc.

Treatment. Asymptomatic patients do not require treatment. Treatment is differentiated according to symptoms and addresses the consequences rather than the duplication itself (reflux, obstruction, ureteral ectopia, etc.). It may include medical and/or surgical measures.

Chapter 52. URETEROCELE

Definition. It represents a cystic dilatation of the distal, intravesical portion of the ureter. The ureteral meatus is stenotic and has an obstructive effect on the overlying urinary system.

The **incidence** is 1:5000-12000 children and is more common in females. It is very often associated with the double collecting system and ureteral meatus ectopia.

Etiopathogenesis. Ureterocele occurs because of incomplete resorption of the Chawla ureteral membrane. This membrane is a structure that temporarily separates the urogenital sinus from the ureter during the embryonic period.

Pathological anatomy. At the distal, intravesical segment of the ureter, there is a thin-walled, membranous dilatation that bulges inside the bladder. There is a small, stenotic orifice on the ureterocele membrane (ureteral meatus). The suprajacent urinary tract is dilated due to obstruction. Ureterocele is very often associated with ureteral duplications.

Clinical picture. Patients may show signs of urinary tract infection due to stasis: polyuria, fever, or urinary retention when the ureter is very large and obstructs the bladder neck. Stones may form inside the ureterocele. When it is very large, it may prolapse through the urethra in girls and become visible.

Paraclinical picture. Laboratory tests show suggests urinary tract infection. Fetal ultrasound can establish the diagnostic of hydronephrosis and sometimes identify intravesical dilatation. Postnatally, ultrasound is the investigation of choice, highlighting the intravesical ureterocele as well as the dilatation of the overlying urinary tract and the condition of the renal

parenchyma. Cystography reveals an intravesical lacuna. When the ultrasound diagnostic is unclear (duplex system, ectopic ureter), additional investigations such as uro-MRI or uro- CT are necessary (Figure 52.1).

The **differential diagnosis** includes any situation that leads to dilation of the urinary tract: vesicoureteral reflux, ureterovesical stenosis, ectopic ureter, duplications or intravesical tumors, bladder malformations, bladder diverticula.

Treatment. Medical treatment refers to the treatment and prevention of urinary tract infections. The first line of treatment is endourological treatment (Figure 52.1). Through a cystoscopic approach, the ureterocele membrane is fenestrated using either electrocautery or a surgical laser. This decompresses the dilatation. If endourological treatment is not effective or if vesicoureteral reflux occurs after fenestration (window too large), surgical treatment is necessary – ureteral reimplantation.

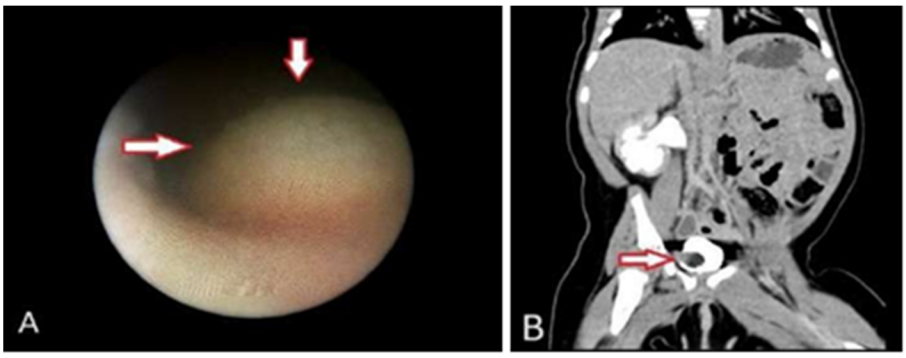


Fig. 52.1. Ureterocele. A: cystoscopic image; B: uro-CT

Chapter 53. BLADDER EXTROPHY

Definition. Bladder exstrophy is a complex congenital malformation characterized by the absence of the anterior abdominal wall in the sub-umbilical region and the absence of the anterior wall of the bladder and urethra.

The **incidence** is 1:25,000-50,000 newborns, with a male: female ratio of 4:1.

Etiopathogenesis. It is due to a defect in the formation or rupture of the anterior cloacal membrane.

Pathological anatomy. The sub-umbilical anterior abdominal wall together with the anterior wall of the bladder are missing (Figure 53.1).

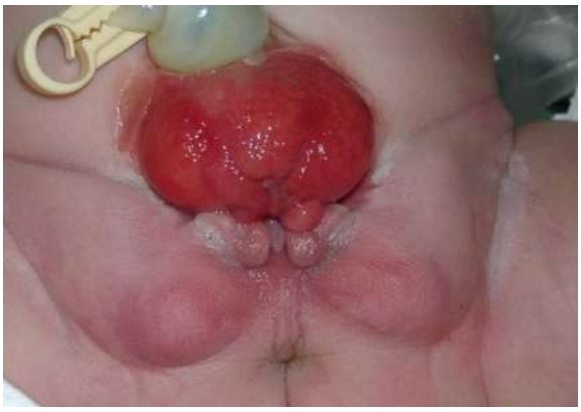


Fig. 53.1. Bladder exstrophy in a female newborn

The urinary bladder continues laterally with normal skin, and the umbilicus is inserted at the upper pole. At the level of the bladder wall, the two urethral orifices are visible, and the bladder mucosa exposed to the external environment becomes edematous, fibrotic, presents polyps, and may become metaplastic. Distally, the bladder wall continues with the urethra, which lacks an anterior wall along its entire length. In girls, a bifid clitoris

appears, the vaginal introitus is directed anteriorly, and there is often a degree of lower vaginal stenosis. In boys, the penis presents epispadias: the penis is open like a book towards the rear, the corpora cavernosa diverges in the proximal portion and are shortened. The pelvis shows pubic diastasis with posterior rotation of the pelvic bones by $12–18^0$.

Positive diagnostic. Ultrasound can diagnose the condition starting at 18–20 weeks of intrauterine life. For more detailed images, fetal MRI can be performed. At birth, the malformation is clinically evident. Additional investigations are necessary to assess the presence of associated conditions and for planning the treatment. Other conditions such as umbilical hernia or omphalocele, anorectal malformation, inguinal hernia, or cryptorchidism may also be present.

Treatment is surgical and involves closing the bladder, bladder neck, and urethra, as well as repairing the pubic symphysis and anterior abdominal wall. It is essential to close the bladder as early as possible, preferably within the first 72 hours after birth, because over time the sacroiliac joints become rigid, and the pelvic bones can no longer be mobilized to repair the pubic symphysis. In girls, this can be done in a single stage. In boys, staged treatment is preferred, with the first stage involving the reconstruction of the bladder, bladder neck, pubic symphysis, and urethra down to the base of the penis. Subsequently, at 6-12 months, the epispadias is corrected.

Prognosis depends on the severity of malformation and the promptitude of treatment. Usually, the bladder has reduced capacity, and the patient has a degree of incontinence. Sexual function may be affected, especially in males.

Chapter 54. POSTERIOR URETHRAL VALVES

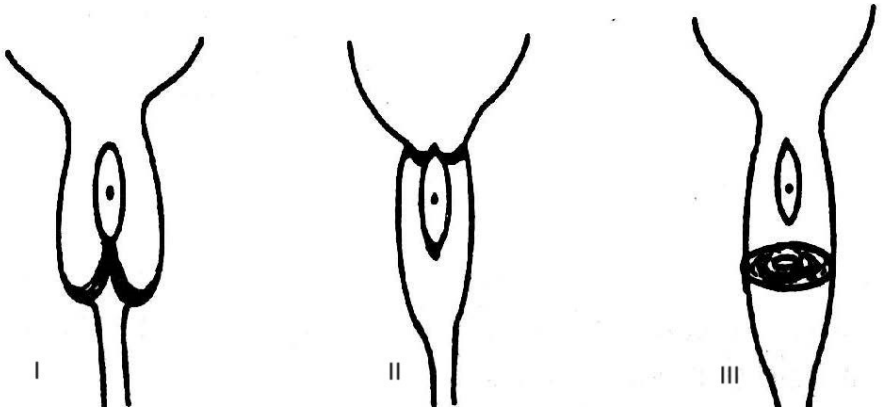
Definition. Posterior urethral valves are abnormal membranous folds in the posterior urethra that create a subvesical obstruction to urine flow.

Epidemiology. Posterior urethral valves occur exclusively in males with an incidence of 1:5000-8000 newborns. They are the most common cause of subvesical obstruction in boys.

Etiopathogenesis. The origin of urethral valves is in the embryonic period, and they occur due to an abnormality in the insertion of the mesonephric duct into the cloaca (too anterior).

Pathological anatomy. Young's classification describes three types (Figure 54.1):

- Type I (90-95%): Folds of the urethra starting immediately below the verumontanum, extending laterally and inferiorly, joining anteriorly
- Type II: Folds, not very prominent, starting from the verumontanum and ascending towards the bladder neck (not obstructive)
- Type III: Diaphragm with central perforation located either above or below the verumontanum



54.1. Posterior urethral valves, Young classification

The severity of subvesical obstruction caused by posterior urethral valves can range from mild obstruction with no impact on the urinary system to severe obstruction with severe bilateral hydronephrosis and significant impairment of the structure and function of the entire renal-urinary system, even during intrauterine life.

The **clinical picture** depends on the severity of the obstruction. The disease is suspected in male newborns from a pregnancy with oligohydramnios, and the clinical picture can take several forms:

- Severe form – newborn and infants have urine retention, fever, palpable bladder and kidneys (trefoil abdomen), chronic renal failure, respiratory failure due to respiratory hypoplasia
- Moderate form in older children: dysuria, weak urine stream, overflow urination, low back pain, bladder globe
- Late form (more difficult to diagnose): discrete obstructive signs, recurrent urinary tract infections

Paraclinical picture. Posterior urethral valves should be suspected in any male fetus presenting with bilateral ureterohydronephrosis, full bladder with thick walls, or oligohydramnios. The characteristic sign on fetal ultrasound is the "keyhole sign" (Figure 54.2).

After birth, ultrasound will show a bladder with thickened walls, bilateral ureterohydronephrosis, and dilated proximal urethra. The gold standard imaging investigation is voiding cystography. The proximal urethra is dilated with a sudden decalibration resembling a "swallow's nest" (Figure 54.3).

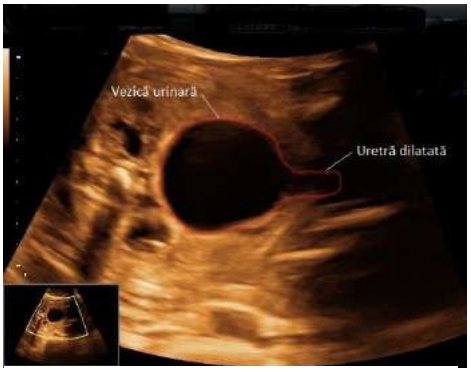


Fig. 54.2. Prenatal ultrasound. The "keyhole sign"

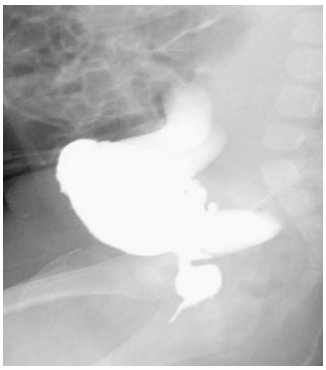


Fig. 54.3. Posterior urethral valves (cystography)

Laboratory tests can detect, depending on the clinical form: nitrogen retention (increased creatinine, urea), signs of urinary tract infection, anemia, and electrolyte imbalances due to renal failure.

A positive diagnosis is established by prenatal ultrasound in most cases. Postnatal voiding cystography is the gold standard investigation.

The differential diagnosis includes neurogenic bladder, Marion disease, bilateral vesicoureteral reflux, secondary stenosis, urethral polyps, urethral meatal stenosis, and phimosis.

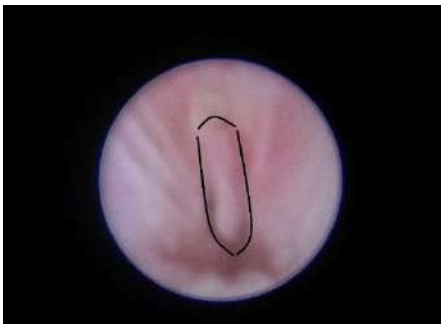


Fig. 54.4. Young I urethral valves, endoscopic view

Treatment. Prenatal diagnosis is essential for appropriate therapeutic management. In cases of severe urinary obstruction diagnosed prenatally, treatment must be initiated during intrauterine life to save the kidneys. A

drainage tube is implanted in the fetus through the abdominal wall into the bladder so that urine drains directly into the amniotic sac, bypassing the obstruction. However, the procedure is not without risks and is only preferred in carefully selected cases.

Postnatally, the treatment of choice for posterior urethral valves is endoscopic ablation of the valves (Figure 54.4). When endoscopic ablation is not possible (lack of appropriate equipment, small urethra size in premature newborns, serious associated diseases), a temporary bladder drainage procedure (Foley catheter, cystostomy) is preferred, and valve ablation is performed later when is possible.

In addition to etiological treatment (valve ablation), therapeutic measures targeting the consequences of the valves are necessary: treatment and prevention of urinary tract infection (proper hygiene and nutrition, antibiotic therapy, and long-term prophylaxis); treatment of hydronephrosis and associated vesicoureteral reflux; treatment of associated renal failure.

The **prognosis** of patients with posterior urethral valves is directly influenced by the severity of the obstruction. The severe form in infants with severe obstruction and significant destruction of the renal parenchyma often has an unfavorable outcome. Chronic renal failure often develops, requiring kidney transplantation. Moderate and tolerated forms generally have a favorable outcome once the obstruction has been removed.

Chapter 55. HYPOSPADIAS, EPISPADIAS

55.1. HYPOSPADIAS

Definition. Hypospadias is a congenital malformation of the penis in which the opening of the urethra is not in its physiological position at the tip of the glans but is ectopic on the anterior (ventral) surface of the penis, at the level of the scrotum or perineum (Figure 55.1).

The **incidence** is 1:250–500 newborns.

Etiopathogenesis. The penis develops between weeks 7 and 14 of gestation. First, the genital tubercle appears in the anterior portion of the urogenital sinus. The urethral meatus is initially located in the perineum, but as the penis develops, the urethral plate elongates and progressively tabularizes until it reaches the tip of the glans in week 16. In hypospadias this process is disturbed, and the development of the urethra is stopped.

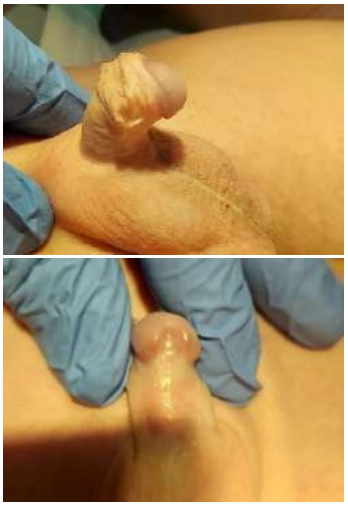
Pathological anatomy. Three pathological changes to the penis are described:

- The urethral meatus is located on the anterior (ventral) surface of the penis away from the tip of the glans, often being stenotic.
- Proximal to the ectopic urethral meatus, there is a fibrous cord that causes the penis to curve anteriorly (chordee).
- The foreskin is not circular, being interrupted anteriorly and crowded on the dorsal side of the penis

Hypospadias is classified according to the location of the urethral meatus (Figure 55.2).

Positive diagnostic. Malformation is evident at birth, with prenatal ultrasound diagnostic being rare. No further investigations are necessary.

The **differential diagnostic** is considered in proximal hypospadias (scrotal, perineal) which ca be confused with disorders of sex differentiation.



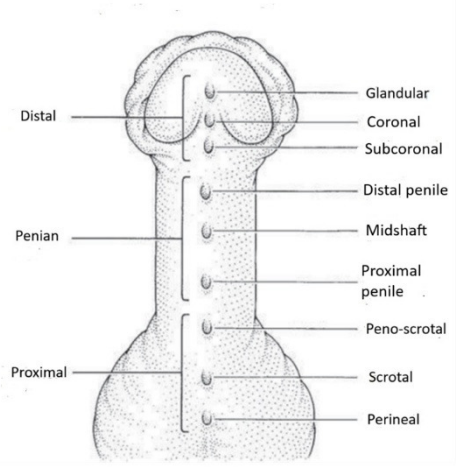


Fig. 55.1. Penile hypospadias **Fig. 55.2.** Classification of hypospadias

Treatment is surgical. The ideal age for hypospadias correction is 6-12 months. Treatment should consider the following objectives:

- Straightening the penis and achieving a straight penis
- Bringing the urethral meatus to the tip of the penis
- Forming a symmetrical, conical glans
- Construction of a new urethra of sufficient and constant caliber
- Achieving an optimal cosmetic appearance, including the foreskin

There is a wide variety of surgical techniques, the choice of procedure depending on the clinical form. In distal forms (glandular, penile), hypospadias correction is performed in a single stage, while proximal forms usually require two stages.

Complications. Frequently, after hypospadias correction, urethral fistulas or urethral strictures may form.

55.2. EPISPADIAS

This is a congenital malformation characterized by ectopic opening of the urethral meatus on the dorsal surface of the penis. It is a rare malformation that can occur alone or in association with bladder exstrophy (Figure 55.3).

Pathological anatomy. Three forms are described: glanular, penile, and penopubic (complete). Distal to the urethral opening, the corpora cavernosa opens posteriorly like a book. Treatment is surgical and is ideally performed at 6-12 months of age.

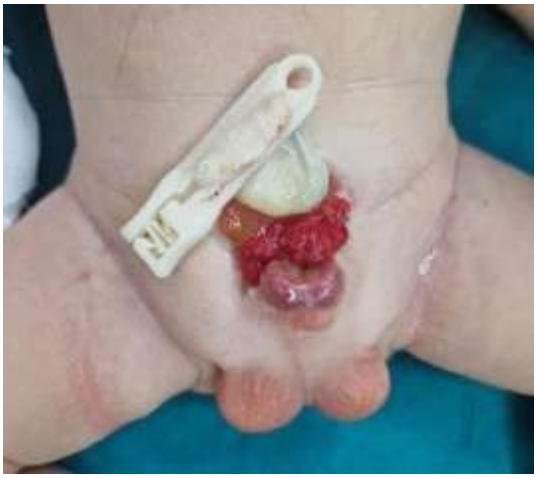


Fig. 55.3. Epispadias and bladder exstrophy

Chapter 56. PHIMOSIS. PARAPHIMOSIS.

56.1. PHIMOSIS

Definition. Phimosis is the inability to retract the foreskin due to stenotic narrowing of the foreskin opening.

Incidence. Approximately one-third of boys have physiological phimosis until the age of 3 years.

Etiopathogenesis. At birth, foreskin retraction is possible in <5% of boys, as the foreskin is physiologically adherent to the glans (balanopreputial adhesions). Retraction and epithelialization of the foreskin occur gradually as a natural, progressive process, and is completed by the age of 3-5 years.

Phimosis can be congenital, when the foreskin is narrowed from birth and this narrowing persists in older children, preventing normal retraction of the penis.

A significant percentage of phimosis cases are acquired, with narrowing of the foreskin caused by a local scarring process (scarring phimosis) following local infections (acute or chronic balanitis) or trauma (untimely, aggressive retraction at an early age, Figure 56.1).

Pathological anatomy. The preputial orifice is narrowed, making retraction of the glans impossible or difficult. Sometimes, a ring of fibrous, inextensible tissue can be observed at the preputial orifice, which cracks during retraction maneuvers.

Clinical picture. The inability to retract the foreskin makes proper local hygiene difficult, which predisposes to local infections (balanitis) or urinary tract infections. In adolescents and adults, phimosis can cause sexual dysfunction. Sometimes pain may occur in the penis, especially when it is

erect. Very rarely, the foreskin opening is so narrow that the child has difficulty urinating.

Positive diagnostic is based on clinical examination: the inability to retract the foreskin, revealing the narrowing of the foreskin opening.

Differential diagnosis includesbalanoposthitis adhesions. Foreskin is adherent to the glans making the retraction impossible.

Treatment. Phimosis in young children, before the age of 3-5, is considered physiological and does not require surgical treatment. Parents are instructed to perform gentle, progressive foreskin retraction daily, followed by cleaning of the glans.

Surgical treatment is recommended for patients in whom foreskin retraction maneuvers have been unsuccessful or for those with cicatricial phimosis. There are two surgical options: Duhamel dorsal preputioplasty (Figure 56.2) or circumcision. Circumcision is preferred in patients with cicatricial phimosis.

56.2. PARAPHIMOSIS

Paraphimosis is a complication of phimosis and occurs when the phimotic foreskin has been retracted, becomes trapped in the coronal sulcus and can no longer be pulled back over the glans. The foreskin acts as a tourniquet on the penis and can lead to ischemia of the glans. Therefore, paraphimosis is an emergency requiring urgent manual reduction (Figure 56.3). If manual reduction is not successful, emergency surgery is required.

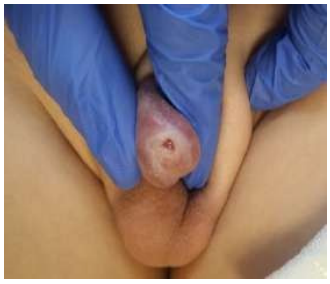


Fig. 56.1. Scarring phimosis

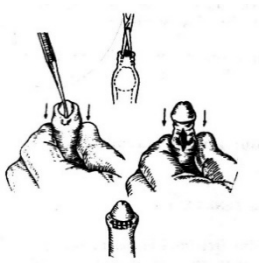


Fig. 56. 2. Preputioplasty

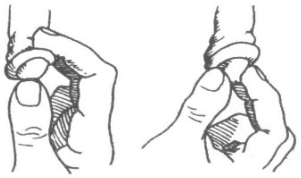


Fig. 56.3. Reduction of paraphimosis

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